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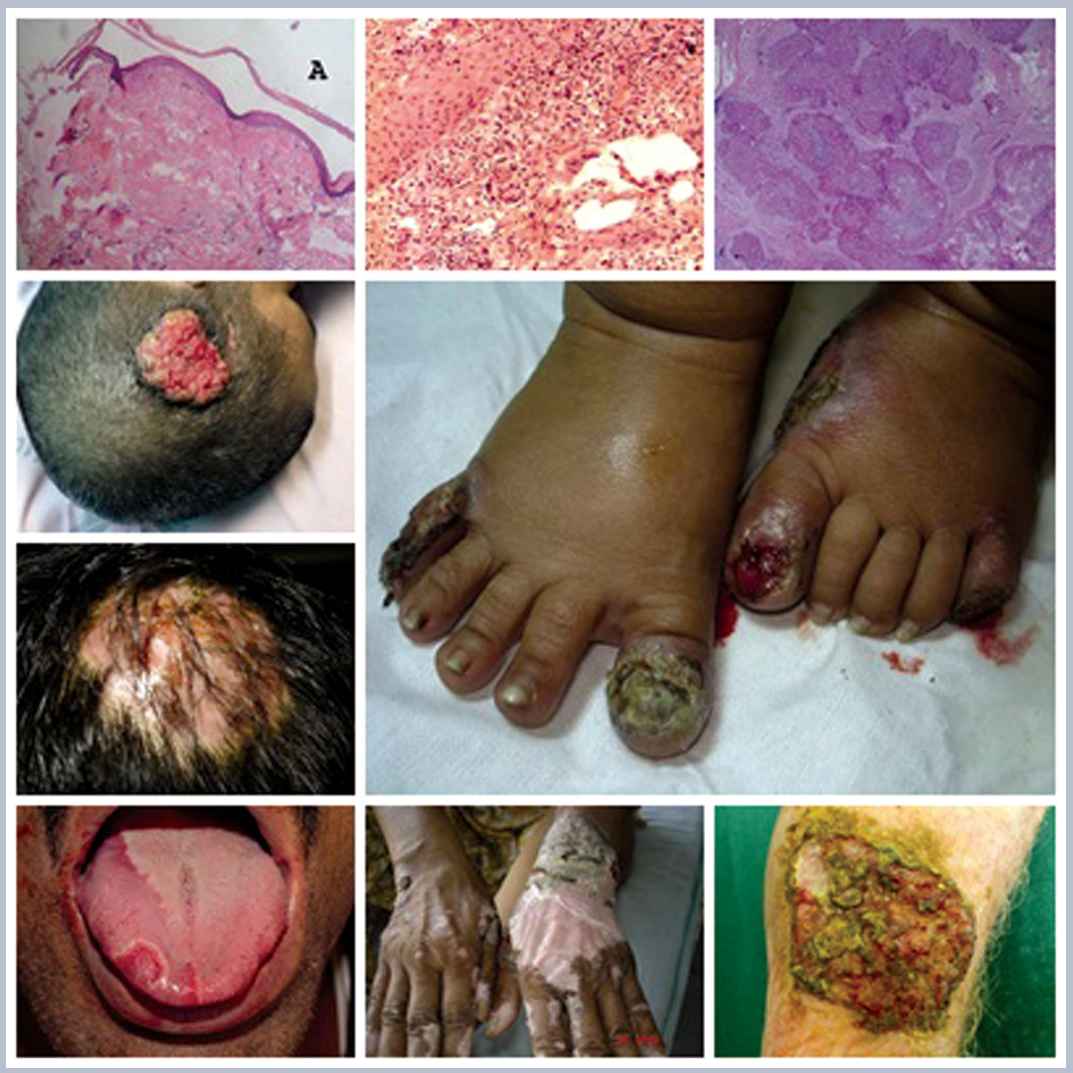

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Editorial Pages

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Editorial Board:

Grupa **Polpharma** dla dermatologii

Flucofast (*Fluconazolum*). **Skład i postać:** 1 kapsułka zawiera 50 mg, 100 mg lub 150 mg flukonazolu. Produkt leczniczy Flucofast 100 mg zawiera laktozę oraz żółcień pomarańczową (E 110). **Wskazania do stosowania:** Flucofast jest wskazany w leczeniu zakażeń grzybiczych. Flucofast jest wskazany do stosowania u pacjentów dorosłych w leczeniu następujących zakażeń: kryptokokowe zapalenie opon mózgowych; kokcydioidomikoza; inwazyjne kandydozy; drożdżakowe zakażenia błon śluzowych, w tym zakażenia gardła, przełyku, występowanie drożdżaków w moczu oraz przewlekłe drożdżakowe zakażenia skóry i błon śluzowych; przewlekłe zanikowe drożdżakowe zapalenie jamy ustnej (związane ze stosowaniem protez zębowych), jeśli higiena jamy ustnej lub leczenie miejscowe są niewystarczające; drożdżyca pochwy, ostra lub nawracająca, gdy leczenie miejscowe jest niewystarczające; drożdżakowe zapalenia żołędzi, gdy leczenie miejscowe jest niewystarczające; grzybice skóry, w tym stóp, tułowia, podudzi, łupież pstry, zakażenia drożdżakowe skóry właściwej, gdy zalecane jest podjęcie leczenia ogólnoustrojowego; grzybica paznokci (onychomikoza), gdy uzna się, że inne leki są nieodpowiednie. Flucofast jest wskazany do stosowania u pacjentów dorosłych w zapobieganiu następującym zakażeniom: nawroty kryptokokowego zapalenia opon mózgowych u pacjentów z podwyższonym ryzykiem nawrotów; nawroty drożdżakowego zapalenia błony śluzowej jamy ustnej, gardła i przełyku u pacjentów zakażonych HIV, u których jest zwiększone ryzyko nawrotów; nawroty drożdżycy pochwy (4 lub więcej zakażeń w ciągu roku); zakażenia grzybicze u pacjentów z przedłużającą się neutropenią (np. u pacjentów z nowotworami krwi, otrzymujących chemioterapię lub u pacjentów po przeszczepieniu krwiotwórczych komórek macierzystych). Flucofast jest wskazany do stosowania w następujących zakażeniach u noworodków, niemowląt, dzieci i młodzieży w wieku od 0 do 17 lat. Flucofast stosuje się w leczeniu drożdżakowego zakażenia błon śluzowych (jamy ustnej, gardła i przełyku), inwazyjnej kandydozy i kryptokokowego zapalenia opon mózgowych oraz w zapobieganiu zakażeniom drożdżakami u pacjentów z osłabioną odpornością. Flucofast można stosować jako leczenie podtrzymujące w celu zapobiegania nawrotom kryptokokowego zapalenia opon mózgowych u dzieci z wysokim ryzykiem nawrotów. Leczenie można rozpocząć przed otrzymaniem wyników posiewu lub innych badań laboratoryjnych. Jednakże po ich otrzymaniu należy odpowiednio dostosować dostępne leczenie. Należy wziąć pod uwagę oficjalne wytyczne dotyczące właściwego zastosowania leków przeciwgrzybiczych. **Dawkowanie i sposób podawania:** Dawkę należy dostosować do rodzaju oraz ciężkości zakażenia grzybiczego. Jeśli w danym zakażeniu konieczne jest stosowanie wielokrotnych dawek, leczenie należy kontynuować do chwili ustąpienia klinicznych lub mikrobiologicznych objawów czynnego zakażenia. Niedostatecznie długi okres leczenia może być przyczyną nawrotu czynnego zakażenia. Kryptokokoza. Leczenie kryptokokowego zapalenia opon mózgowych: Dawka nasycająca: 400 mg pierwszej doby, następna dawka: 200 mg do 400 mg na dobę, zwykle przez 6 do 8 tygodni. W zakażeniach zagrażających życiu dawkę dobową można zwiększyć do 800 mg. Leczenie podtrzymujące w zapobieganiu nawrotom kryptokokowego zapalenia opon mózgowych u pacjentów z podwyższonym ryzykiem nawrotów: 200 mg na dobę. Nieograniczony czas stosowania w dawce dobowej 200 mg. Kokcydioidomikoza: 200 mg do 400 mg przez 11 do 24 miesięcy lub dłużej, w zależności od pacjenta. W niektórych zakażeniach, zwłaszcza w zapaleniu opon mózgowych, można rozważyć zastosowanie dawki 800 mg na dobę. Kandydozy inwazyjne: Dawka nasycająca: 800 mg pierwszej doby. Następna dawka: 400 mg na dobę. Zalecana zwykle długość leczenia zakażenia drożdżakowego krwi wynosi 2 tygodnie po pierwszym negatywnym wyniku posiewu krwi oraz ustąpieniu objawów przedmiotowych i podmiotowych charakterystycznych dla kandydemii. Leczenie kandydozy błon śluzowych. Kandydoza jamy ustnej: Dawka nasycająca: 200 mg do 400 mg pierwszej doby. Następna dawka: 100 mg do 200 mg na dobę przez 7 do 21 dni (do czasu ustąpienia kandydozy jamy ustnej). Można stosować dłużej u pacjentów z ciężkim osłabieniem czynności układu immunologicznego. Kandydoza przełyku: Dawka nasycająca: 200 mg do 400 mg pierwszej doby. Następna dawka: 100 mg do 200 mg na dobę przez 14 do 30 dni (do czasu ustąpienia kandydozy przełyku). Można stosować dłużej u pacjentów z ciężkim osłabieniem czynności układu immunologicznego. Występowanie drożdżaków w moczu: 200 mg do 400 mg na dobę przez 7 do 21 dni. Można stosować dłużej u pacjentów z ciężkim osłabieniem czynności układu immunologicznego. Przewlekła zanikowa kandydoza: 50 mg na dobę przez 14 dni. Przewlekła kandydoza skóry i błon śluzowych: 50 mg do 100 mg na dobę do 28 dni. Można stosować dłużej u pacjentów z ciężkim osłabieniem czynności układu immunologicznego. Zapobieganie nawrotom drożdżakowego zapalenia błony śluzowej u pacjentów zarażonych HIV, u których jest zwiększone ryzyko nawrotów. Kandydoza jamy ustnej: 100 mg do 200 mg na dobę lub 200 mg 3 razy na tydzień. Nieograniczony czas stosowania u pacjentów z przewlekłym osłabieniem czynności układu immunologicznego. Kandydoza przełyku: 100 mg do 200 mg na dobę lub 200 mg 3 razy na tydzień. Nieograniczony czas stosowania u pacjentów z przewlekłym osłabieniem czynności układu immunologicznego. Kandydoza narządów płciowych. Ostra drożdżyca pochwy oraz drożdżakowe zapalenie żołędzi: 150 mg, pojedyncza dawka. Leczenie i zapobieganie nawrotom drożdżycy pochwy (4 lub więcej zakażeń w roku): 150 mg co trzeci dzień, w sumie 3 dawki (doba 1., 4. i 7.), a następnie dawka podtrzymująca 150 mg raz na tydzień. Dawka podtrzymująca przez 6 miesięcy. Grzybice skóry. Grzybica stóp, grzybica tułowia, grzybica podudzi, drożdżyca skóry: 150 mg raz na tydzień lub 50 mg raz na dobę przez 2 do 4 tygodni; w grzybicy stóp może być konieczne stosowanie do 6 tygodni. Łupież pstry: 300 mg do 400 mg raz na tydzień przez 1 do 3 tygodni lub 50 mg raz na dobę przez 2 do 4 tygodni. Grzybica paznokci (onychomikoza): 150 mg raz na tydzień. Leczenie należy kontynuować aż do zastąpienia zakażonego paznokcia przez nowy, niezakażony. Czas potrzebny do odrostu nowego paznokcia dłoni lub stopy wynosi odpowiednio 3 do 6 lub 6 do 12 miesięcy. Szybkość odrostu może jednak różnić się u poszczególnych pacjentów, także w zależności od wieku. Po wyleczeniu przewlekłego zakażenia paznokcie czasami mogą pozostać zniekształcone. Zapobieganie zakażeniom drożdżakowym u pacjentów z przedłużającą się neutropenią: 200 mg do 400 mg. Leczenie należy rozpocząć kilka dni przed spodziewanym początkiem neutropenii i kontynuować przez 7 dni po jej ustąpieniu, kiedy liczba neutrofili zwiększy się powyżej 1000 komórek na mm^3. Szczególne grupy pacjentów: Pacjenci w podeszłym wieku. Dawkowanie należy zmodyfikować w zależności od czynności nerek (patrz „Pacjenci z zaburzeniami czynności nerek"). Pacjenci z zaburzeniami czynności nerek: Jeśli stosuje się pojedynczą dawkę, nie jest konieczna zmiana dawkowania. Pacjentom (w tym dzieciom i młodzieży) z zaburzeniami czynności nerek, otrzymujących wielokrotne dawki flukonazolu, na początku należy podać dawkę od 50 mg do 400 mg, bazując na zalecanej dla danego wskazania dawce dobowej. Po podaniu tej dawki nasycającej dawkę dobową (zgodnie ze wskazaniem) należy ustalić na podstawie poniższych danych: Klirens kreatyniny >50 ml/min – 100% zalecanej dawki. Klirens kreatyniny ≤50 ml/min (bez dializ) – 50% zalecanej dawki. Regularne dializy – 100% zalecanej dawki po każdej dializie. Pacjentom regularnie dializowanym należy po każdej dializie podawać 100% zalecanej dawki; w dniach, w których nie wykonuje się dializy, należy podawać dawkę zmniejszoną odpowiednio do klirensu kreatyniny. Pacjenci z zaburzeniami czynności wątroby: Dane dotyczące stosowania u pacjentów z zaburzeniami czynności wątroby są ograniczone, dlatego flukonazol należy stosować ostrożnie u pacjentów z zaburzeniami czynności wątroby. Dzieci i młodzież. U dzieci i młodzieży nie należy przekraczać maksymalnej dawki dobowej wynoszącej 400 mg. Podobnie jak w zakażeniach u pacjentów dorosłych, długość leczenia zależy od klinicznej oraz mikologicznej odpowiedzi pacjenta. Flucofast podaje się w pojedynczych dobowych dawkach. Dawkowanie u dzieci z zaburzeniami czynności nerek - patrz „Pacjenci z zaburzeniami czynności nerek". Nie przebadano farmakokinetyki flukonazolu u dzieci i młodzieży z niewydolnością nerek (dawkowanie u noworodków (0 do 27 dni), u których często czynność nerek nie jest w pełni rozwinięta – patrz poniżej). Noworodki, niemowlęta i dzieci (w wieku od 28 dni do 11 lat). Kandydoza błon śluzowych. Dawka początkowa: 6 mg/kg m.c. Następna dawka: 3 mg/kg m.c. na dobę. Dawkę początkową można stosować w pierwszym dniu leczenia w celu szybszego osiągnięcia stanu równowagi. Kandydozy inwazyjne oraz kryptokokowe zapalenie opon mózgowych. Dawka: 6 do 12 mg/kg m.c. na dobę, w zależności od ciężkości choroby. Leczenie podtrzymujące w celu zapobiegania nawrotom kryptokokowego zapalenia opon mózgowych u dzieci z dużym ryzykiem nawrotu. Dawka: 6 mg/kg m.c. na dobę, w zależności od ciężkości choroby. Zapobieganie zakażeniom drożdżakami u pacjentów z osłabioną odpornością. Dawka: 3 do 12 mg/kg m.c. na dobę. W zależności od stopnia oraz czasu trwania neutropenii (patrz „Dawkowanie u dorosłych"). Młodzież (w wieku od 12 do 17 lat). W zależności od masy ciała oraz dojrzałości lekarz przepisujący powinien ocenić, które dawkowanie (dla dorosłych czy dla dzieci) jest najbardziej odpowiednie. Dane kliniczne wskazują, że klirens flukonazolu u dzieci jest większy niż u dorosłych. Dawki 100, 200 i 400 mg u dorosłych odpowiadają dawkom 3, 6 i 12 mg/kg m.c. u dzieci, umożliwiającym uzyskanie porównywalnego stopnia narażenia. Nie określono profilu bezpieczeństwa stosowania ani skuteczności flukonazolu w leczeniu kandydozy narządów płciowych u dzieci i młodzieży. Aktualnie dostępne dane dotyczące bezpieczeństwa stosowania u dzieci i młodzieży w innych wskazaniach opisano w punkcie: Działania niepożądane. Jeżeli konieczne jest leczenie kandydozy narządów płciowych u młodzieży (w wieku od 12 do 17 lat), należy zastosować takie samo dawkowanie jak u dorosłych. Noworodki (w wieku od 0 do 27 dni). Noworodki wolniej wydalają flukonazol. Istnieją nieliczne dane farmakokinetyczne potwierdzające sposób stosowania u noworodków. Noworodki (0 do 14 dni): Taką samą dawkę w mg/kg m.c. jak u niemowląt i dzieci należy podawać co 72 godziny. Nie należy przekraczać maksymalnej dawki 12 mg/kg m.c., podawanej co 72 godziny. Noworodki (od 15 do 27 dni): Taką samą dawkę w mg/kg m.c. jak u niemowląt i dzieci należy podawać co 48 godzin. Nie należy przekraczać maksymalnej dawki 12 mg/kg m.c., podawanej co 48 godzin. Flukonazol można podawać w postaci doustnej lub dożylnej (w zależności od preparatu); droga podania zależy od stanu klinicznego pacjenta. W przypadku zmiany drogi podania z dożylnej na doustną i odwrotnie nie jest konieczna zmiana dawkowania. Kapsułki należy połykać w całości, niezależnie od przyjmowanych posiłków. **Przeciwwskazania:** Nadwrażliwość na substancję czynną, pokrewne związki azolowe lub na którąkolwiek substancję pomocniczą produktu. Z badań dotyczących interakcji po podaniu wielokrotnym wynika, że przeciwwskazane jest podawanie terfenadyny pacjentom otrzymującym flukonazol w dawkach wielokrotnych, wynoszących 400 mg na dobę lub więcej. Stosowanie innych leków, które wydłużają odstęp QT i są metabolizowane przez cytochrom P450 (izoenzym CYP3A4), takich jak: cyzapryd, astemizol, pimozyd, chinidyna oraz erytromycyna, jest przeciwwskazane u pacjentów otrzymujących flukonazol. **Ostrzeżenia i zalecane środki ostrożności:** Grzybica skóry owłosionej głowy. Badano stosowanie flukonazolu w leczeniu grzybicy skóry owłosionej głowy u dzieci. Nie wykazano wyższej skuteczności niż gryzeofulwiny, a ogólny odsetek wyzdrowień był mniejszy niż 20%. Dlatego produktu Flucofast nie należy stosować w leczeniu grzybicy skóry owłosionej głowy. Kryptokokoza. Dane dotyczące skuteczności flukonazolu w leczeniu kryptokokozy oraz zakażeń o innych lokalizacjach (np. kryptokokoza płuc lub skóry) są ograniczone, przez co brak dokładnych zaleceń dotyczących dawkowania. Głębokie grzybice endemiczne. Dane dotyczące skuteczności flukonazolu w leczeniu innych postaci grzybicy endemicznej, takich jak parakokcydioidomkoza, sporotrychoza limfatyczno-skórna i histoplazmoza, są ograniczone, przez co brak dokładnych zaleceń dotyczących dawkowania. Nerki. Należy zachować ostrożność podczas podawania flukonazolu u pacjentów z zaburzeniami czynności nerek. Wątroba i drogi żółciowe. Należy zachować ostrożność podczas podawania flukonazolu u pacjentów z zaburzeniami czynności wątroby. Stosowanie produktu Flucofast wiązało się rzadko z ciężkim toksycznym uszkodzeniem wątroby, w tym ze skutkiem śmiertelnym, głównie u pacjentów z ciężkimi chorobami podstawowymi. W przypadkach hepatotoksycznego działania flukonazolu nie obserwowano jednoznacznego związku z całkowitą dawką dobową leku, długością terapii oraz z płcią ani wiekiem pacjentów. Działanie hepatotoksyczne flukonazolu zwykle ustępowało po zaprzestaniu terapii. Jeśli w trakcie leczenia flukonazolem wystąpią zaburzenia wyników badań czynności wątroby, należy dokładnie obserwować, czy u pacjenta nie wystąpi cięższe uszkodzenie tego narządu. Należy poinformować pacjenta, jakie mogą wystąpić objawy świadczące o silnym działaniu na wątrobę (znaczna astenia, jadłowstręt, przedłużające się nudności, wymioty i żółtaczka). Stosowanie flukonazolu należy niezwłocznie przerwać, a pacjent powinien skonsultować się z lekarzem. Układ sercowo-naczyniowy. Stosowanie niektórych azoli, w tym flukonazolu, było związane z wydłużeniem odstępu QT w zapisie elektrokardiograficznym. W badaniach przeprowadzonych po wprowadzeniu produktu do obrotu u pacjentów przyjmujących flukonazol rzadko notowano wydłużenie odstępu QT i zaburzenia rytmu typu torsade de pointes. Dotyczyło to ciężko chorych pacjentów z wieloma czynnikami ryzyka, takimi jak: choroby mięśnia sercowego, zaburzenia elektrolitowe oraz jednoczesne przyjmowanie leków mogących powodować zaburzenia rytmu serca. Należy zachować ostrożność podczas stosowania flukonazolu u pacjentów, u których występują powyższe czynniki ryzyka zaburzeń rytmu serca. Jednoczesne stosowanie innych leków wydłużających odstęp QT oraz metabolizowanych przez cytochrom P450 (CYP3A4) jest przeciwwskazane. Halofantryna. Wykazano, że halofantryna stosowana w zalecanej dawce terapeutycznej wydłuża odstęp QT oraz jest substratem dla izoenzymu CYP3A4. Nie zaleca się jednoczesnego stosowania flukonazolu i halofantryny. Reakcje skórne. Podczas leczenia flukonazolem rzadko obserwowano występowanie skórnych reakcji alergicznych, takich jak zespół Stevensa-Johnsona i martwica toksyczno-rozpływna naskórka. Pacjenci z AIDS są bardziej skłonni do ciężkich reakcji skórnych po zastosowaniu różnych produktów leczniczych. Jeśli u pacjenta z powierzchowną grzybicą, leczonego flukonazolem, wystąpi wysypka, należy przerwać podawanie flukonazolu. Jeżeli wysypka rozwinie się u pacjenta leczonego flukonazolem z powodu inwazyjnego, układowego zakażenia grzybiczego, pacjenta należy uważnie obserwować; w razie wystąpienia zmian pęcherzowych lub rumienia wielopostaciowego należy przerwać podawanie flukonazolu. Nadwrażliwość. Rzadko opisywano przypadki wystąpienia reakcji anafilaktycznej. Cytochrom P450. Flukonazol jest silnym inhibitorem CYP2C9 i umiarkowanym inhibitorem CYP3A4. Ponadto jest również inhibitorem CYP2C19. Należy kontrolować pacjentów przyjmujących jednocześnie flukonazol i leki o wąskim indeksie terapeutycznym metabolizowane z udziałem izoenzymów CYP2C9, CYP2C19 i CYP3A4. Terfenadyna. Należy dokładnie kontrolować pacjentów przyjmujących jednocześnie terfenadynę i flukonazol w dawkach mniejszych niż 400 mg na dobę. Substancje pomocnicze. Kapsułki zawierają laktozę jednowodną, dlatego nie należy ich stosować u pacjentów z rzadko występującą dziedziczną nietolerancją galaktozy, niedoborem laktazy (typu Lapp) lub zespołem złego wchłaniania glukozy-galaktozy. **Działania niepożądane:** Najczęściej zgłaszanymi działaniami niepożądanymi (>1/10) są: ból głowy, ból brzucha, biegunka, nudności, wymioty, zwiększenie we krwi aktywności aminotransferazy alaninowej, aminotransferazy asparaginianowej, fosfatazy alkalicznej oraz wysypka. Podczas stosowania flukonazolu obserwowano i raportowano przedstawione niżej działania niepożądane z następującą częstotliwością: bardzo często (≥1/10), często (≥1/100 do <1/10), niezbyt często (≥1/1000 do <1/100), rzadko (≥1/10 000 do <1/1000), bardzo rzadko (<1/10 000), częstość nieznana (nie może być określona na podstawie dostępnych danych). Zaburzenia krwi i układu chłonnego: Niezbyt często: niedokrwistość. Rzadko: agranulocytoza, leukopenia, trombocytopenia, neutropenia. Zaburzenia układu immunologicznego: Rzadko: anafilaksja. Zaburzenia metabolizmu i odżywiania: Niezbyt często: zmniejszenie łaknienia. Rzadko: hipercholesterolemia, hipertriglicerydemia, hipokaliemia. Zaburzenia psychiczne: Niezbyt często: bezsenność, senność. Zaburzenia układu nerwowego: Często: ból głowy. Niezbyt często: drgawki, parestezje, zawroty głowy, zmiany smaku. Rzadko: drżenie. Zaburzenia ucha i błędnika: Niezbyt często: zawroty głowy pochodzenia obwodowego. Zaburzenia serca: Rzadko: zaburzenia rytmu serca typu torsade de pointes, wydłużenie odstępu QT. Zaburzenia żołądka i jelit: Często: ból brzucha, wymioty, biegunka, nudności. Niezbyt często: zaparcia, niestrawność, wzdęcia, suchość w jamie ustnej. Zaburzenia wątroby i dróg żółciowych: Często: zwiększenie aktywności aminotransferazy alaninowej, aminotransferazy asparaginianowej, fosfatazy alkalicznej we krwi. Niezbyt często: cholestaza, żółtaczka, zwiększenie stężenia bilirubiny. Rzadko: niewydolność wątroby, martwica komórek wątrobowych, zapalenie wątroby, uszkodzenie komórek wątroby. Zaburzenia skóry i tkanki podskórnej: Często: wysypka. Niezbyt często: wysypka polekowa, pokrzywka, świąd, zwiększona potliwość. Rzadko: martwica toksyczno-rozpływna naskórka, zespół Stevensa-Johnsona, ostra uogólniona osutka krostowa, złuszczające zapalenie skóry, obrzęk naczynioruchowy, obrzęk twarzy, łysienie. Zaburzenia mięśniowo-szkieletowe i tkanki łącznej: Niezbyt często: ból mięśni. Zaburzenia ogólne i stany w miejscu podania: Niezbyt często: zmęczenie, złe samopoczucie, astenia, gorączka. Dzieci i młodzież. Profil i częstość występowania działań niepożądanych oraz nieprawidłowych wyników badań laboratoryjnych obserwowanych podczas badań klinicznych z udziałem dzieci i młodzieży, z wyjątkiem stosowania w leczeniu kandydozy narządów płciowych, są porównywalne do obserwowanych u dorosłych. **Podmiot odpowiedzialny:** Medana Pharma S.A., 98-200 Sieradz, ul. Władysława Łokietka 10. Pozwolenie na dopuszczenie do obrotu dla Flucofast 50 mg, Flucofast 100 mg i Flucofast 150 mg odpowiednio nr 4905, nr 14865 i nr 4906, wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. Cena urzędowa detaliczna dla dawki 50 mg x 7 kaps.; 50 mg x 14 kaps.; 100 mg x 7 kaps.; 100 mg x 28 kaps.; 150 mg x 1 kaps. wynosi odpowiednio w PLN: 14,73; 28,43; 29,00; 109,86; 6,32. Kwota dopłaty pacjenta wynosi odpowiednio w PLN: 8,29; 15,54; 16,11; 58,30; 3,56. ChPL: 2012.09.24.

Trioxal (*Itraconazolum*). **Skład i postać:** Jedna kapsułka twarda zawiera 100 mg itrakonazolu. **Wskazania:** Kandydoza sromu i pochwy; łupież pstry; grzybica skóry wywołana przez dermatofity wrażliwe na itrakonazol (Trichophyton spp., Microsporum spp., Epidermophyton floccosum), np. grzybica stóp, grzybica podudzi, grzybica tułowia, grzybica rąk; grzybica paznokci spowodowana dermatofitami lub drożdżakami; kandydoza jamy ustnej; grzybicze zakażenie rogówki; grzybice układowe: histoplazmoza, aspergiloza, kandydoza, kryptokokoza (w tym kryptokokowe zapalenie opon mózgowych), sporotrychoza, parakokcydioidomikoza, blastomikoza, chromomikoza i inne rzadko występujące grzybicze zakażenia układowe. **Dawkowanie i sposób podawania:** Preparat stosuje się doustnie. W celu uzyskania maksymalnego wchłaniania powinien być przyjmowany niezwłocznie po posiłku. Dorośli: Kandydoza sromu i pochwy: 200 mg dwa razy na dobę przez 1 dzień lub 200 mg raz na dobę przez 3 dni. Łupież pstry: 200 mg raz na dobę przez 7 dni. Grzybica tułowia i grzybica podudzi: 100 mg raz na dobę przez 15 dni lub 200 mg raz na dobę przez 7 dni. Grzybica rąk, grzybica stóp: 100 mg raz na dobę przez 30 dni lub 200 mg raz na dobę przez 7 dni. Kandydoza jamy ustnej: 100 mg raz na dobę przez 15 dni. Grzybicze zapalenie rogówki: 200 mg raz na dobę przez 21 dni. Grzybica paznokci: terapia pulsowa – 200 mg itrakonazolu dwa razy na dobę przez 7 dni. Okresy przyjmowania preparatu (pulsy) są oddzielone 3-tygodniową przerwą w przyjmowaniu leku. W leczeniu grzybicy paznokci rąk stosuje się 2 pulsy, w leczeniu grzybicy paznokci stóp – 3 pulsy. Terapia ciągła: 200 mg raz na dobę przez 3 miesiące. Optymalne efekty kliniczne i przeciwgrzybicze osiągane są po 2 do 4 tygodni po zakończeniu leczenia w przypadku infekcji skóry, sromu i pochwy, jamy ustnej i gardła oraz po 6 do 9 miesięcy po zakończeniu leczenia zakażenia paznokci. Grzybice układowe – Aspergiloza: 200 mg raz na dobę przez okres od 2 do 5 miesięcy. Kandydoza: 100-200 mg raz na dobę przez okres od 3 tygodni do 7 miesięcy. W leczeniu obu powyższych typów grzybic należy zwiększyć dawkę do 200 mg 2 razy na dobę, jeśli przebieg choroby jest ostrzejszy lub zmiany są rozsiane. Kryptokokoza: 200 mg raz na dobę przez okres od 2 do 12 miesięcy. Kryptokokowe zakażenie opon mózgowo-rdzeniowych: 200 mg 2 razy na dobę przez okres od 2 do 12 miesięcy; dawka podtrzymująca (zakażenie opon mózg.-rdz.) 200 mg raz na dobę. Histoplazmoza: 200 mg 1 do 2 razy na dobę przez 8 mies. Sporotrychoza: 100 mg raz na dobę przez 3 miesiące. Parakokcydioidomikoza: 100 mg raz na dobę przez 6 miesięcy. Chromomikoza: 100-200 mg raz na dobę przez 6 miesięcy. Blastomikoza: 100 mg raz na dobę do 200 mg 2 razy na dobę przez 6 miesięcy. Nie stosować u dzieci i osób w podeszłym wieku. **Przeciwwskazania:** Nadwrażliwość na itrakonazol lub inny składnik leku. Leki przeciwwskazane do jednoczesnego podawania z preparatem Trioxal: terfenadyna, astemizol, cisaprid, chinidyna, pimozyd, sertindol, lewacytylometadol, dofetylid, inhibitory CoA-HMG metabolizowane przez CYP3A4, takie jak simwastatyna, lowastatyna, atorwastatyna, triazolam i doustnie podawany midazolam, alkaloidy sporyszu: dihydroergotamina, ergometryna (ergonowina), ergotamina i metyloergometryna (metyloergonowina). Stosowanie preparatu Trioxal jest przeciwwskazane u kobiet w ciąży z wyjątkiem sytuacji zagrożenia życia, gdy potencjalne korzyści przewyższają ewentualny szkodliwy wpływ na płód. Kobiety w wieku rozrodczym powinny stosować odpowiednie środki antykoncepcyjne aż do wystąpienia kolejnej menstruacji po zakończeniu leczenia preparatem. **Ostrzeżenia i zalecane środki ostrożności:** W badaniach na zdrowych ochotnikach z zastosowaniem itrakonazolu podawanego dożylnie obserwowano bezobjawowe zmniejszenie frakcji wyrzutowej lewokomorowej. Itrakonazol wykazywał działanie inotropowe ujemne. Podczas stosowania itrakonazolu występowała zastoinowa niewydolność serca. Nie należy więc stosować preparatu u pacjentów z zastoinową niewydolnością serca występującą aktualnie lub w przeszłości, chyba że potencjalne korzyści przewyższają ryzyko powikłań. Oceniając indywidualnie stosunek korzyści do ryzyka, należy wziąć pod uwagę czynniki, takie jak ciężkość zakażenia, dawkowanie i czas trwania leczenia preparatem Trioxal oraz osobnicze czynniki zagrażające wystąpieniem zastoinowej niewydolności serca. Leczenie tych pacjentów należy prowadzić ostrożnie. Pacjentów zagrożonych rozwojem zastoinowej niewydolności serca należy poinformować o tym, jakie są objawy zastoinowej niewydolności serca, oraz monitorować ich pod względem wystąpienia tych objawów. Jeśli objawy zastoinowej niewydolności serca wystąpią w czasie leczenia itrakonazolem, należy lek odstawić. Należy ostrożnie stosować itrakonazol z antagonistami kanału wapniowego. Itrakonazol może wykazywać klinicznie istotne interakcje z innymi lekami. Wchłanianie itrakonazolu po podaniu doustnym jest zmniejszone w przypadku zmniejszonej kwaśności soku żołądkowego. Leki zobojętniające kwas żołądkowy (np. wodorotlenek glinu) powinny być podawane co najmniej 2 godziny po przyjęciu preparatu Trioxal. U pacjentów z achlorhydrią oraz u pacjentów otrzymujących leki hamujące wydzielanie kwasu żołądkowego (np. antagoniści receptora H2, inhibitory pompy protonowej) zaleca się podawanie preparatu Trioxal z napojem typu „cola". Rzadko występują przypadki hepatotoksyczności, w tym zakończonej zgonem, głównie u pacjentów leczonych itrakonazolem dłużej niż jeden miesiąc. W związku z tym podczas leczenia zaleca się monitorowanie czynności wątroby. Jeśli podczas leczenia u pacjenta wystąpią objawy zapalenia wątroby, takie jak anoreksja, nudności, wymioty, uczucie zmęczenia, bóle brzucha lub ciemny mocz, należy natychmiast zaprzestać stosowania preparatu i ocenić czynność wątroby. U pacjentów ze zwiększoną aktywnością enzymów wątrobowych lub czynną chorobą wątroby, lub u których zaobserwowano działanie hepatotoksyczne innych leków, nie należy rozpoczynać leczenia, chyba że oczekiwane korzyści przewyższają ryzyko uszkodzenia wątroby. W takich przypadkach konieczne jest monitorowanie aktywności enzymów wątrobowych. Itrakonazol jest metabolizowany głównie w wątrobie. U pacjentów z marskością wątroby zaobserwowano niewielkie zmniejszenie biodostępności po podaniu doustnym, które nie było istotne statystycznie. Końcowy okres półtrwania uległ jednak statystycznie istotnemu wydłużeniu. W takich przypadkach konieczne jest dostosowanie dawki. U pacjentów z niewydolnością nerek biodostępność itrakonazolu po podaniu doustnym może być mniejsza. W takim przypadku można rozważyć dostosowanie dawki. W przypadku wystąpienia neuropatii, którą można wiązać z przyjmowaniem itrakonazolu, leczenie należy przerwać. Nie ma dostępnych informacji dotyczących nadwrażliwości krzyżowej pomiędzy itrakonazolem a innymi lekami przeciwgrzybiczymi z grupy azoli. Preparat Trioxal należy przepisywać ostrożnie pacjentom z nadwrażliwością na inne leki z grupy azoli. W kandydozie układowej, jeśli podejrzewa się zakażenie opornymi na flukonazol szczepami Candida, przed rozpoczęciem leczenia należy zbadać wrażliwość tych szczepów na itrakonazol. U niektórych pacjentów z osłabioną odpornością (np. pacjenci z neutropenią, AIDS, po przeszczepach narządowych) biodostępność itrakonazolu może być zmniejszona. Preparat nie jest zalecany w celu rozpoczęcia leczenia bezpośrednio zagrażających życiu grzybic układowych. U pacjentów z AIDS leczonych z powodu grzybic układowych, takich jak sporotrychoza, blastomikoza, histoplazmoza lub kryptokokoza (postać z zajęciem opon mózgowo-rdzeniowych lub bez), u których istnieje ryzyko rozwoju infekcji, należy rozważyć pozostawienie leczenia podtrzymującego. Ze względu na zawartość sacharozy preparatu nie należy stosować u pacjentów z rzadkimi dziedzicznymi zaburzeniami związanymi z nietolerancją fruktozy, zespołem złego wchłaniania glukozy-galaktozy lub niedoborem sacharazy-izomaltazy. **Działania niepożądane:** Podczas leczenia itrakonazolem około 9% pacjentów zgłaszało działania niepożądane. U pacjentów otrzymujących itrakonazol nieprzerwanie przez dłuższy czas (około 1 miesiąca) częstość występowania działań niepożądanych była większa (ok. 15%). Do najczęstszych należały działania pochodzące za strony układu pokarmowego, wątroby i skóry. Ponadto obserwowano inne, które niemniej występowały bardzo rzadko: hipokaliemia, neuropatia obwodowa, bóle głowy, zawroty głowy; zastoinowa niewydolność krążenia; obrzęk płuc, bóle w nadbrzuszu, wymioty, niestrawność, nudności, biegunka i zaparcie, bardzo ciężkie działania hepatotoksyczne z przypadkami ostrej niewydolności wątroby zakończonej zgonem, zapalenie wątroby, przemijające zwiększenie aktywności enzymów wątrobowych, zespół Stevensa-Johnsona, obrzęk naczyniowy, pokrzywka, wypadanie włosów, wysypka i świąd, zaburzenia cyklu miesiączkowego, reakcje alergiczne, obrzęki. **Podmiot odpowiedzialny:** ZF Polpharma S.A. Pozwolenie nadopuszczenie do obrotu nr 10538, wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. Cena urzędowa detaliczna dla opakowania 4 kaps.; 28 kaps. odpowiednio wynosi: 19,60; 102,99 PLN. Kwota dopłaty pacjenta dla opakowania 4 kaps.; 28 kaps. odpowiednio wynosi: 12,60; 53,98 PLN. ChPL: 2007.07.18.

Pirolam (*Ciclopiroxi olaminum*). **Skład i postać:** *Ciclopiroxi olaminum*: 1% żel, 1% zawiesina, 1% roztwór. **Wskazania do stosowania:** Leczenie grzybic skóry gładkiej, owłosionej, grzybic stóp, paznokci, podudzia, spowodowanych przez: *Trichophyton rubrum, T. mentagrophytes, Epidermophyton floccosum, Microsporum canis, M. gypseum, Candida albicans, C. tropicalis, C. krusei, C. parapsilosis, Cryptococcus neoformans, Aspergillus fumigatus*. Zakażenia bakteriami Gram-dodatnimi i Gram-ujemnymi oraz mykoplazmami. **Dawkowanie i sposób podawania:** Do stosowania zewnętrznego: dorośli: 2 razy dziennie na zmienioną chorobowo skórę, lekko wcierając lub pozostawiając do wyschnięcia. Leczenie powinno się prowadzić do ustąpienia objawów chorobowych, optymalny czas leczenia wynosi 3 tygodnie. Zaleca się kontynuowanie leczenia przez okres do 10 dni po ustąpieniu zmian (zapobieganie nawrotom). Nie stosować opatrunków i plastrów; wyjątek stanowi grzybica paznokci. W grzybicy paznokci preparat należy nanieść na zmienione chorobowo miejsca i zastosować opatrunek zamknięty. **Przeciwwskazania:** Nadwrażliwość na cyklopiroks i/lub inne składniki podłoża; grzybicze zapalenie oka i okolic oka. **Ostrzeżenia i zalecane środki ostrożności:** U kobiet ciężarnych można stosować tylko w koniecznych przypadkach, nie należy stosować w czasie karmienia piersią; chronić oczy przed kontaktem z lekiem. Preparat nie jest przeznaczony do leczenia grzybic pochwy. Swędzących zakażonych miejsc nie wolno drapać. **Działania niepożądane:** U nielicznych chorych (1–4%) pieczenie, podrażnienie skóry, rumień, świąd, obrzęk. Brak danych na temat stosowania leku u dzieci. **Podmiot odpowiedzialny:** Medana Pharma SA, 98-200 Sieradz, ul. Władysława Łokietka 10. Pozwolenie na dopuszczenie do obrotu: nr 7401 (1% żel), nr 7400 (1% zawiesina), nr 7399 (1% roztwór), wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. ChPL: 2008.09.29.

Pirolam (*Ciclopiroxum*). **Skład i postać:** 1 g lakieru zawiera 80 mg cyklopiroksu. **Wskazania:** Grzybicze zakażenia paznokci. **Dawkowanie i sposób podawania:** Do stosowania miejscowego. Preparat Pirolam należy nakładać na paznokieć przez pierwszy miesiąc leczenia co drugi dzień, w drugim miesiącu leczenia stosowanie lakieru można ograniczyć do dwóch razy w tygodniu, a od trzeciego miesiąca stosować raz w tygodniu. Cienką warstwę lakieru należy nanieść równomiernie na płytkę paznokciową i pozostawić do wyschnięcia. Raz w tygodniu należy zmyć całą warstwę lakieru kosmetycznym zmywaczem do paznokci. W przypadku uszkodzenia lub starcia warstwy lakieru wystarczy polakierować odsłonięte miejsca. Czas trwania leczenia zależy od ciężkości zakażenia grzybiczego, nie należy jednak stosować go dłużej niż 6 miesięcy. **Przeciwwskazania:** Nadwrażliwość na substancję czynną lub którąkolwiek substancję pomocniczą. Nie stosować u dzieci ze względu na brak wystarczających danych klinicznych. **Ostrzeżenia i zalecane środki ostrożności:** Stosować wyłącznie na paznokcie. Produkt łatwopalny, używać z dala od źródeł ciepła i otwartego ognia. Unikać kontaktu z oczami i błonami śluzowymi. Podczas leczenia nie należy stosować na płytkę paznokciową produktów kosmetycznych. **Działania niepożądane:** U nielicznych chorych obserwowano miejscowe podrażnienie (pieczenie, świąd, rumień, obrzęk, zaczerwienienie i łuszczenie) po kontakcie preparatu ze skórą otaczającą paznokieć. **Podmiot odpowiedzialny:** Medana Pharma SA, 98-200 Sieradz, ul. Władysława Łokietka 10. Pozwolenie na dopuszczenie do obrotu nr 15279, wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. ChPL: 2009.03.31.

Telfexo 120 mg; Telfexo 180 mg *(Fexofenadini hydrochloridum)*. **Skład i postać:** Jedna tabletka powlekana zawiera 120 mg lub 180 mg feksofenadyny chlorowodorku. **Wskazania:** Dla dawki 120 mg – łagodzenie objawów sezonowego alergicznego zapalenia błony śluzowej nosa; 180 mg – łagodzenie objawów przewlekłej pokrzywki idiopatycznej. **Dawkowanie i sposób podawania:** Dorośli i dzieci w wieku 12 lat i starsze – 1 tabletka raz na dobę; lek nie jest zalecany do stosowania u dzieci poniżej 12 lat w związku z brakiem danych dotyczących bezpieczeństwa i skuteczności; u pacjentów w podeszłym wieku, pacjentów z zaburzeniami czynności wątroby i nerek lek należy stosować ostrożnie. Tabletkę należy połknąć, popijając wystarczającą ilością wody. **Przeciwwskazania:** Nadwrażliwość na substancję czynną lub którąkolwiek substancję pomocniczą. **Ostrzeżenia i zalecane środki ostrożności:** Pacjentów z chorobami układu krążenia w wywiadzie należy ostrzec, że leki przeciwhistaminowe z tej grupy mogą mieć związek z występowaniem działań niepożądanych, takich jak tachykardia i kołatanie serca. Dostępne są jedynie ograniczone dane dotyczące stosowania u pacjentów w podeszłym wieku i u pacjentów z zaburzeniami czynności nerek lub wątroby. W tych grupach pacjentów chlorowodorek feksofenadyny powinien być stosowany ostrożnie. **Działania niepożądane:** W kontrolowanych badaniach klinicznych częstość występowania częstych działań niepożądanych była podobna do obserwowanej w grupie placebo. Zaburzenia układu nerwowego: często – bóle głowy, senność, zawroty głowy. Zaburzenia żołądka i jelit: często – nudności, suchość w jamie ustnej. Zaburzenia ogólne i stany w miejscu podania: niezbyt często – zmęczenie. Następujące działania niepożądane były zgłaszane u osób dorosłych w ramach nadzoru po wprowadzeniu produktu do obrotu. Częstość ich występowania nie jest znana (nie może być określona na podstawie dostępnych danych): Zaburzenia układu immunologicznego: reakcje nadwrażliwości z objawami, takimi jak: obrzęk naczynioruchowy, uczucie ucisku w klatce piersiowej, duszność, nagłe zaczerwienienie skóry i uogólnione objawy anafilaksji. Zaburzenia psychiczne: bezsenność, nerwowość, zaburzenia snu lub koszmary senne/nieprawidłowe sny. Zaburzenia serca: tachykardia, palpitacje. Zaburzenia żołądka i jelit: biegunka. Zaburzenia skóry i tkanki podskórnej: wysypka, pokrzywka, świąd. **Podmiot odpowiedzialny:** ZF Polpharma S.A. Pozwolenie na dopuszczenie do obrotu: Telfexo 120 mg i 180 mg odpowiednio nr 14235 i 14236, wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. ChPL: 2011.03.30.

Heviran (*Aciclovirum*). **Skład i postać:** Jedna tabletka powlekana zawiera odpowiednio 200 mg, 400 mg lub 800 mg acyklowiru oraz substancje pomocnicze. **Wskazania:** Leczenie zakażeń skóry i błon śluzowych wywołanych przez wirus opryszczki pospolitej (*Herpes simplex virus*), w tym pierwotnego i nawracającego opryszczkowego zakażenia narządów płciowych. Zapobieganie nawrotom opryszczki pospolitej (*Herpes simplex virus*) u pacjentów z prawidłową odpornością. Leczenie zakażeń wywołanych przez wirus ospy wietrznej i półpaśca (*Varicella-Zoster virus*). **Dawkowanie i sposób podawania:** Dawkowanie u dorosłych: Leczenie zakażeń wywołanych przez wirus opryszczki pospolitej (*Herpes simplex virus*): Produkt należy podawać w dawce 200 mg pięć razy na dobę, mniej więcej co 4 godziny, z przerwą nocną. Produkt stosować przez 5 dni, a w przypadku ciężkich zakażeń pierwotnych kurację można przedłużyć. Pacjentom ze znacznie zmniejszoną odpornością (np. po przeszczepieniu szpiku) lub z zaburzeniami wchłaniania z przewodu pokarmowego dawkę można podwoić do 400 mg. Podawanie acyklowiru należy rozpocząć jak najwcześniej po rozpoznaniu zakażenia. W zakażeniach nawracających szczególnie ważne jest rozpoczęcie leczenia w okresie prodromalnym lub tuż po pojawieniu się pierwszych zmian. Zapobieganie nawrotom opryszczki pospolitej (*Herpes simplex virus*): U pacjentów z prawidłową odpornością produkt należy podawać w dawce 200 mg trzy – cztery razy na dobę, mniej więcej co 6-8 godzin. U wielu pacjentów skuteczne oraz wygodniejsze może być podawanie produktu dwa razy na dobę, co 12 godzin, w dawce 400 mg. Zapobiegawczo może również działać dawka zmniejszana stopniowo do 200 mg trzy razy na dobę, mniej więcej co 8 godzin, lub nawet dwa razy na dobę, mniej więcej co 12 godzin. U niektórych pacjentów reakcja na zapobiegawcze podawanie leku występuje po zastosowaniu całkowitej dawki dobowej produktu wynoszącej 800 mg. Stosowanie leku należy przerywać co 6 do 12 miesięcy w celu obserwacji ewentualnych zmian w przebiegu choroby. Leczenie zakażeń wywołanych przez wirus ospy wietrznej i półpaśca (VZV): Produkt należy podawać w dawce 800 mg cztery – pięć razy na dobę (mniej więcej co 4 godziny), z przerwą nocną. Leczenie należy kontynuować przez 5 do 7 dni. Należy rozważyć dożylne podawanie leku pacjentom ze znacznie zmniejszoną odpornością (np. po przeszczepieniu szpiku) lub z zaburzeniami wchłaniania z przewodu pokarmowego. Leczenie trzeba rozpocząć bezzwłocznie po rozpoznaniu zakażenia. Najlepsze wyniki lecznicze zarówno w przypadku ospy wietrznej, jak i półpaśca uzyskuje się, podając lek w ciągu 24 godzin po wystąpieniu pierwszych zmian skórnych – wysypki. Dawkowanie u dzieci: Leczenie zakażeń wywołanych przez wirus opryszczki pospolitej (*Herpes simplex virus*): Dzieci w wieku 2 lat i powyżej – dawka jak dla dorosłych. Leczenie zakażeń wirusem ospy wietrznej: Dzieci w wieku od 6 lat – 800 mg cztery razy na dobę; dzieci w wieku 2 do 5 lat – 400 mg cztery razy na dobę. Dawkę można też określić dokładniej, podając 20 mg/kg masy ciała (do maksymalnej dawki 800 mg) cztery razy na dobę. Leczenie należy kontynuować przez 5 dni. Brak szczegółowych danych na temat zapobiegania nawrotom zakażeń wirusem opryszczki pospolitej oraz leczenia półpaśca u dzieci z prawidłową odpornością. Dawkowanie u pacjentów w podeszłym wieku: U pacjentów w podeszłym wieku należy brać pod uwagę możliwość zaburzenia czynności nerek i odpowiednio dostosować dawkę leku (patrz: Dawkowanie u pacjentów z zaburzeniami czynności nerek). Należy utrzymywać stan właściwego nawodnienia organizmu. Dawkowanie u pacjentów z zaburzeniami czynności nerek: Zaleca się ostrożność podczas stosowania acyklowiru u pacjentów z zaburzeniami czynności nerek. Należy utrzymywać odpowiedni poziom nawodnienia organizmu u tych pacjentów. Podczas leczenia zakażeń wirusem opryszczki pospolitej lub zapobiegania im u pacjentów z umiarkowanym i ciężkim zaburzeniem czynności nerek stosowanie zalecanych dawek doustnych nie doprowadzi do kumulacji acyklowiru w organizmie w stężeniach wyższych niż uznane za bezpieczne podczas podawania leku dożylnie. Jednak pacjentom z ciężką niewydolnością nerek (klirens kreatyniny mniejszy niż 10 ml/min) zaleca się zmniejszenie dawki do 200 mg dwa razy na dobę, mniej więcej co 12 godzin. Podczas leczenia zakażeń wirusem ospy wietrznej i półpaśca u pacjentów z umiarkowaną niewydolnością nerek (klirens kreatyniny 10-25 ml/min) zaleca się zmniejszenie dawki do 800 mg trzy razy na dobę, mniej więcej co 8 godzin, a u pacjentów z ciężką niewydolnością nerek (klirens kreatyniny mniejszy niż 10 ml/min) zaleca się zmniejszenie dawki do 800 mg dwa razy na dobę, mniej więcej co 12 godzin. **Przeciwwskazania:** Nadwrażliwość na acyklowir, walacyklowir lub na jakikolwiek składnik leku. **Ostrzeżenia i zalecane środki ostrożności:** Acyklowir jest wydalany z organizmu przez nerki, toteż dawkę leku u osób z niewydolnością nerek należy zmniejszyć. U pacjentów w podeszłym wieku należy brać pod uwagę zaburzenia czynności nerek, toteż w tej grupie pacjentów należy rozważyć zmniejszenie dawki leku. Osoby w podeszłym wieku oraz pacjenci z niewydolnością nerek należą do grupy zwiększonego ryzyka działań niepożądanych dotyczących układu nerwowego i z tego względu obie te grupy należy uważnie obserwować. Zgłaszane dotąd objawy zwykle ustępowały po przerwaniu podawania leku. Stan nawodnienia: należy zachować szczególną ostrożność podczas stosowania doustnie dużych dawek acyklowiru, aby utrzymać właściwy stan nawodnienia organizmu, np. w przypadku leczenia półpaśca (4 g na dobę), aby uniknąć ryzyka uszkodzenia nerek. Obecnie dostępne dane z badań klinicznych nie są wystarczające, aby wnioskować, że leczenie acyklowirem zmniejsza możliwość wystąpienia powikłań związanych z półpaścem u pacjentów z prawidłową odpornością. Długotrwałe podawanie acyklowiru lub powtarzane cykle leczenia u pacjentów ze znacznie osłabioną odpornością mogą spowodować selekcję szczepów wirusa o zmniejszonej wrażliwości na acyklowir. **Ciąża i laktacja:** Podawanie acyklowiru kobietom w ciąży należy rozważać jedynie wtedy, gdy oczekiwane korzyści ze stosowania leku u matki przewyższają możliwość wystąpienia zagrożeń u płodu. Karmienie piersią: Po doustnym podaniu acyklowiru w dawce 200 mg pięć razy na dobę stężenie jego w mleku kobiecym wynosiło od 60% do 410% stężenia leku mierzonego w osoczu. Takie stężenie acyklowiru w mleku matki mogłoby spowodować przyjmowanie leku przez oseska w dawce dobowej do 0,3 mg/kg masy ciała. Dlatego należy zachować ostrożność, podając lek kobietom karmiącym piersią. **Działania niepożądane:** Następujące zasady zostały przyjęte dla sporządzenia klasyfikacji częstości występowania: bardzo często (≥1/10), często (≥1/100 do <1/10), niezbyt często (≥1/1 000 do <1/100), rzadko (≥1/10 000 do <1/1 000), bardzo rzadko (<1/10 000). Zaburzenia krwi i układu chłonnego: *Bardzo rzadko*: niedokrwistość, zmniejszenie liczby krwinek białych, małopłytkowość. Zaburzenia układu immunologicznego: *Rzadko*: reakcje anafilaktyczne. Zaburzenia psychiczne i zaburzenia układu nerwowego: *Często*: bóle głowy, zawroty głowy; *bardzo rzadko*: pobudzenie, dezorientacja, drżenia, niezborność ruchowa, zaburzenia wymowy, omamy, objawy psychotyczne, drgawki, senność, encefalopatia, śpiączka. Powyższe objawy są zwykle przemijające i występowały głównie u pacjentów z zaburzeniami czynności nerek lub z innymi czynnikami predysponującymi. Zaburzenia oddechowe, klatki piersiowej i śródpiersia: *Rzadko*: duszność. Zaburzenia żołądka i jelit: *Często*: nudności, wymioty, biegunka, bóle brzucha. Zaburzenia wątroby i dróg żółciowych: *Rzadko*: przemijające zwiększanie stężenia bilirubiny oraz aktywności enzymów wątrobowych we krwi; *bardzo rzadko*: zapalenie wątroby, żółtaczka. Zaburzenia skóry i tkanki podskórnej: *Często*: świąd, wysypki (w tym z nadwrażliwością na światło); *niezbyt często*: pokrzywka, przyspieszone, rozsiane wypadanie włosów. Ponieważ przyspieszone, rozsiane wypadanie włosów może być związane z wieloma chorobami oraz ze stosowaniem wielu leków, jego związek z działaniem acyklowiru nie jest pewny; *rzadko*: obrzęk naczynioruchowy. Zaburzenia nerek i dróg moczowych: *Rzadko*: zwiększenie stężenia mocznika i kreatyniny we krwi; *bardzo rzadko*: ostra niewydolność nerek, ból nerek. Ból nerek może być związany z niewydolnością nerki (nerek). Należy utrzymywać właściwy stan nawodnienia chorego. Zaburzenie czynności nerek zwykle szybko ustępuje po uzupełnieniu płynów u pacjenta i (lub) po zmniejszeniu dawki lub odstawieniu leku. W wyjątkowych przypadkach może jednak dojść do ostrej niewydolności nerek. Zaburzenia ogólne i stany w miejscu podania: *Często*: zmęczenie, gorączka. **Podmiot odpowiedzialny:** ZF Polpharma S.A. Dopuszczenie do obrotu dla dawek 200, 400 i 800 mg odpowiednio nr 8400, 8401, 8402, wydane przez MZ. Dodatkowych informacji o leku udziela: Polpharma Biuro Handlowe Sp. z o.o., ul. Bobrowiecka 6, 00-728 Warszawa, tel. +48 22 364 61 00, fax +48 22 364 61 02, www.polpharma.pl. Lek wydawany na podstawie recepty. Cena urzędowa detaliczna dla dawek: 200 mg x 30 tabl.; 400 mg x 30 tabl.; 800 mg x 30 tabl. odpowiednio wynosi w PLN: 16,39; 28,49; 49,83. Kwota dopłaty pacjenta odpowiednio wynosi w PLN: 10,16; 16,03; 24,92. ChPL: 2012.02.22.

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CONTENTS

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EFFECT OF ANTIRETROVIRAL THERAPY ON SURVIVAL OF HIV/TB-INFECTED PATIENTS IN UKRAINE

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Abstract
Introduction: There is little information from Ukraine about the effect of highly active antiretroviral therapy (HAART) on survival of HIV/TB-infected patients. We evaluated the survival and the effect of HAART on mortality on these patients.
Methods: Prospective cohort study of HIV patients who developed TB from January 2005 to December 2006 in a Zaporizhzhya AIDS Center, and were tracked for 60 months after start HAART. Survival was determined by Kaplan-Meier method and effect of HAART on survival was evaluated using Cox proportional hazards models.
Results: Eighty patients were studied (mean age 34 years, 50% male, median CD4 count 103 cell/μL). In 60 months of HAART 14 patients died. The probability of survival was 82%. In multivariate analysis, patients with a CD4 cell count <100 μL had a 5-fold higher risk of mortality (HR 5, 2; 95% CI 1.4-19, 4) and those with extra pulmonary tuberculosis a 2, 2-fold increased risk (HR 2, 2, 95% CI 1, 1-8, 3).
Conclusions: HAART significantly increased probability of survival and reduced the risk of death for HIV/TB-infected patients in Ukraine.

Key words: TB; HIV; HAART; survival; Ukraine

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Introduction

The global HIV pandemic has a dramatic impact on the epidemiology of tuberculosis (TB). It has been estimated that global prevalence of active TB was greater than one third of the estimated 36 million patients infected with HIV. The risk of TB is dramatically increased in HIV-infected patients as a result of a higher probability of either primary progression or reactivation of latent infection [1-3]. The HIV and TB epidemics overlap to a great degree in Eastern Europe countries, including Ukraine. In Ukraine tuberculosis is the most frequent major pportunistic infection (OI) and the leading cause of mortality among HIV-infected patients. In developed countries, prior to the introduction of highly active antiretroviral therapy (HAART), a wide range of survival times in people with TB-HIV was reported [4-7]. Some of these reports showed associations between immunosuppression, history of AIDS and TB location with risk of death [5,6]. In the HAART era morbidity and mortality of people living with HIV and AIDS has been reduced significantly, in both industrialized and less developed regions [8,9]. Further, in settings of widespread use, HAART appears to have been responsible for a significant reduction in the incidence of TB, even in places with high prevalence of this disease [10,11]. There is also increasing evidence about the efficacy or effectiveness of HAART when used together with anti-TB therapy [12].

To date, there have been limited clinical data regarding survival rates among HIV/TB-infected patients and the impact of HAART on clinical outcomes in Ukraine. We therefore, conducted the present study to determine the survival rate among HIV/TB-infected patients who received HAART. This study also aimed to determine possible risk factors that related to death among these patients and the appropriate timing for initiating HAART after TB diagnosis.

Material and Methods

A prospective cohort study was conducted among HIV-infected patients who were diagnosed with active TB between January 2005 and December 2006.

In study we included HIV-infected patients older than 18 years. TB diagnosis was confirmed by direct (positive Ziehl-Nielsen in sputum smear) or pathological (typical granuloma in biopsy) examination.

Pulmonary TB was the one with positive sputum or positive culture of sputum or bronchial aspirate, without clinical or radiological evidence of extra-pulmonary extension.

Extra-pulmonary TB was considered when there was clinical evidence and positive Ziehl-Nielsen in biopsy from at least one extra-pulmonary location, with or without pulmonary TB.

In general anti-TB therapy was standard according to the World Health Organization (WHO). All patients included in the study received antituberculosis medications as directly observed therapy and in accordance with standard Category 1 regimens.

Prophylactic regimens against opportunistic infections were provided in accordance with national Ukrainian guidelines during all time of the study.

The HAART regimens consisted of three-drug therapy using combinations listed by the WHO as approved first-line regimens.

Statistical analysis

Median (interquartile range, IQR), and frequencies (%) were used to describe patients' characteristics in each group. The Kaplan-Meier test was used to estimate the probability of death and the median time to death. To compare survival by baseline immunological status, the Kaplan-Meier analysis was further stratified by baseline CD4 count (<100, 100–200, and >200 cells/μL). The hazard ratio (HR) and its 95% confidence interval (CI) derived from univariate and multivariate Cox proportional hazards models. All analyses were performed using STATISTICA 6.0 and SPSS 17.0. A P value less than 0.05 was considered statistically significant.

Results

A total of 80 patients who met the inclusion criteria were identified. Time of follow-up was 60 months. Mean age at initial presentation was 34 years (range 18-54 years). Forty patients (50%) were female. The median (IQR) CD4 cell count was 103 μL (63-182). Thirty-seven patients (46%) had an initial CD4 cell count <100/ μL, 22 (28%) a count of 100 - 200/ μL, 21(26 %.) a count of >200/ μL (Tabl. I).

Injection drug use was the most common route (65%) of HIV transmission. The number of patients co-infected with the hepatitis C virus (HCV) was 57 (71%). Specific HAART regimens included zidovudine-lamivudine-efavirenz, administered to 70 (88%) of the patients; stavudine-lamivudine-efavirenz, administered to 10 (12%). The baseline demographic and clinical characteristics are provided in Table I.

The probabilities of survival after start HAART estimated by the Kaplan-Meier method is shown in Figure 1. Survival rates at 1, 2, 3, 4 and 5 years were 95%, 92%, 86%, 86%, and 82%. Among 14 patients who died, 12 patients (86%) died during the first 36 months of HAART. Five of the 14 patients who died, had CD4 counts less than 50 cells/μL. The causes of death were TB (12 cases), primary CNS lymphoma and cryptococal meningitis (one case for each). Among the 5 patients with HIV-related deaths that occurred during the first 12 months of HAART, we identified 4 in whom death could be attributed to IRIS (all case of tuberculosis meningitis).

Univariate analysis revealed that a baseline CD4 cell count <100/ μL was a significant predictor of mortality (HR 5,6;95% CI 1,6-20,1,p=0.02) (Tabl. II). Patients who died had a mean pre-ART CD4 cell count of 77 (43-95) μL compared with 130 (65-213) μL for those who were alive at the conclusion of the study. Also, in univariate analysis, patients with extrapulmonary tuberculosis had an increased mortality risk (HR 2, 4; 95% CI 1, 2-10, 4, p=0.032).

Figure 2 shows a Kaplan-Meier curve depicting the probability of death over the 5-year period of observation according to CD4 cell count stratum. The log-rank test indicates (P<0, 01) that the probability of death was statistically significantly different among the different CD4 cell count strata, with the lowest survival (70%) among patients who initiated ART with a CD4 cell count <100 cells/uL.

In multivariate analysis, patients with a CD4 cell count <100 μL had a 5-fold higher risk of mortality (HR 5,2;95% CI 1.4-19,4, p<0.05) and those with extrapulmonary tuberculosis a 2,2-fold increased risk (HR 2,2, 95% CI 1,1-8,3, p<0,05) (Tabl. II). Ages, gender, route of HIV transmission were not predictive of mortality.

Mean age (years)	34 (18-54)
Male (%)	50
Route of HIV transmissionHeterosexual IDU	28 (35%) 52 (65%)
Pulmonary TB location (%)	35
CD4 T-lymphocyte count (cells/μL) Median (IQR)	103 (63-182)
<100	37 (46%)
100-200	22 (28%)
>200	21 (26%)

Table I. Clinical characteristics of HIV/TB-patients

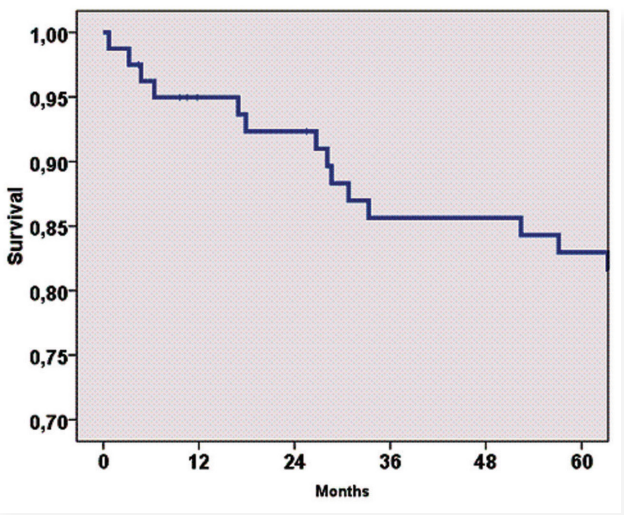

Figure 1. Survival time of HIV/TB- patients by antiretroviral therapy

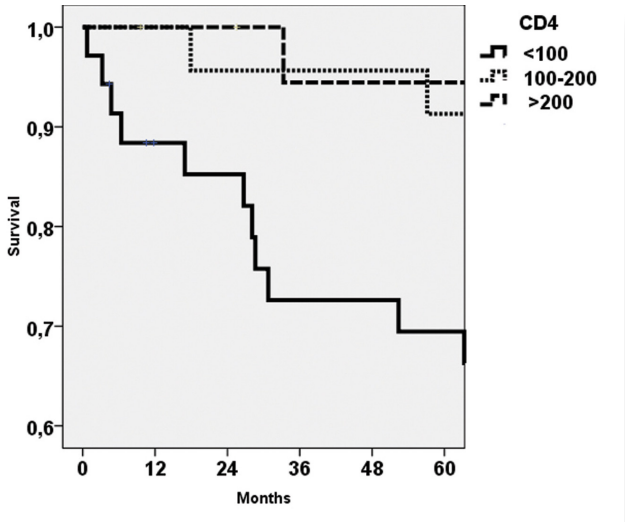

Figure 2. Survival time of HIV/TB- patients by CD4 T-lymphocyte count

Variable	Category	Unadjusted HR (95% CI)	Adjusted HR (95% CI)
Age	>34	1,8 (0,6-5,2)	1,4 (0,4-4,4)
	<34	1	1
Gender	Female	0,5 (0,16-1,6)	0,4 (0,13-1,4)
	Male	1	1
TB location	Extrapulmonary	2,4 (1,2-10,4)	2,2 (1,1-8,3)
	Pulmonary	1	1
Route of HIV transmission	IDU	1,2 (0,3-3,2)	1,1 (0,4-3,1)
	non IDU	1	1
CD4 count	< 100 cells/uL	5,6 (1,6-20,1)	5,2 (1,4-19,4)
	> 100 cells/uL	1	1

Table II. Cox proportional hazards for baseline predictors of HIV/TB- patients

HR - Cox proportional hazards ratio: 95% CI = 95% confidence interval.

Unadjusted - Cox proportional hazards ratio comparing group 1 v. group 2 in a univariate model with only one risk factor included.

Adjusted - Cox proportional hazards ratio estimated in a multivariate model including age. Gender, TB location. Route of HIV transmission, CD4 cell count

Discussions

To date, combined ART has been widely used for the treatment for HIV/TB-infected patients in the world. A plenty of studies precisely demonstrate the impact of ART on the survival outcomes among HIV-infected patients with successful immune restoration and reductions in morbidity and mortality [13-15]. However, the data regarding survival rates among HIV/TB-coinfected patients and the impact of HAART on clinical outcomes in Ukraine are still limited.

For HIV/TB-infected patients who did not receive ART, approximately half of them died within 1 year after TB diagnosis. As known, the simultaneous use of HAART with anti-TB therapy in patients with TB-HIV significantly reduces the risk of death in the short and long term compared with the risk of death for those receiving only anti-TB therapy [16-17].

The results from the present study point out that HAART is crucial to improve survival in HIV/TB-infected patients.

We observed that degree of immunosuppression (CD4+ <100 cells/μL) were associated with increased risk of dying and exstapulmonary TB as a risk factor of death. Whalen et al. [8] have shown that survival time was shorter in HIV-infected patients with extrapulmonary TB. This finding may be explained by the fact that patients with extrapulmonary TB have a higher bacterial load of M. tuberculosis and much more severe immunodeficiency status.

Most patients in the present study died of TB-related conditions. The previous studies have demonstrated that death within the first few months of TB treatment may be related to TB, whereas late deaths are attributable to HIV disease progression.

Initiation of HAART and immunological restitution may „unmask» extrapulmonary sites of TB explains the high mortality in first 6 months of HAART.

We observed 4 deaths that could be attributed to IRIS, of which 3 were attributable to central nervous system syndromes. The IRIS deaths primarily involved the central nervous system, suggesting that central nervous system manifestations may not be as well tolerated as manifestations in other body compartments and may require more-urgent attention.

In conclusion, HAART significantly reduced the risk of death for people with TB-HIV in Ukraine. These findings add further evidence to emphasize the importance of access to HAART, especially in regions where HIV infection and TB disease are highly prevalent.

Contributors

All authors contributed to conceptualization, design, data collection, and revision of the final draft of the study, which was written by M. Andreichyn and D. Zhyvytsia.

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CLASSICAL FUSIFORM EXCISION OF MELANOCYTIC NEVI: OUR EXPERIENCE

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Abstract
Introduction: Facial melanocytic nevi whether acquired or congenital may pose an aesthetic problem for many patients, especially women. There are many methods of removal of melanocytic nevi including surgical and non-surgical. However, surgical excision by classical fusiform excision remains the most widely used and one of the best methods taking all perspectives into consideration.
Aim: To evaluate efficacy of classical fusiform excision for the removal of facial melanocytic nevi.
Methods: In a prospective study, 55 facial melanocytic nevi were removed by fusiform excision technique. Incision was given around the nevus in an elliptical pattern, three times as long as it was wide and along the relaxed skin tension lines. Dissection was carried down to mid-subcutaneous tissue to remove the nevus down to its full depth. Wound was closed by simple interrupted sutures using 5-0 prolene with adequate undermining of the wound edges. Sutures were removed on 7th postoperative day.
Results: Complete removal of nevi was achieved in all patients with good to excellent cosmetic results. The scar mark if any, would fade in 3-4 months and were imperceptible in 6-9 months.
Conclusions: Fusiform excision is one of the best, most widely used and time-tested procedure for complete surgical excision of melanocytic nevi.

Key words: melanocytic naevi; naevi; fusiform excision

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Iffat Hassan, Shazia Jeelani, Abid Keen, Mashkoor Wani: Classical fusiform excision of melanocytic nevi: our experience. Our Dermatol Online. 2013; 4(2): 153-156.

Introduction

Melanocytic naevi are common lesions that can be found on the integument of almost all individuals. Some patients present with few lesions, while others have hundreds. The number on a given individual increases in rough proportion to the degree of skin pigmentation.

Facial melanocytic naevi whether acquired or congenital may pose an aesthetic problem for many patients especially women. There are many methods of removal of melanocytic naevi including surgical and non-surgical. However, surgical excision by classical fusiform excision remains the most widely used and one of the best methods taking into consideration all perspectives. We have found good cosmetic result and complete removal of naevus with no recurrence while removing the facial naevus by means of fusiform excision and here by report our findings.

Material and Methods

A prospective study was carried out in the Department of Dermatology, STD & Leprosy (Associated teaching hospital of Govt. Medical College, Srinagar) from January 2010 to December 2011 involving 50 patients (45 female and 5 male) in the age range of 18-51 years having a total of 55 melanocytic naevi on their face which were excised. In two female patients, multiple facial melanocytic naevi were excised and in all others single naevus was excised. The size of the naevi varied from 1 to 1.5 cm diameter.

Routine laboratory investigations including complete hemogram, fasting blood sugar, bleeding and clotting time were done in all patients. Xylocaine sensitivity was performed in all patients. Close-up photographs were taken and an informed consent was obtained. The area was cleaned with povidine iodine and spirit. Under local anaesthesia using 2% lignocaine, incision was given around the naevus in an elliptical pattern using scalpel blade.

The length of the incision being approximately 3 times its width. Care was taken to make the incision always along the relaxed skin tension lines.

The precise inclination of these lines was confirmed by having the patient engaged in a variety of exaggerated facial expressions and by pinching the skin in between thumb and forefinger. A total of 10 naevi were on the natural facial folds including nasolabial folds, on forehead along the wrinkles in a middle aged woman. These lesions healed with excellent cosmetic results obtained as early as 15 days after the excision.

Dissection was carried down to mid-subcutaneous tissue to remove the naevus down to its full depth. In case of larger naevi on relatively less mobile skin subcutaneous sutures using absorbable Vicryl were given so as to approximate the wound margins. Wound was closed by simple interrupted sutures using non-absorbable polypropylene monofilament suture (5-0 Prolene) and with adequate undermining of the wound edges. Wound was dressed with sterile gauze packs. Systemic antibiotics were prescribed for 1 week post-operation to minimize the chances of post-operative infection. The specimen obtained was sent for histopathological examination in all cases. Sutures were removed on 7th postoperative day. After the suture removal, scar was immediately treated with topical silicone gel to fade out the suture marks.

Results

Complete removal of naevi was achieved in all patients with good to excellent cosmetic results (Fig. 1a-d, 2a-c, 3a-c, 4a,b). The complications noted during the procedure were minor and included hematoma formation and slight wound infection in 3 patients each and stitch granuloma in one patient. The scar mark, if any faded in 2-3 months and was imperceptible in 6-7 months. No recurrences were noted. Patients were very much satisfied with the result outcome to the extent that patients with multiple facial melanocytic naevi requested for the removal of other facial naevi.

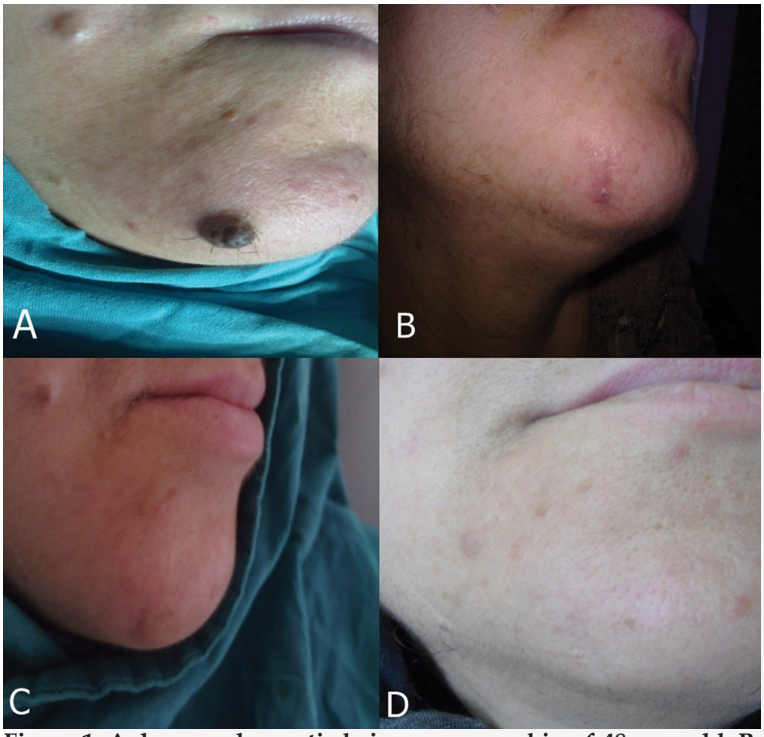

Figure 1. A. large melanocytic hairy nevus on chin of 48 year old; B. weeks post-excision; C. 4 weeks post excision; D. excellent cosmetic result after 12 weeks

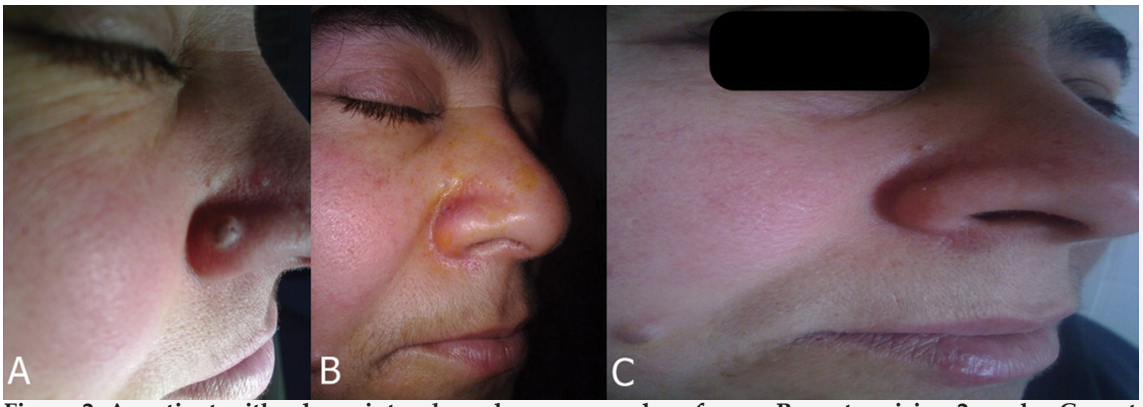

Figure 2. A. patient with a large intra-dermal neavus on alae of nose; B. post excision 2 weeks; C. post excision 4 weeks

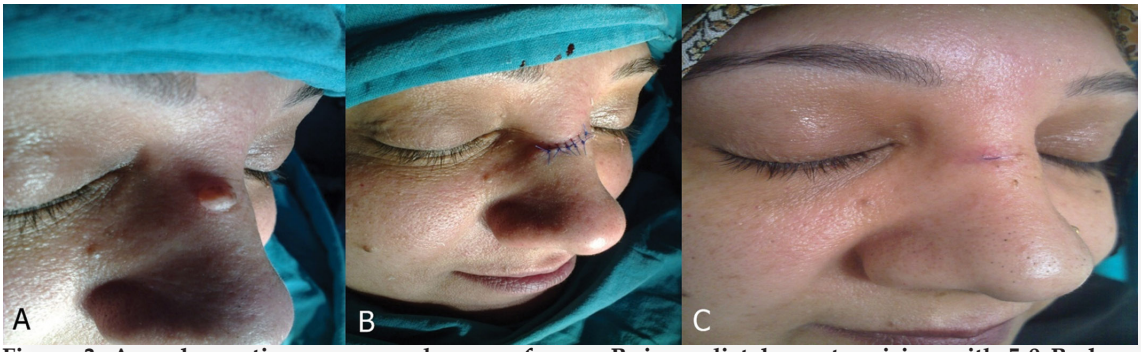

Figure 3. A. melanocytic naevus on dorsum of nose; B. immediately post-excision with 5-0 Prolene sutures; C. same lesion after 2 weeks, subcutaneous Vicryl coming out from the wound which was cut off, with good cosmesis

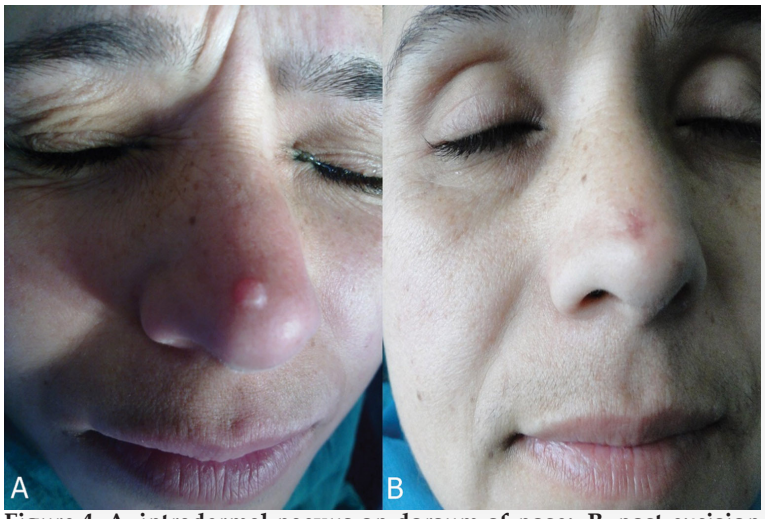

Figure 4. A. intradermal naevus on dorsum of nose; B. post-excision 2 weeks

Discussions

Along with recent advances in the knowledge of pigment formation and of pigment cell biology, there has been an increasing interest in the pigmented mole or naevus. Melanocytic naevi are normal benign proliferations of melanocytes. Although the risk of a naevus evolving into a melanoma is extremely small, melanocytic naevi are both risk factors for melanoma and precursors of melanoma. The prevalence of pigmented lesions present at birth varies considerably between published series; this is principally because of the ethnic mix of the patients examined, as congenital melanocytic naevi may be more common in black or Asian children [1], but they are generally considered to be present in between 1 and 2% of newborns [2]. Some patients present with few lesions, while others have hundreds. Melanocytic naevi develop through childhood and twin studies provide good evidence that naevus number is predominantly genetically determined [3] with a smaller effect of sun exposure [4].

Melanocytic naevi can be broadly divided into congenital and acquired types. Congenital melanocytic naevi vary considerably in size and are classified according to American National Institutes of Health (NIH) consensus definition [1] as small (< 1.5cm), intermediate (1.5-20 cm), or large/giant (>20cm). Conventional or common acquired melanocytic naevi are generally less than 1cm in diameter and evenly pigmented.

Not all melanocytic naevi that change are malignant, especially if change is noted in a person younger than 40 years. However, change that is perceptible over a short time is an indicator of potential malignancy and designates a lesion deserving a biopsy. An Australian study found that 16% of benign lesions changed (as measured by sequential digital dermoscopic imaging) over an interval of 2.5-4.5 months. The proportion of benign lesions that changed was higher in persons aged 0-35 years than in those aged 36-65 years but rose again in the elderly (age >65 years) [5].

Since it is an extremely common lesion, clinically often disfiguring, many patients are seen who desire cosmetic removal of their moles. Removal of a medium size melanocytic naevus whether congenital or acquired over exposed parts, especially over face is warranted for its cosmetic, embarrassment rather than for its potential to cause malignancy.

Several methods of dealing with the common mole are described in the literature. These may be divided into two main types: deep excision, which removes the entire lesion, and other methods, which do not completely remove it. Out of the many methods available (viz, surgical resection, shave excision, laser removal) cosmetic result of surgical resection with primary suturing is always preferable [6]. This is technically less demanding and can be performed even by a novice cutaneous surgeon if basic principles of cosmetic surgery are taken care of.

Melanocytic naevi removed for cosmesis are often removed by tangential or shave excision however such a procedure has its potential disadvantages as there are chances of incomplete removal and subsequent recurrence of the naevus. Moreover, shave excisions can sometimes heal with a permanent scar formation.

Punch excision can be used for relatively small dome-shaped lesions [7]. However when used for larger lesions, they may lead to the formation of dog ears.

Melanocytic naevi can also be removed by means of CO2 laser. However there is a high incidence of repigmentation following CO2 laser treatment of melanocytic naevi [8]. Cost implications of laser treatments should also be considered when choosing treatment options for the removal of melanocytic naevi. Finally, laser-induced malignant transformation of naevus cells remains a theoretical concern but such concerns have not been found to be true in clinical practice [9].

Large lesions may require complete excision with sutured closure, even if known to be benign, because lesions exceeding 1 cm in diameter often are not amenable to the shave technique.

A simple conservative excisional biopsy with a sutured closure is usually the most expeditious means to diagnosis if concern exists regarding the possibility of melanoma. If the lesion is found to be benign, then, ordinarily, no further treatment is required. Providing the pathologist with a complete excisional specimen affords him or her best opportunity to make an accurate diagnosis because all available criteria (including low-magnification attributes such as size, circumscription and symmetry) can be applied to the lesion.

The elliptical or fusiform, excision is the classic approach to removing an approximately round or linear lesion [10,11]. An excision of this shape is preferred because of the subsequent ease of its closure. A round excision larger than a 4-mm punch is difficult to close without leaving excess outpouchings of skin or dog ears.

The ellipse should be long enough so it can be sutured together without dog ears but no longer than necessary to minimize the length of the inevitable scar. The process begins by marking the lesion to be removed. A permanent marker or other skin marker may be used to circle the target lesion. Often, a free margin of several millimetres must be removed from around the primary lesion, and the mark for this margin can be drawn as a larger concentric circle. Then, an ellipse should be marked around the larger circle. The diameter of the circle should form the short axis of the ellipse, and the longer axis should be 3-4 times this length.

The orientation of the ellipse is important. Ideally, the long axis should lie parallel to the relaxed skin tension lines at the body site involved. Diagrams that display the common skin tension lines on the face and body are available. The precise inclination of these lines can be confirmed by having the patient engage in a variety of exaggerated facial expressions. On the arms and legs, flexion and extension can substitute for such manoeuvres. The surgeon can obtain this information at the surgical site by deliberately wrinkling the skin at the surgical site between his or her thumb and forefinger. By placing the ellipse along relaxed skin tension lines, the surgeon ensures that the final scar is parallel to these lines. If the scar is thin and small and if the patient is elderly, it may be barely noticeable, appearing as merely another wrinkle or crease to the untrained observer.

The most commonly used and versatile suture in cutaneous surgery is the simple interrupted suture [12]. Compared with running sutures, interrupted sutures are easy to place, have greater tensile strength, and have less potential for causing wound oedema and impaired cutaneous circulation. Interrupted sutures also allow the surgeon to make adjustments as needed to properly align wound edges as the wound is sutured. Disadvantages of interrupted sutures include the length of time required for their placement and the greater risk of crosshatched marks (ie, train tracks) across the suture line. The risk of crosshatching can be minimized by removing sutures early to prevent the development of suture tracks.

To sum up, the classical fusiform excision of melanocytic naevi offers certain advantages in the form of a better cosmetic scar, no chances of recurrences, histopathological examination of the lesion, minimal expertise in performing the simple elliptical excisions, and cost-effectiveness of the procedure as compared to repeated laser treatments.

Based on our results, we can conclude that treatment of melanocytic naevi whether acquired or congenital with classical fusiform excision along the relaxed skin tension lines remains an effective and time tested procedure. There are least chances of scarring provided the basic principles of skin cosmesis are not violated.

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A STUDY ON TINEA CAPITIS IN THE PRE SCHOOL AND SCHOOL GOING CHILDREN

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Abstract
Introduction: Tinea capitis is a superficial fungal infection of scalp and hair caused by various species of dermatophytes. The incidence of Tinea capitis varies from country to country and region to region.
Material and Methods: Fifty patients from the preschool going population were selected for the study.
Results and Discussion: Clinical presentation of disease revealed that black dot to be the commonest (32%) followed by grey patch (28%), kerion (20%) and favus type was the least (1%). Direct microscopy of hair in KOH preparations revealed that all clinically suspected patients of Tinea capitis had endothrix type in 56% of cases and ectothrix type in 44%.

Key words: Tinea capitis; fungal; hair; ectothrix; endothrix

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Introduction

Tinea capitis (TC) is a fungal infection of the scalp, hair follicles and hair shafts, especially common in the pediatric population and under tropical conditions [1,2]. The highest incidence is seen in children 3-7 years of age. The presence of symptoms like hyperkeratosis of scalp, seborrhea-like symptoms, excoriation secondary to pruritus, alopecia, broken hair or „black dot" appearance, cervical lymphadenopathy, pustules, or indurated or boggy plaques in a child should alert the dermatologist toward the possibility of TC [3,4].

TC is caused by various dermatophytes. The prevalence of various causative fungi varies according to the geographical area being studied [5,6]. TC can present as noninflammatory or inflammatory morphological variants. An early diagnosis is important to prevent transmission between children, especially siblings, and also to avoid possible scarring and permanent hair loss. The presence of tinea corporis or an id reaction in a child should also prompt a search for TC. As is true for most infectious diseases, the epidemiology of TC is in a constant state of flux, and varies considerably with respect to geography and specific population groups [7]. This fact prompted the present study, aimed at evaluating the clinical and etiologic profile of TC.

Aims

The major aim of this study was to determine the incidence, clinical presentation, age and sex distribution and seasonal variation of Tinea capitis among school going children. One objective was to relate the aetiological agent to the type of lesions.

Material and Methods

Fifty patients from the preschool going population were selected for the study from the dermatology outdoor department.There were 35 male children and 15 female children between 3 to 10 years of age. A detailed history was taken regarding the duration and pattern of hair loss. Demographic and socioeconomic data of the patients were recorded. Socioeconomic status of the children included patients in low income group, middle income group and high income group. Factors predisposing toward the spread of TC were analyzed. This included an assessment of the living conditions, history of any other family members affected and the hair care practices being followed. The patients were specifically asked about pets or any other prolonged contact with animals.Examination of the whole scalp was carried out to assess the type and extent of hair loss. The children included in our study were those presenting with patchy hair loss and easy pluckability of hair, with or without any associated inflammatory changes. Patients on any oral or topical antifungal therapy for the past 6 weeks were excluded from the study.

Patients were classified according to the morphological types of TC as non inflammatory black dot (BD), gray patch (GP), inflammatory (pustular, kerion or favus) or mixed infection (any combination of the above). The patients were thoroughly examined to assess for any evidence of tinea corporis, nail involvement, id reaction or lymphadenopathy. Siblings of affected children as well as close family contacts were also examined to assess for any hair loss. The patients were asked to come after head wash so as to remove any oil from the scalp. For all the patients, skin scrapings and hair fragments were collected from the affected areas in an aseptic manner. The material collected on a slide was immersed in potassium hydroxide (10% KOH) to prepare smears for microscopic examination. Both macroscopical and microscopical examinations were performed to identify the dermatophyte isolates. The slides were assessed under a low-power microscope to look for fungal arthrospores or any hyphae. If the spores were located on the surface of the hair shaft, without causing any distortion of hair architecture, the infection was classified as ectothrix. If the spores were seen inside the shaft and were destroying the hair fragment architecture, the infection was classified as endothrix. Presence of both the patterns in the same specimen was also recorded. The samples were also inoculated on Sabouraud's Dextrose Agar; with and without antibiotics (chloramphenicol, gentamicin and cycloheximide). This was done to identify the causative species involved. The clinical, microbiological and etiologic data were collected and correlated.

Results

The data was collected and the results were analysed.

Sr no	Morphological pattern	Number	Percentage
1	Black dot type	16	32
2	Grey patch	14	28
3	Kerion	10	20
4	Pustular	6	12
5	Mixed	4	8
	Total	50	100

Table I. Morphological pattern of patients

Sr no	Clinical type	Endothrix (28)	Ectothrix (22)
1	Black dot type (n=16)	10 (62.5%)	6 (37.5%)
2	Grey patch type (n=14)	6 (42.8%)	8 (57.1%)
3	Kerion (n = 10)	6 (60%)	4 (40%)
4	Pustular type (n=6)	4 (66.6%)	2 (33.3%)
5	Mixed (n=4)	2 (33.3%)	2 (33.3%)

Table II. Clinical and microscopic types of tinea capitis

Sr no	Species	Black dot	Grey patch	Kerion	Pustular	Mixed
1	Trichophyton violaceum	14	10	6	4	3
2	Trichophyton rubrum	-	1	-	-	-
3	Trichophyton tonsurans	-	-	2	-	-
4	Microsporum audounii	-	2	-	-	1
5	No growth	2	4	2	2	-

Table III. Causative agent of tinea capitis

Discussion

It was seen that the majority of children were in the age group of 3-10 years.Beyond this age group, the incidence declines because of the onset of puberty and seborrhea. Regarding distribution, male children patients numbered female children and male : female ratio was 2:1. Various conflicting views exist regarding the sexual predominance of TC. The low frequency in the females could be due to custom of regular application of vegetable oil over the scalp which has fungistatic properties. Some authorities believe that TC may be common in boys due to shorter hair, allowing easy access for circulating spores, while others believe that it may be more common in girls due to tight hair braiding. 60% patients showed inflammatory (Fig. 1) tinea capitis and 32% patients showed non inflammatory tinea capitis. Family history of TC which was seen in 29% of patients may be due to sharing of articles like towels, combs, cloth cap etc. by other family members. Fungi was recovered on culture in 65% patients and regarding the causative agent of fungi no growth was seen in 20% patients.

82% of the children belonged to the lower middle and 22% belonged to the lower income groups. There were 15 sets of siblings among these patients, with each set having two to three children. A history of sharing of combs and hair accessories was elicited in 68% of the patients. Only 15% gave a history of pets at home or prolonged contact with animals.

An attempt was made to correlate the clinical and microscopic types of TC. It was seen that although the endothrix pattern was more common for BDTC, the ectothrix pattern is also seen in a large number of cases. Similarly, ectothrix invasion is more common for gray patch tinea capitis (GPTC), although endothrix cases can also be seen. Also, most of the cases with a mixed pattern of invasion on KOH belonged to the GP + BD morphology. Culture specimens from all the cases were examined. No growth at the end of 6 weeks was recorded in 20% of the cases (10 cases). Of those showing growth of fungal elements, *Trichophyton violaceum* was the most common isolate in 74% (37 cases). This was followed by *T. rubrum* (one case), *T. tonsurans* (two cases), *Microsporum audouinii* (two cases). A correlation of fungal species isolated with the clinical type of TC was drawn out. It can be seen that Trichophyton violaceum was responsible for BDTC (Fig. 2) in most of the cases. *M. audouinii* were responsible for a GPTC pattern and *T. tonsurans* was isolated from cases of kerion (Fig. 3). Majority of TC cases were from urban area (78%) and family history of dermatophytoses was present in 25% of cases. There was seasonal incidence of cases of TC. Incidence of disease was slightly higher (45%) in post monsoon period (July-October) and the percentage of cases from January to April was also 38%. Incidence was low, 5% and 3.5% in extreme summer (May-June) and begining of winter (Novermber-December) respectively.

Clinical presentation of disease revealed that black dot to be the commonest (32%) followed by grey patch (28%), kerion (20%) and favus type was the least (1%). Direct microscopy of hair in KOH preparations revealed that all clinically suspected patients of TC had endothrix type in 56% of cases and ectothrix type in 44%. The percentage of endothrix infection in black dot type was 62.5%, grey patch 42.8%, Kerion 60% and favus 1%, whereas ectothrix hair involvement was seen only in grey patch (57.2%) and kerion (40%). It was observed that *T. violaceum* was main isolate from black dot type while *T. mentagrophyte* was the main isolate from grey patch and kerion lesions. TC is a common fungal infection, particularly among children in urban regions [8-10]. More often than not, it presents with mild scaling and little hair loss, which is reversible. However, in a few cases, it may be characterized by intense inflammation and subsequent cicatricial alopecia, which causes permanent cosmetic disfigurement. Also, the infection is highly contagious and, hence, needs to be recognized and treated early to prevent transmission to siblings and costudents [11-13]. Awareness in patients regarding disease was good (88%) and also incidence was higher in urban areas compared to rural population.

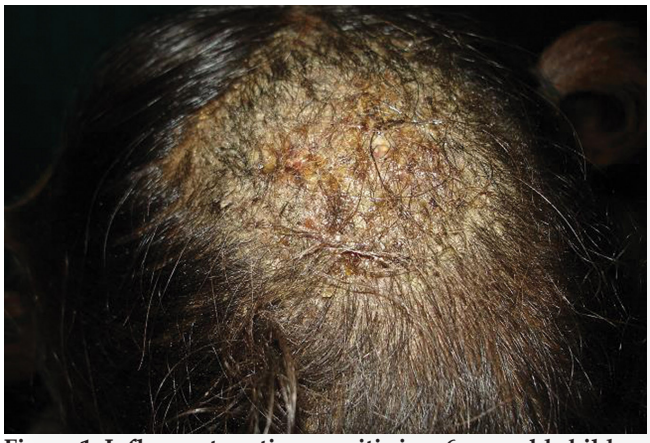

Figure 1. Inflammatory tinea capitis in a 6 year old child

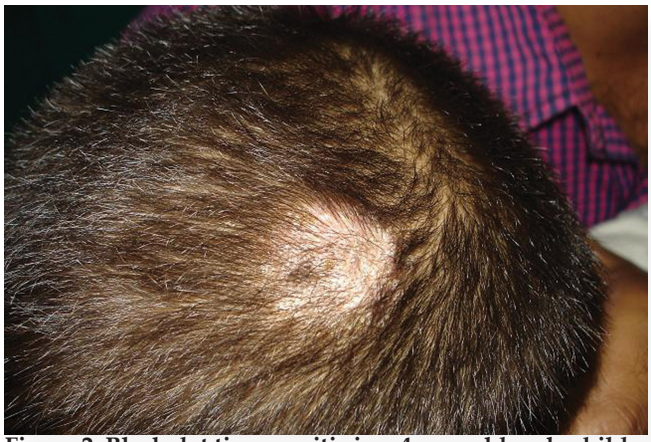

Figure 2. Black dot tinea capitis in a 4 year old male child

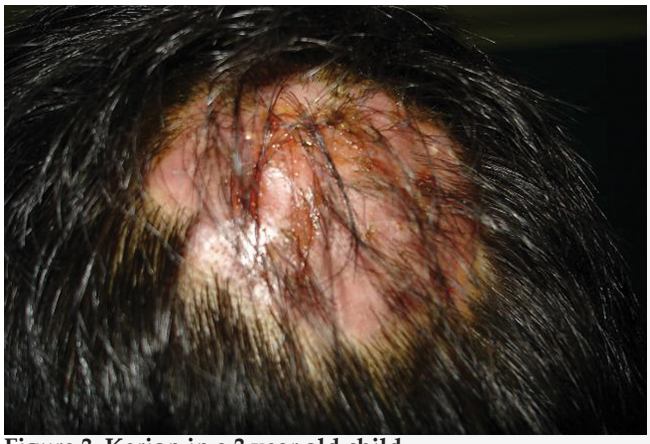

Figure 3. Kerion in a 3 year old child

Conclusion

Our study threw up interesting findings. BDTC was the most common type of TC and *Trichophyton violaceum* was the most common species isolated. KOH examination and culture were useful diagnostic methods. However, the clinical morphology or KOH findings were not found to be clearly predictive of the species involved. Mixed patterns were observed both on clinical examination as well as on KOH examination. In our study results of microscopic examination of hair in KOH mount when correlated with culture, it was seen that majority of dermatophytes isolated was *Trichophyton violaceum* which caused endothrix type of hair infection. Also, the percentage of dermatophytes on culture in our study was 65%.

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A STUDY ON TINEA CAPITIS IN THE PRE SCHOOL AND SCHOOL GOING CHILDREN

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This study by Puri N & Puri A provides an overview of the prevalence of Tinea capitis among school children in some parts of India.

Tinea capitis (TC) (scalp ringworm) poses a significant health problem in many underdeveloped and developing countries. It is the most common dermatophyte infection in childhood and primarily caused by fungal dermoatophytes in the *Trichophyton* and *Microsporum* genus. The epidemiology of TC varies within different geographical areas throughout the world and a number of studies have clearly demonstrated that there is a significant increase in its incidence and a change in the pattern of infectious agents in different countries and a similar trend can be observed in India as well.

Data provided by the authors are interesting but the results could have been expanded and compared with other recent studies. Given the sporadic nature of the causal agents, which vary within different geographical areas, a proper comparison is warranted so that early diagnosis and adequate treatment can be done promptly. While the authors here found that Black dot type and T. *violaceum* were more prevalent; more males were infected than females as the latter frequently apply vegetable oil over scalp, such is not the case in other parts of the world, even in different states in India.

For instance, a recent study assessing the prevalence of *Tinea capitis* among urban school children in Kolkata, (India), Bindu et al (2012) found that the prevalence rate was significantly high among boys but there was no significant difference in prevalence of infection among coconut oil users and castor oil users. In addition, they also reported that the commonest clinical type of infection found was dull grey patches as compared to Black dot type observed by Puri N & Puri A. However, it should be mentioned that the use of oils to reduce TC is quite well known as these oils contain different percentages of various saturated and unsaturated fatty acids which largely determine their toxicity against dermatophytes (Garg & Muller, 1992).

In another study, Ayaya et al (2001) reported there were more males school children in Africa that were infected than females (ratio of 2:1) and this corroborates with what Puri N and Puri A found. In addition, most of the cases reported were of endothrix and similar findings are reported by Puri N and Puri A. The major difference between the two studies was in the infectious agent. While Ayaya et al (2001) commonly isolated *T. tonsurans*, Puri N and Puri A reported *T. violaceum* as more prevalent. Other studies that have reported *violaceum* as more prevalent include those of Chepchirchir et al (2009) in Kenya. The latter also found that the grey patch form was the dominant clinical manifestation contrary to Puri N and Puri A. In Kuwait, the non-inflammatory "grey patch" variety has also been reported to be the most common clinical type, followed by black-dot variant and that *T. violaceum* was the most common fungus responsible for the black-dot variety (Nawaf et al 2003).

It is noteworthy to mention that not only *Trichophyton* might be more prevalent than *Microsporum* and this may vary from regions to regions. Mycological data on TC collected in urban and rural areas of the Dominican Republic by Arenas et al (2010) reported a higher occurrence of *Microsporum audouinii* than *Trichophyton tonsurans*, *Microsporum canis* and *Trichophyton violaceum* from children in urban areas, whereas *T. tonsurans* and *T. mentagrophytes* were isolated from those in rural areas and in addition *T. violaceum* was rarely reported (in contrast to Puri N and Puri A). Similarly, Ross et al (1993) found that the dermatophytes responsible for TC in Puerto Rico were mainly *M. canis*.

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A STUDY ON EFFICACY OF ORAL ZINC THERAPY FOR TREATMENT OF ACRODERMATITIS ENTEROPATHICA

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Abstract
Introduction: Acrodermatitis enteropathica (AE) is a disorder of zinc metabolism that occurs in one of two forms: an inborn (congenital) form and an acquired form. The inborn form of AE is a rare genetic disorder characterized by intestinal abnormalities that leads to inability to absorb zinc from the intestine. The lack of zinc presents, characteristically, as skin inflammation with pustules occurring around the mouth and/or anus, diarrhea.
Aims: To study the efficacy of oral zinc therapy on thirty patients of acrodermatitis enteropathica.
Methods: Thirty clinically diagnosed patients of acrodermatitis enteropathica were taken for the study. The criterion of diagnosis of this condition was the clinical picture of symmetrical vesiculo-pustular dermatitis, in upper and lower limbs and periorificial regions**.**
Results and Discussion: In our study maximum (50%) patients were below 1 year of age, 33.3% patients were between 13-24 months of age, 6.66% patients were between 25- 36 months and 37- 48 months each and 3.33% patients were between 49- 60 months of age. Nail changes were seen in 60% children. Also, it was seen that perioral area was the commonest (86.6%) site involved, followed by anogenital area in 80% patients, palms and soles were involved in 66.6% patients, arms in 46.6% patients and legs were seen in 40% patients. Regarding clinical features, dermatitis was seen in 100%patients, alopecia was seen in 40% patients, diarrhea in 60% patients and mental disturbances were seen in 30 % patients.

Key words: acrodermatitis; enteropathica; zinc

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Introduction

Acrodermatitis enteropathica is a rare genetic disorder characterised by diarrhoea, an inflammatory rash around the mouth and/or anus and hair los [1]. Acrodermatitis enteropathica is due to malabsorption of zinc through the intestinal cells.The precise cause is not known, but it may relate to mutations in a gene (SLC39A4) that codes the zinc transporter protein, ZIP4 [2,3]. It is thought that the missing protein may be responsible for decreased zinc uptake and abnormal zinc metabolism. To have congenital acrodermatitis enteropathica you must inherit two defective genes (one from each parent) i.e. the inheritance is autosomal recessive [4]. If an individual receives one normal gene and one defective gene, the person will be a carrier for the disease, but usually will not show symptoms. Symptoms usually occur in bottle-fed infants within a few days or weeks after birth and breast-fed infants soon after weaning. Both males and females are equally affected.

Zinc is an essential component of the diet. Zinc in human milk is more absorbable than that from infant formulas or cow's milk, hence the later onset of acrodermatitis enteropathica in breast-fed babies compared to formula-fed babies [5]. Zinc is also found in meat, shellfish and wheat germ. Foods of plant origin are mostly low in zinc. Phytates present in cereals and soy and high levels of calcium, can reduce the absorption of zinc through the duodenum. Zinc is needed to assist metalloenzymes that are involved in many cellular processes throughout the body [6,7]. These include the production of anti-inflammatory agents (cytokines and antioxidants) and the normal functioning of the brain. If zinc deficiency is suspected, the following investigations may be helpful [8].

- Serum/plasma zinc levels confirm the diagnosis (normal levels are 60-140 microgram/dL);
- Urinary zinc excretion may be reduced;
- Blood count may reveal anaemia;
- Skin biopsy may show characteristic features.

Acrodermatitis enteropathica is characterized by chronic diarrhea which may be mild or severe, and the presence of fatty substances in the feces (steatorrhea) [9]. In the congenital form symptoms start gradually, frequently at the time of weaning of an infant.

The skin around body openings such as the mouth, anus, and eyes; and the skin on elbows, knees, hands, and feet becomes inflamed. Skin lesions are usually blistered (vesicobullous) and after drying out become psoriasis-like. The skin around the nails may also be inflamed and the nail may be abnormal due to malnourished tissue [10]. Hair loss on the scalp, eyelids, and eyebrows may be total (alopecia). Inflammation of the membrane that lines the eyelid (conjunctivitis), usually also occurs.

Material and Methods

Thirty clinically diagnosed patients of acrodermatitis enteropathica were taken for the study. The criterion of diagnosis of this condition was the clinical picture of symmetrical vesiculo-pustular dermatitis, in upper and lower limbs and periorificial regions. Another diagnostic criteria was prompt response to oral zinc therapy.The patents selected were below 5 years of age. Written informed consent was taken from the parents of the children before the start of the study. Prior approval of the hospital ethical committee was taken for the study. The routine haemogram was performed of all the patients. Histopathological examination was done in those patients wherever diagnosis was in doubt. Serum zinc levels were calculated before the treatment was started. The children were put on oral zinc therapy, if serum zinc levels were found to be below 50 µgm/dl. The children with zinc deficiency were given oral zinc supplements in the form of syrup zinc sulphate given in the dose of 5mg/kg body weight. The oral zinc was continued for 4weeks or till the clearance of dermatitis, whichever was earlier. Post treatment serum zinc levels were again estimated.

The specimens for plasma zinc were collected in plastic syringes or acid washed vacutainer tubes with no rubber stopper to prevent exogenous contamination that could lead to spuriously normal measurements. Plasma zinc concentrations of less than 50 µgm/dl were suggestive, but not diagnostic of acrodermatitis enteropathica.

Results and Discussion

The results were tabulated and the data was analysed.

Maximum (50%) patients were below 1 year of age, 33.3% patients were between 13-24 months of age (Tabl. I).

Table II shows that there were 60% male children and 40% female children.

Nail changes were seen in 60% children. 60% patients showed nail ridging, 40% patients had paronychia and 6.66% patients had nail dystrophy (Tabl. III).

Table IV shows that perioral area was the commonest (86.6%) site involved, followed by anogenital area in 80% patients, palms and soles were involved in 66.6% patients, arms in 46.6% patients and legs were seen in 40% patients.

Cermatitis was seen in 100% patients, alopecia was seen in 40% patients, diarrhea in 60% patients and mental disturbances were seen in 30 % patients (Tabl. V).

Sr no	Age (months)	Number	Percentage
1	< 12 months	15	50%
2	13-24	10	33.3%
3	25-36	2	6.66%
4	37-48	2	6.66%
5	49-60	1	3.33%
	Total	30	100

Table I. Age distribution of children with acrodermatitis enteropathica

Sr No	Sex	Number	Percentage
1	Male children	18	60%
2	Female children	12	40%
	Total	30	100

Table II. Sex distribution of children with acrodermatitis enteropathica

Sr No	Sex	Number	Percentage
1	Nail ridging	18	60%
2	Paronychia	12	40%
3	Nail dystrophy	2	6.66%

Table III. Nail changes in patients with acrodermatitis enteropathica

Sr No	Site	Number	Percentage
1	Perioral	26	86.6%
2	Anogenital	24	80%
3	Palms and soles	20	66.6%
4	Arms	14	46.6%
5	Legs	12	40%
6	Trunk	8	26.6%

Table IV. Site of involvement of skin lesions

Sr No	Clinical Features	Number	Percentage
1	Dermatitis	30	100
2	Alopecia	12	40
3	Diarrhoea	18	60
4	Mental disturbances	9	30

Table V. Clinical features of acrodermatitis enteropathica

Acrodermatitis enteropathica is a rare inherited form of zinc deficiency, characterized by periorificial and acral dermatitis, alopecia, and diarrhea [11]. The inherited form of acrodermatitis enteropathica was usually fatal until the role of zinc was discovered in 1973. It should be treated with 1 mg/kg body wait of oral zinc supplementation per day for life. Zinc gluconate is better tolerated than zinc sulfate. Zinc can be given during pregnancy. After zinc replacement the skin lesions heal within one to two weeks, diarrhoea ceases and irritability and depression of mood improve within 24 hrs.Secondary bacterial and/or fungal infection of lesions require appropriate antibiotic therapy. Additionally, zinc deficiency can present in full-term breastfed infants as a result of low maternal serum zinc levels or a defect in mammary zinc secretion [12]. Thus, not all infants who have an acrodermatitis enteropathica–like presentation have the genetic disorder.The clinical findings of infant with acrodermatitis enteropathica are as follows: Infants are typically irritable and often inconsolable, and they show a slowing or cessation of growth and development.

The skin shows erythematous, dry and scaly patches and plaques which may evolve into crusted, vesiculobullous, erosive, psoriasiform, and pustular lesions. Lesions are predominantly distributed in a periorificial and acral pattern and may become secondarily infected with *Staphylococcus aureus* or *Candida albicans*. Red and inflamed patches of dry and scaly skin, particularly around body openings such as the mouth, anus, and eyes, and the skin on elbows, knees, hands, and feet. It may look like atopic dermatitis. Patches evolve into crusted, blistered, pus-filled and eroded lesions. There is usually a sharp demarcation between the affected area and normal skin. The mucosa shows angular cheilitis, glossitis, conjunctivitis, blepharitis, punctate keratopathy and photophobia. The nails shows paronychia and nai dystrophy is typical.Also the patients have loss of scalp hair, eyebrows, and eyelashes.

Other features of acrodermatitis enteropathica includes conjunctivitis, sensitivity to light, loss of appetite, diarrhoea, mild or severe, irritability (babies cry and whine incessantly), depressed mood and growth failure. Histological evaluation of a skin biopsy specimen is characteristic, but the same findings can be seen in other nutritional disorders [13]. The histological findings vary with the age of the lesion. Early lesions show confluent parakeratosis associated with a reduced granular layer. Often, exocytosis of neutrophils into the epidermis is noted, which may be acanthotic and exhibit slight spongiosis. The intracellular edema eventuates into pallor of the upper third of the epidermis. Subsequently, subcorneal and intraepidermal clefts may develop as a result of massive ballooning and reticular degeneration, with necrosis of the keratinocytes. In late lesions, psoriasiform hyperplasia of the epidermis and less epidermal pallor are noted.

In our study maximum (50%) patients were below 1 year of age, 33.3% patients were between 13-24 months of age, 6.66% patients were between 25-36 months and 37-48 months each and 3.33% patients were between 49- 60 months of age. There were 60% male children and 40% female children. Nail changes were seen in 60% children. 60% patients showed nail ridging, 40% patients had paronychia and 6.66% patients had nail (Fig. 1, 2). These children had low body weight, were apathetic and irritable. Alopecia of scalp and eyebrow hair was seen in 38% children. Also, it was seen that perioral area was the commonest (86.6%) site involved, followed by anogenital area (Fig. 3) in 80% patients, palms and soles were involved in 66.6% patients, arms in 46.6% patients and legs were seen in 40% patients.

Regarding clinical features, dermatitis was seen in 100%patients, alopecia was seen in 40% patients, diarrhea in 60% patients and mental disturbances were seen in 30 % patients. Most of the children had low body weight,were apathetic and irritable. Out of 30 cases, one patient had intractable diarrhea and was severely dehydrated and died of electrolyte imbalance. Three cases had positive family history.Two of their siblings haad suffered from similar disease. Some of the children had anaemia and ascariasis infestation. In our study,clinical improvement started within 2 - 3 weeks of zinc supplementation (Fig. 4, 5).

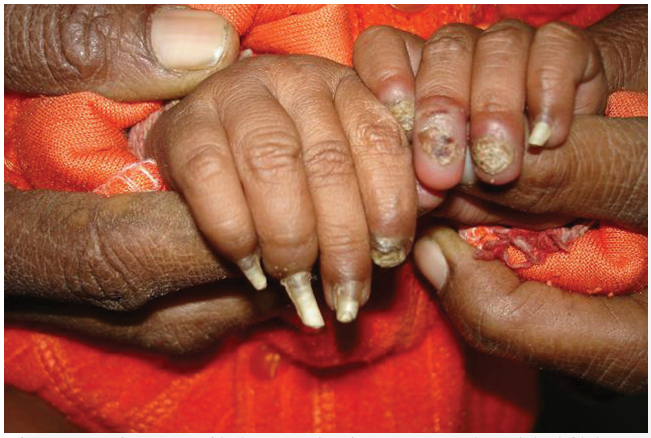

Figure 1. Finger nail dystrophy in a 3 months old child

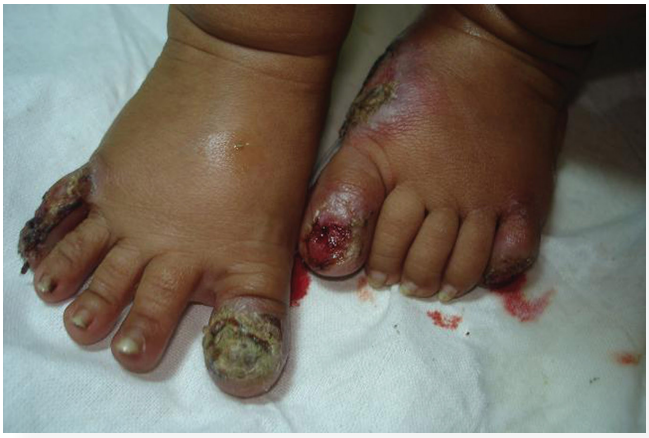

Figure 2. Toe nail with subungual haemorrhage in toe nails

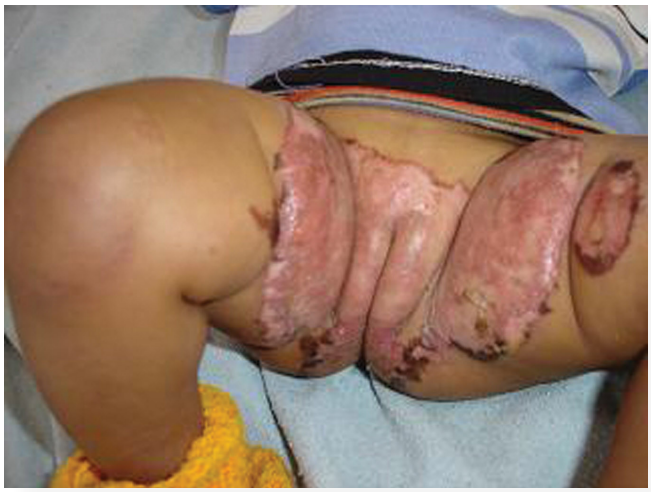

Figure 3. Erythematous plaque in a 7 months old female child

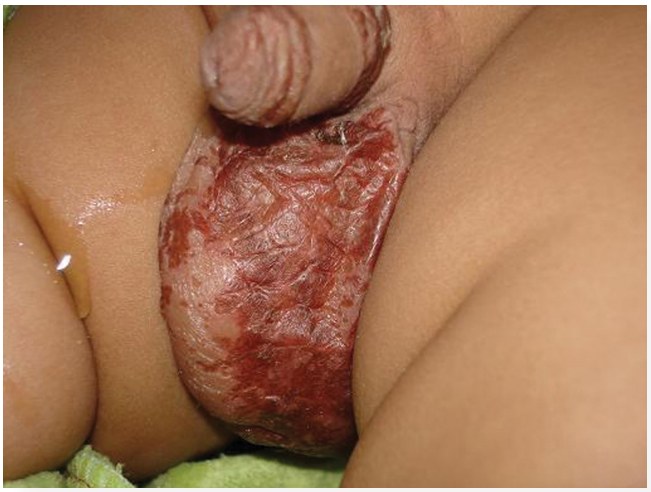

Figure 4. Erythema over the genital area in a 5 months old infant befeore treatment

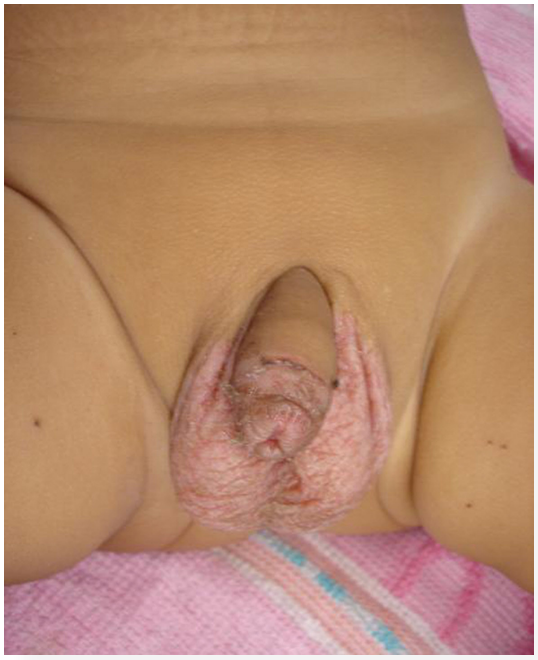

Figure 5. After treatment with zinc therapy in the same child

Treatment of acrodermatitis enteropathica requires lifelong zinc supplementation. Acrodermatitis enteropathica is treated with zinc supplements in the form of zinc sulfate. These supplements should be given as soon as diagnosis of the disorder is made and they have to be continued for life. The drug Diodoquin (iodoquinol) is another treatment that usually clears up symptoms within a week. If the disorder is caused by intravenous feeding, adding zinc supplements to the nutritional regimen can prevent and/or clear up manifestations of AE.Typically, 3-5 mg/kg of zinc gluconate or sulfate is administered orally each day. Clinical improvement occurs prior to any significant change in the plasma zinc levels, usually within days to weeks of initiating treatment [14]. Monitor serum zinc levels and alkaline phosphatase values every 3-6 months [15,16]. Acrodermatitis enteropathica exacerbation during pregnancy or the stress of disease may require an increase in therapy. Warm compresses to remove the scale crust, followed by application of white petrolatum to eroded skin lesions, may enhance reepithelialization when used concurrently with zinc replacement [17]. Genetic counseling is recommended for families of patients with the congenital form of acrodermatitis enteropathica. Although no special diet is required for acrodermatitis enteropathica patients, as long as zinc supplementation is continued, certain foods contain increased levels of zinc, including oysters, crab, beef, pork, and fowl. Zinc content is directly related to protein [18,19].

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A STUDY ON EFFICACY OF ORAL ZINC THERAPY FOR TREATMENT OF ACRODERMATITIS ENTEROPATHICA

by Neerja Puri

comment: Subtitle: **IMPORTANCE OF ZINC IN MEDICINE OF 21ST CENTURY**

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Zinc is an essential mineral which belongs to the most important and non-substitutable micronutrients in human. Its significance for health was invented in 60s years of 20th century. Zinc possesses many important characteristics and plays an important role in the maintenance of equilibrium in organism. It is a part of many enzymes and possesses complex antioxidant and anti-inflammatory activity. Another very important function of zinc is its role in the immune system. Zinc stimulates the processes of phagocytosis, activated the complement system, supports the maturation of B- and T-lymphocytes and is very important for the production of immunoglobulins [1]. It is also essential for the regulation of inflammatory processes in the organism [2]. In case of zinc deficiency, the immune dysfunction can be observed. The zinc deficiency is one of the most important acquired secondary immunodeficiencies worldwide.

The zinc deficiency was observed in several diseases and chronic conditions. It was showed that the severe asthmatics and the patients with severe forms of atopic eczema have the zinc deficiency and its supplementation could support the standard anti-allergic therapy [3,4]. Zinc could be also used for the treatment and prevention of acute and recurrent respiratory tract infections [5-7].

The zinc deficiency still represents the very important problem worldwide. It is usually associated with malnutrition, especially in case of high contents of phytates in the food. Zinc deficiency is a part of genetically-determined disease – acrodermatitis enteropathica. Despites the progression of the modern medicine, it is necessary to consider this diagnosis in the context of the differential diagnosis of chronic dermatosis. As the current published study shows, the simple and cheap therapy consisting of supplementation of zinc is effective in the treatment of this disease [8]. Author clearly described the complexity of clinical manifestation of acrodermatitis enteropathica, which involves not only the skin changes but also gastrointestinal and psycho-neurological changes and disturbances. Despites the severe chronical changes in the different organ systems, the oral therapy with zinc could significantly improve the clinical status of these patients.

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CUTANEOUS MANIFESTATIONS OF DERMATOMYOSITIS IN MALE PATIENT: A RARE REPORT

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Abstract
Introduction: Dermatomyositis is an idiopathic inflammatory myopathy. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, the esophagus, the lungs, and, less commonly, the heart.
Case report: It is presented with characteristic cutaneous findings like skin rash as well as progressive symmetrical proximal muscle weakness. Its prevalence rate is approximately one per 100,000 in the general population with a female to male predominance of about 2:1.
Conclusion: We report a male patient with the classical features of dermatomyositis in whom cutaneous changes preceded muscle weakness.

Key words: dermatomyositis; Gottron's sign; helitrop sign; poikiloderma; shawl sign

Cite this article:
Ganesh Dhavalshankh, Archana Dhavalshankh, Vaishali Masvekar: Cutaneous manifestations of dermatomyositis in male patient: a rare report. Our Dermatol Online. 2013; 4(2): 168-171.

Introduction

Dermatomyositis is an idiopathic inflammatory myopathy (IIM) [1,2]. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, the esophagus, the lungs, and, less commonly, the heart [3,4]. It is presented with characteristic cutaneous findings like skin rash as well as progressive symmetrical proximal muscle weakness. It has extramuscular manifestations such as joint contractures, dysphagia, cardiac disturbance, pulmonary symptoms, and subcutaneous calcifications. DM also has an association with malignant disease, and various autoimmune and connective tissue diseases. The average age at diagnosis is 40 yr, and almost twice as many women are affected as men [3,4]. It's prevalence rate is approximately one per 100,000 in the general population with a female to male predominance of about 2:1. DM is usually associated with an underlying malignancy, and its prevalence is even rarer without coexistent cancer [5,6]. Dermatomyositis is a connective tissue disorder constituting inflammatory myopathy along with characteristic cutaneous markers. The diagnostic criterias for the disease has been defined [7]. The various cutaneous manifestations in dermatomyositis, may precede or follow myositis [8,9]. However dermatomyositis can present without muscle weakness [10].

Objectives

We report a male with the classical features of dermatomyositis in whom cutaneous changes preceded muscle weakness. The presenting cutaneous lesions in dermatomyositis include a heliotrope rash with edema, photosensitivity, Gottron's papules and poikiloderma.

Case Report

A 32 years male patient with previous healthy condition presented to us 2 months back with red colored rash & swelling around both eyes and cheeks. He noticed similar type of rash on both hands, both thighs and both shoulders extending up to mid back. He also complained of severe muscle weakness & pain in both shoulder joints .He noticed difficulty in combing his hair and aggravation of rash when exposed to sun. The initial maculopapular erythematous rash started on dorsal aspect of both hands and then spread to involve face, back and abdomen since 6 months. Since last two months he noticed difficulty in climbing stairs, getting up from squatting position and combing his hair. The muscle weakness was bilateral and gradually progressive without any fasciculations. The clinical examination revealed bilateral periorbital erythematous rash and edema covered with fine white scales suggestive of heliotrope rash (Fig. 1) which is highly suggestive of dermatomyositis.

Similar type of rash was also present on both malar prominences, dorsii of both hands, abdomen and V of neck mostly covering sun-exposed areas suggestive of poikiloderma (Fig. 2). The rash extended on upper part of the back suggestive of shawl sign (Fig. 3). There were hyperpigmentation papules found on bony prominences particularly the metacarpophalangeal and iterphalangeal joints suggestive of Gottron papules and Gottron's sign (Fig. 4).

The systemic examination revealed mild pallor. There was weakness of various group of muscles up to grade II. None of the muscles were atrophic but tenderness was present in the muscles of extremities. No fasciculations were seen in any of the muscles. Deep tendon, abdominal and plantar reflexes were normal. The breasts, genitalia and gastrointestinal tract were normal. No periinguinal erythema and nailfold telangiectasia or cuticular dystrophy observed. No mechanics hand lesions (fissured scaly hyperkeratosis) were found. Laboratory data revealed haemoglobin 13.88 gm/dl, total leukocyte count 5900/mm^3, P73L19M4E4B0, platelets 1.9 lacs/mm^3 and ESR 16 mm/hr. Urine analysis, blood glucose, blood urea, serum creatinine and serum uric acid were within normal limits. LE cells and antinuclear factors (by indirect immunoflourescence) were not detected (0.66) and anti-dsDNA was negative (19.13 IU/ml). CPK was 195 units/dl (n= 10-70), CPK (MB) 36.78 units/dl (n<5 percent of total), LDH 299 units/dl (n=200-450) and SGOT was 23 units/ml (n=15-45). RA test was negative. Ultrasonography of abdomen including pelvis, and ECG were normal. A skin biopsy showed mild hyperkeratosis, patchy parakeratosis, spongiosis, focal thinning, hydropic degeneration of basal cell layer with lymphocytic exocytosis. The upper dermis showed band-like lymphocytic infiltrate, incontinence of pigment and dilated capillaries (Fig. 5).

The patient received 10mg of oral prednisolone twice daily along with 200mg of hydroxychloroquine twice a day and once weekly 10mg of methotrexate. The therapy was supplemented with oral antioxidants, folic acid and topical diluted mometasone furoate at night and broad spectrum sunscreen during daytime. The muscle weakness was gradually improved over a period of four weeks and the muscle enzymes also reduced. The prednisolone was then gradually tapered to 5mg over a span of four weeks. Presently he is off the steroids and is being maintained on 5mg of methotrexate once a week and same dose of hydroxychloroquine. The patient has regained his muscle power and there is a drastic improvement in his rash.

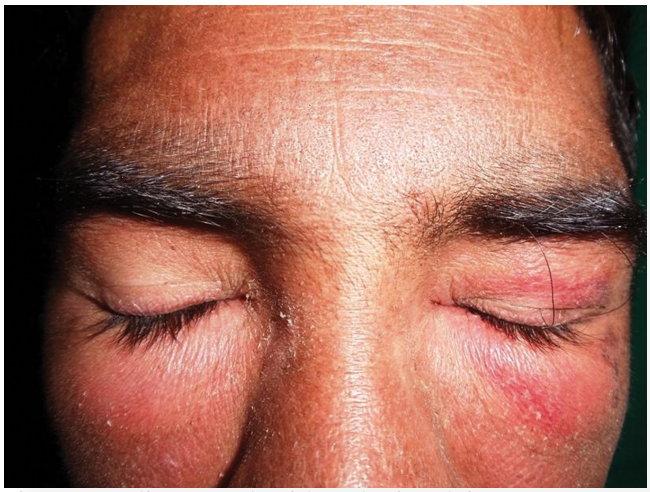

Figure 1. Helitrope rash with scales involving periorbital skin

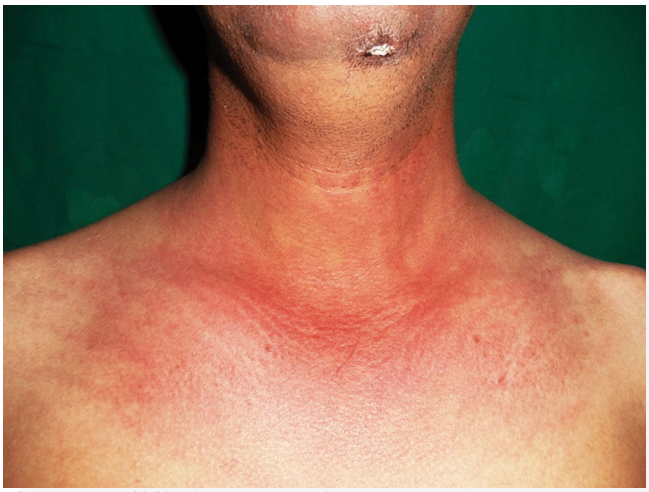

Figure 2. Poikiloderma – Erythematous rash on sun exposed area

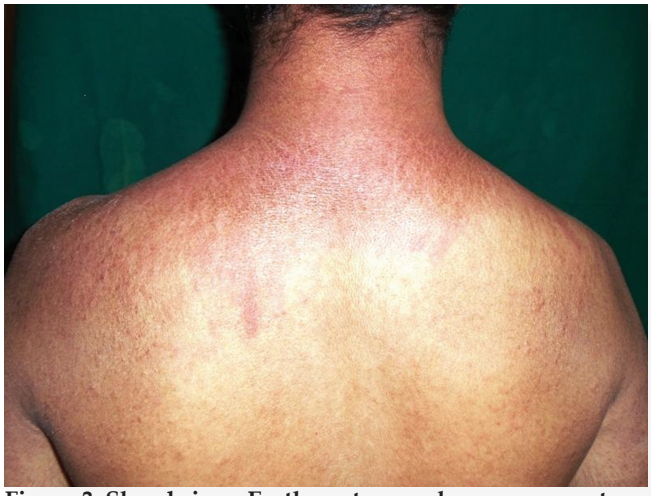

Figure 3. Shawl sign - Erythematous rash on upper part of back

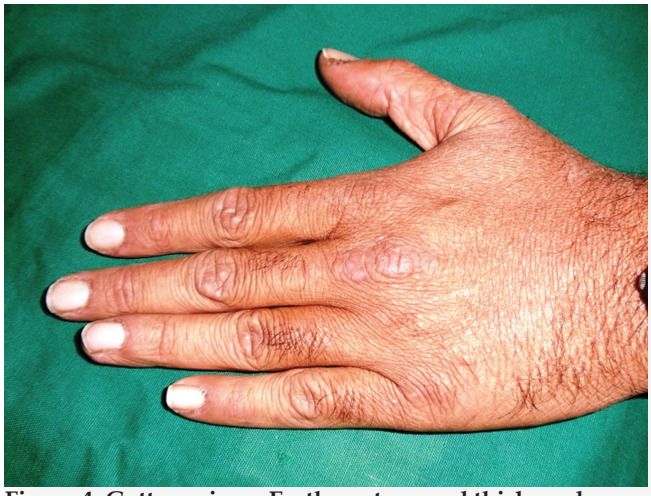

Figure 4. Gottron sign – Erythematous and thickened skin at joints

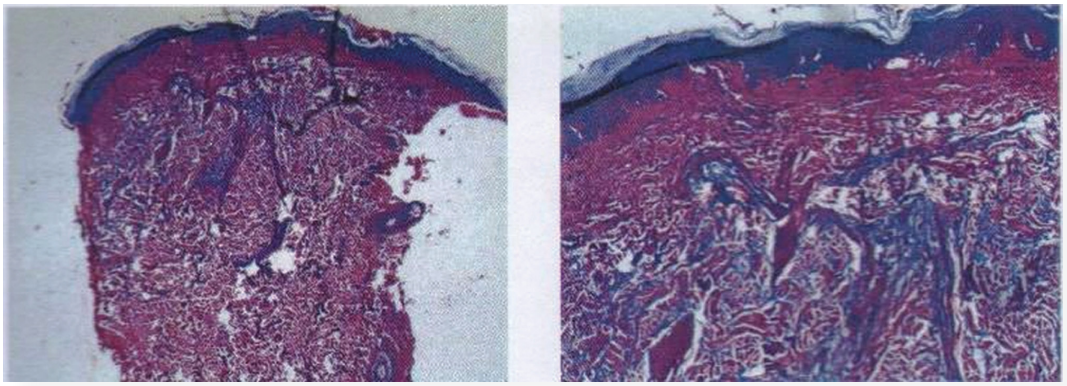

Figure 5. Histopatological changes

Discussion and Conclusion

Bohan and Peter (1975) suggested a set of criteria which aid in diagnosing dermatomyositis. Dermatomyositis and polymyositis is diagnosed in patient with typical cutaneous features, progressive proximal symmetrical muscle weakness, elevated muscle enzyme levels and abnormal findings from muscle biopsy. Patient of dermatomyosititis often present with skin diseaseas an initial manifestations. In nearly 40% patients skin disease may be the sole manifestation as the onset. This patient is presented and having mostly all characteristic skin manifestations, including Gottron's sign, the shawl sign, the heliotrope rash, poikiloderma and a generalized erythroderma. This patient manifested all classic skin signs with his flare up of DM [1,2,5,7]. Muscle disease may occur concurrently or it may precede the skin disease or it may follow the skin disease by weeks to years (Bohan 1975). Skin rash often precedes the onset of weakness by weeks to months [11]. Early in disease course, rash and muscle enzyme elevations may be the sole manifestations of DM [12]. In our study, patient had proximal muscle weakness since1 week but the rash preceded muscle weakness and started since 1 year. Rockerbie NR et al [8] reviewed 50 patients of dermatomyositis retrospectively and was found that cutaneous changes sometimes preceded muscle weakness more than a year before the onset of muscle weakness. These findings suggest that the characteristic dermatomyositis eruption without muscle weakness should not preclude a diagnosis of dermatomyositis. This patient presented to us with similar complaints that rash followed by muscle weakness. Our case satisfied all the criteria for the definite diagnosis of dermatomyositis. The various cutaneous lesions, in dermatomyositis, may precede, occur simultaneously, or follow the onset of muscle weakness. Dermatomyositis sine myositis has also been documented [2]. The cutaneous lesions usually precede the onset of weakness by 3 to 6 months, [6] however, Pearson [11] has described one case, with skin involvement for 13 years. It is known that DM has a bimodal age distribution: one peak occurs in children between 5-14 yr of age and a second, larger peak occurs between 45-64 yr of age [13,14]. In our study, onset age was 32 years. It is reported that females outnumber males by 2:1 [13,14]. We reported this in male patient. The association between dermatomyositis (and possibly polymyositis) and cancer has long been recognized [15-19], but no any clinicopathological findings seen relevant in this case. Though etiology of the disease is unknown, it is believed to be initiated by viral infection and altered immune response. Lymphocyte mediated muscle cell damage and small vessel damage are important central pathogenetic factors. Usually vascular deposits of immune complexes and complement are associated with endothelial cell injury and small vessel obstruction. Dermatomyositis is usually associated with CD4 T cells and B cells infiltrating the muscles whereas polymyositis is associated with CD8 cytotoxic T cells. This case shows similar biopsy changes like lymphocytic infiltrate, incontinence of pigment and dilated capillaries.

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ATROPHIC TYPE OF MORPHEA PROFUNDUS - AN INDIAN EXPERIENCE

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Abstract
Localized scleroderma (also called morphea) is a term encompassing a spectrum of sclerotic autoimmune diseases that primarily affect the skin, but might also involve underlying structures such as the fat, fascia, muscle, and bones. Morphea profundus presenting with atrophic lesions has rarely been reported in the literature. Here we report two cases of morphea profundus presenting with noninflammatory depressed plaques, without any significant skin induration, pigmentation or textural change. Histopathology was confirmatory for morphea profundus.

Key words: morphea profundus; atrophic; multiple

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Introduction

The term „morphea" includes a wide spectrum of clinical entities, varying from localized plaques of only cosmetic importance to deep lesions resulting in considerable morbidity for the patient. Four different types of deep morphea have been distinguished: Subcutaneous Morphea, Eosinophilic Fasciitis, Disabling pansclerotic morphea and Morphea Profundus [1]. Morphea profundus is a rare disease and it often has a progressive course with physical and psychological sequelae [2]. Morphea profundus usually presents with early induration followed by atrophy of subcutaneous tissue and on occasion that of muscle [3]. Here we report two cases of atrophic morphea profundus presenting without any significant skin induration, pigmentation or textural change, the published literature on this type of presentation is sparse.

Case Report

Two female patients presented to us with asymptomatic atrophy of skin on multiple sites of the body.

Case-1:

Our first patient was 24 years old and first noticed circumscribed swelling on left arm 2 years back. It was small to begin with and later gradually increased in size and further, after 4-5 months she noticed atrophy of the skin in the same region. She noticed similar lesions adjacent to the old lesion on left arm and also on right arm and on left side of face at inner margin of eye. On examination, there were atrophic areas on her left upper arm (Fig. 1) and right upper arm (Fig. 2) with ill-defined margins. The skin overlying the lesions showed no obvious induration or tenderness. Multiple nodules were present varying in size from 2cm × 1cm to 5cm ×3 cm, hard in consistency, mobile, with skin pinchable over them, distal to the atrophic areas. Swelling of the face involving left lower eyelid and maxillary region measuring about 6 cm × 8 cm was present (Fig. 3). Examination of the surrounding skin, hair, nails and other systems did not reveal any abnormality.

Case-2:

Our second patient was 16 years old and presented with asymptomatic atrophy of the skin of both upper arms and right side of face since seven months. It was first noticed on her right arm followed in a few months by similar lesions gradually appearing on her left arm and her face.
On examination, atrophic areas on right side of face (Fig. 4), with ill-defined margins and patchy loss of hair was present in temporal, maxillary and mandibular region.
Atrophic areas were also present on right upper arm (Fig. 5) and left upper arm (Fig. 6).The skin overlying the lesions showed no induration or tenderness. Both patients did not complain of pain, redness or itching on these areas.

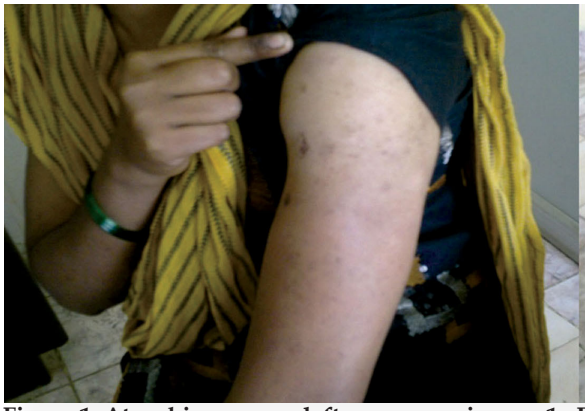

Figure 1. Atrophic areas on left upper arm in case 1

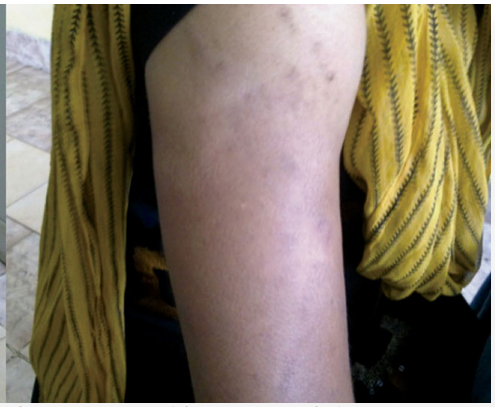

Figure 2. Atrophic areas on right upper arm in case 1

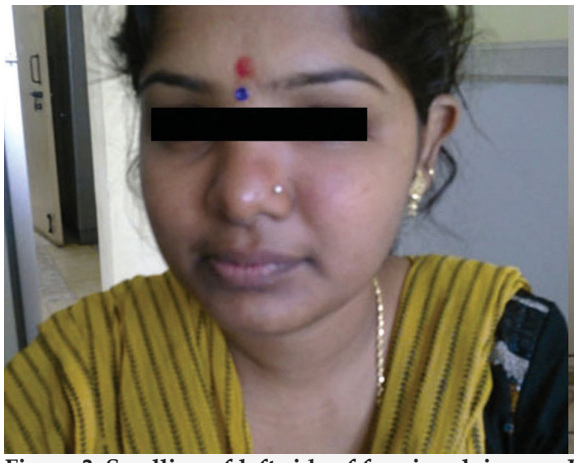

Figure 3. Swelling of left side of face involving left lower eyelid and maxillary region

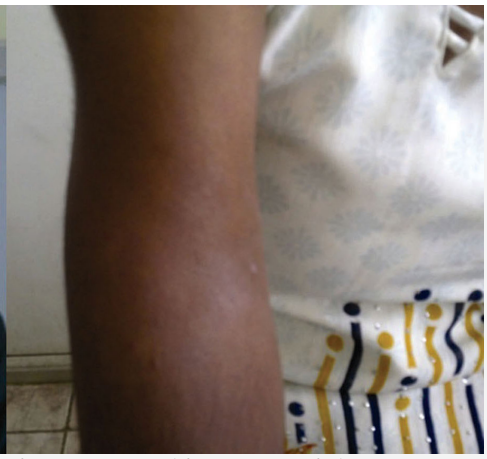

Figure 4. Atrophic areas on right upper arm in case 2

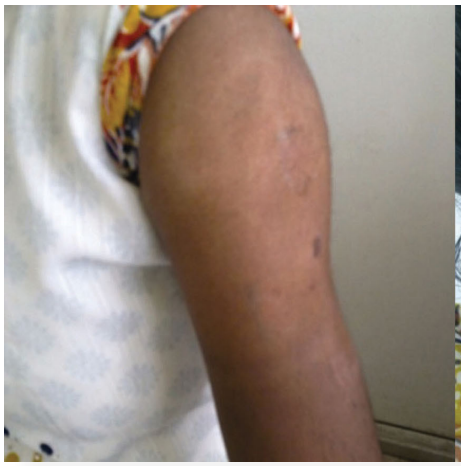

Figure 5. Atrophic areas on left upper arm in case 2

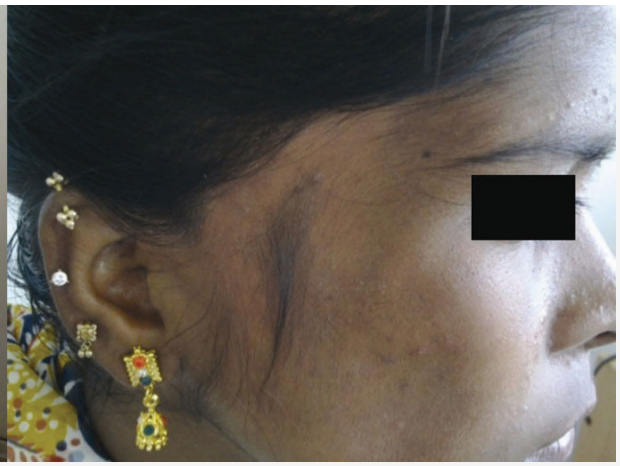

Figure 6. Atrophic areas on right side of face in case 2

There was no history of trauma or any other skin lesions on these sites prior. There was no history of any systemic illness, and similar lesions in their families. They were not under any medication. Routine blood examination including hemogram, random blood sugar, urea, creatinine, sodium, potassium and liver function tests were normal. Anti-nuclear antibodies, Anti ds-DNA and serologies for Syphilis and HIV were nonreactive. Borrelia serology was not done due to unavailability of laboratory facilities. ESR was raised in both the patients and anti-histone antibody was positive in the first patient.

Histopathological examination (Fig. 7, 8) of punch biopsies in both patients showed keratinized stratified squamous epithelium and increased bands of collagenous tissue in the papillary and reticular dermis. Subcutis revealed increased collagen bands. There was perivascular mononuclear infiltrate in the papillary and reticular dermis. A diagnosis of morphea profundus was made based on these typical histological features.

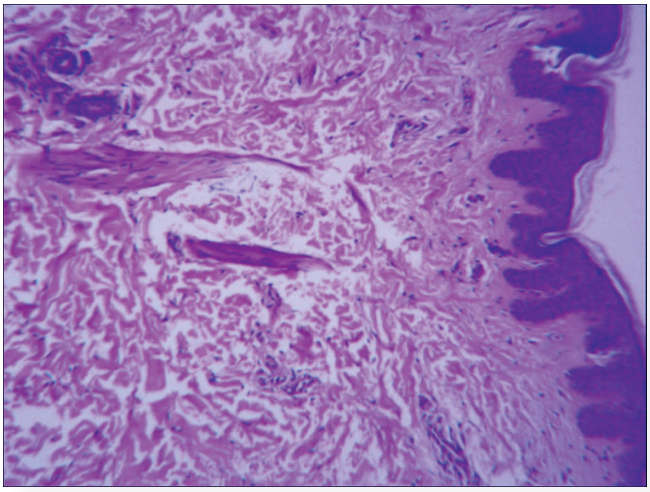

Figure 7. Histopathology from atrophic area in case 1 showing keratinized stratified squamous epithelium and increased bands of collagenous tissue in the papillary and reticular dermis. Perivascular and periadenexal mononuclear infiltrate in the papillary and reticular dermis is seen

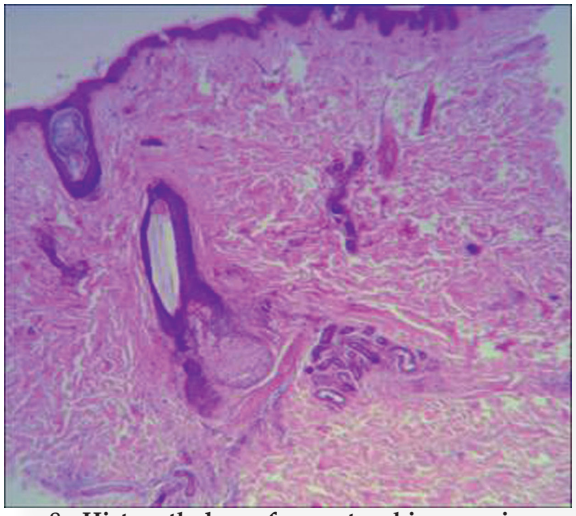

Figure 8. Histopathology from atrophic area in case 2 showing normal epithelium and increased bands of collagenous tissue in the papillary and reticular dermis. Perivascular and periadenexal mononuclear infiltrate in the papillary and reticular dermis is seen

Discussion

Scleroderma is a chronic disease of unknown aetiology in which sclerosis of the skin develops with or without involvement of internal organs. Based on the presence or absence of systemic disease, scleroderma is divided into very distinct clinical categories: systemic scleroderma or systemic sclerosis, and localized scleroderma or morphea [4]. There are five principal forms of localized scleroderma: plaques, generalized morphea, bullous morphea, linear morphea and morphea profundus [5]. Morphea profundus is a rare variant of morphea and it was first described by Whittaker et al in 1989 as a solitary fibrotic plaque. Morphea profundus usually affects middle aged people and has approximately equal sex distribution. It commonly presents with a single fibrotic plaque and is usually located over the back, shoulder and neck or paraspinal area involving the skin and deeper tissue [8]. Other rare variants of morphea profundus include multiple deep atrophic lesions of the skin without preceding inflammatory changes [7] and noninflammatory cupuliform depressed plaques at sites of previous intramuscular vaccination without significant skin induration, pigmentation or texture change [8]. Bullae arising from plaques of morphea profundus have also been reported [9].

Both of our patients were young women and presented with depressed ill-defined plaques on various sites of the body without induration of skin.

We considered a differential diagnosis of morphea profundus and lupus profundus in our first case. Skin biopsy showed features of morphea profundus. Lupus band test on direct immunofluorescence was negative. Lupus band is deposition of immunoglobulins and complement components in the skin of patients with lupus erythematosus as a linear band at the basement membrane zone [10]. Anti-nuclear and Anti ds-DNA antibodies were negative. Antihistone antibody was positive. Antihistone antibodies have been reported in 32% of patients with linear morphea and 25% with localized morphea [11] and it indicates disease activity [12]. High resolution ultrasonography and X-ray showed no involvement of muscle and bone respectively. Based on the investigative reports and typical histopathological findings a diagnosis of morphea profundus was made.

In our second patient, differential diagnosis of morphea and localized involutional lipoatrophy was made. Involutional type of lipoatrophy usually presents with a solitary lesion that exhibits decrease in size of the individual adipocytes and is separated from each other by abundant eosinophilic hyaline material, or in some instances by mucoid material [13]. The histopathology in our patient was consistent for morphea.

This is the first report on atrophic lesions in morphea profundus from India, to the best of our knowledge. Dermatologists should be aware of this rare variant of morphea profundus, and should consider this in the differential diagnosis of cases which present with atrophic skin lesions.

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Dr Sushma MD - Assistant Professor, Department of Pathology, Rajarajeswari Medical college and Hospital Kambipura, Kengeri Hobli, Mysore road, Bangalore-560074

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SYMPTOMATIC MACROGLOSSIA AND TONGUE MYOSITIS IN DERMATOMYOSITIS

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Abstract
The involvement of the tongue in dermatomyositis is rarely described in the literature. We report the case of a patient having a macroglossia whose etiologic was a dermatomyositis. The diagnosis was established by biopsy of the tongue which showed an interstitial lymphocytic infiltration associated with destruction of muscle fibers and perifascicular atrophy. The treatment was based on corticosteroids. The functional prognosis was dominated by the gene to speech and the swallowing disorders.

Key words: macroglossia; tongue myositis; dermatomyositis

Cite this article:
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Introduction

The macroglossia is observed in certain congenital muscle disease such us Becker's and Duchenne's dystrophies or Pompe's disease. It rarely occurs in polymyositis. We report the case of a symptomatic macroglossia with myositis of the tongue associated with dermatomyositis.

Case Report

A 65 years old man was admitted in our service for macroglossia and macrochely (Fig. 1, 2) associated with a swallowing disorders with false routes and frequent tongue-biting during mastication lasting for a year. He had also an incomprehensible speech and complained of muscle weakness involving the shoulder and pelvic girdles and the limbs. The patient was in a good general condition. Blood pressure was 120/70 mmHg and cardiovascular examination was normal. On physical examination, a slight decrease in muscle strength in all 4 limbs and girdles was noted. He had a significant macroglossia with a falling of the lower lip and a nasal speeech. No skin abnormalities were observed and the neurological examination was strictly normal.

The cell blood count was normal and there was no biological inflammatory syndrome. The creatinine phosphokinase (CPK) level was elevated at 606 IU/l and lactate dehydrogenase (LDH) at 416 IU/l. The hepatic and renal function was normal.

Electromyography confirmed a diffuse typical myositic process. Muscle biopsy revealed necrotic muscle fibers, regenerating fibers, an endo- and perimysial inflammatory infiltrate and a perifascicular atrophy. The different etiologies of macroglossia have been eliminated by appropriate investigations. Thyroid balance was normal and biopsy of the tongue with congo red staining showed no amyloid deposits. Enzymatic assays were not made in the absence of clinical signs towards genetic myopathies.

The biopsy of the tongue showed a phenotype T interstitial inflammatory infiltrate and perifascicular atrophy without neoplastic cells (Fig. 3).

The diagnosis of dermatomyositis sine dermatisis revealed by tongue myositis was certain according to the Bohan and Peter criteria. The search for possible neoplasia was negative (thoraco-abdominal scan and tumor markers). The diagnosis of tongue carcinoma has been ruled out by the tongue biopsy.

The patient was treated by prednisone at a dose of 1 mg/kg/day associated with methotrexate (20 mg/week) for five years with no improvement of the symptoms. Imunoglobulin infusions were administered so at a dose of 2 g/kg by cure. The patient received six cures spaced by one month. Evolution was marked by the disappearance of the muscular deficit, standardization of CPK and LDH levels, the partial reduction of macroglossia, improvement of speech and the disappearance of false routes. The current decline is of 12 months.

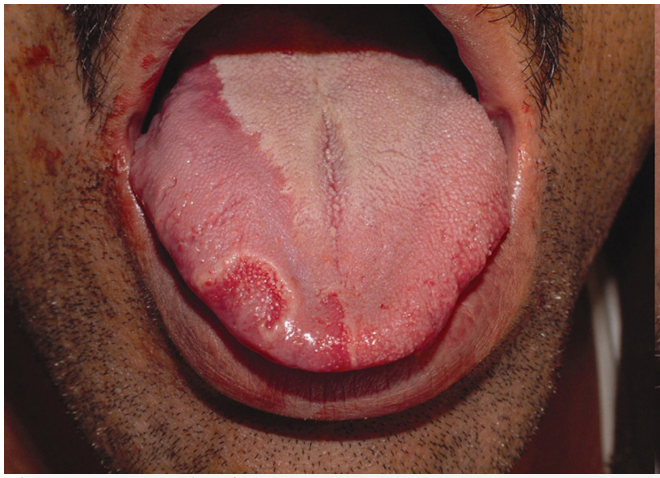

Figure 1. Macroglossia

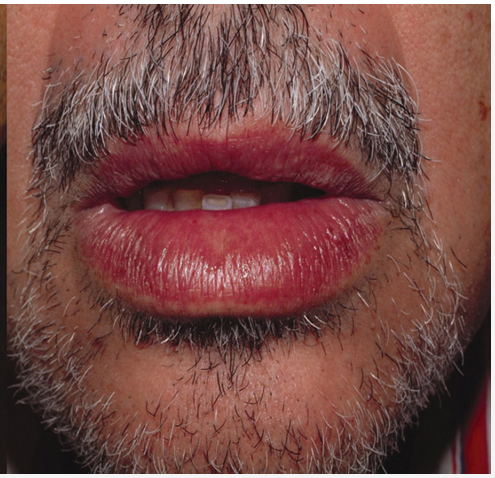

Figure 2. Machrochely

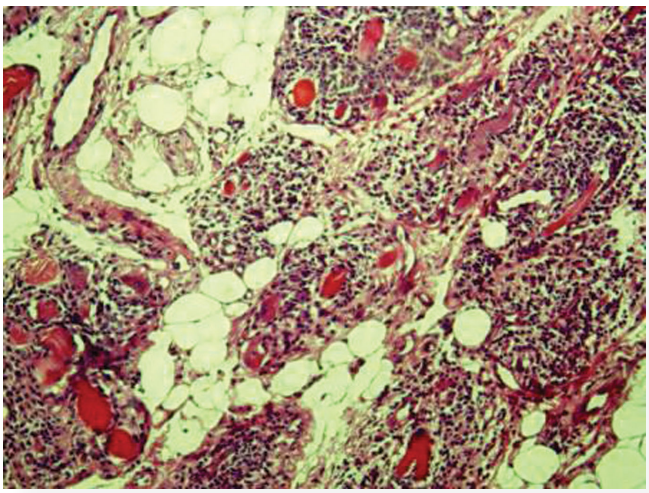

Figure 3. HEX160: The biopsy of the tongue showing a phenotype T interstitial inflammatory infiltrate and perifascicular atrophy without neoplastic cells

Discussion

Macroglossia is defined by hypertrophy or hyperplasia of the muscles of the tongue and it is due to congenital or acquired pathologies [1]. It can be observed in several pathologies such us endocrinopathies (hypothyroidism, acromegaly), granulomatoses (Sarcoidosis, crohn's disease, amyloidosis), genetic syndromes (Myopathies, Mucopolysaccharidoses, glycogen storage diseases, neurofibromatosis) and tumours particularly the tongue carcinoma. The involvement of the tongue in dematomyositis is rare [2-5].

In our patient the diagnosis of dermatomyositis sine dermatisis was certain according to the Bohan and Peter criteria [6]: girdles muscular deficit, the elevation of the CPK level, myositic process in electromyography and perifascicular atrophy in muscle biopsy. In our case the challenge was to link the macoglossia to dermatomyositis and eliminate other causes of tongue hypertrophy. The most common causes of macroglossia (amyloidosis, hypothyroidism, acromegaly) were ruled out.

Concerning genetic myopathies (Duchenne's and Becker's dystrophies, pompe's disease), our patient's age was against these diagnoses. Moreover, the physical examination did not show a waddling walk and difficulty of position change. In addition the biopsy of the tongue did not show dystrophy of the muscle fibers.

Other pathology that could simulate a myositis of the tongue and be associated with dermatomyositis is the tongue carcinoma [7-9]. Thus the biopsy of the tongue was necessary to eliminate an eventual neoplasia. In our case,the macroglossia was probably a localization of dermatomyositis because we have noted an improvement of symptoms by corticosteroid treatment associated to immunoglobulin infusions.

The magnetic resonance imaging (MRI) of the tongue could help to establish the diagnosis by showing a homogeneous tongue hypertrophy without fat deposits or oedema but the diagnosis is based on the tongue biopsy that showed an inflammatory infiltrate of the lingual parenchyma.

The treatment is based on corticosteroid therapy associated with the methotrexate and immunoglobulin infusions relayed by azathioprine in case of resistance to the initial treatment [5]. Thus, our patient demonstrates a rare case of the literature with a real tongue myositis revealing a dermatomyositis.

Indeed, a case report similar to ours was published in the literature [5]. It was a 58 years old woman. She had diabetes mellitus and hypertension treated by captopril. She consulted for diffuse myalgia and muscular weakness. The physical examination showed a deficit of pelvic girdles and limbs without skin lesions and a macroglossia. The creatinine phosphokinase (CPK) level was elevated at 2086 IU/l. Electromyography confirmed a diffuse typical myositic process. Muscle biopsy revealed necrotic muscle fibers, regenerating fibers, and an endo- and perimysial inflammatory infiltrate. The diagnosis of polymyositis was retained according to the Bohan and Peter criteria and the patient was treated only by methotrexate at the dose of 10mg/week with partial improvement. After 6 months the patient reported progressive dysarthria, frequent tongue-biting during mastication, dysphagia, and noisy breathing. The physical examination showed a majoration of macroglossia, a proximal and distal muscular deficit. The CPK level was 1642 IU/l. Captopril-induced angioedema was suspected and the captopril treatment was stopped. Blood α glucosidase activity was normal.

The MRI of the tongue showed a homogenous hypertrophy of the tongue and electromyography revealed signs of a diffuse myogenic process. Tongue biopsy was performed and showed a multifocal inflammatory infiltrate with septal and endomysial fibrosis. Congo red staining of tongue, muscle, bone, salivary gland, and skin biopsy specimens showed no evidence of amyloidosis.

Immunoglobulin infusions was administered at a dose of 2 g/kg/ cure with a total of six cures and were relayed by azathioprine.

In our patient, muscle weakness disappeared by treatment. On the other hand there was a slight improvement of macroglossia and of the speech. These results were similar to the case reported in the literature. Indeed the oropharyngeal symptoms in our patient were probably due to the hypomotility of the tongue caused by myositis and not only due to macroglossia.

A surgical reduction of the tongue had been proposed by some authors to improve the functional complications of macroglossia. The glossectomy was the most used technique and consists of making an elliptical incision in the middle of the tongue and a resection at the level of anterior corner, and then sew the sides in a straight line [10]. However the complications were frequent and included a risk of excessive bleeding, airway obstruction due to the tongue oedema, a loss of taste that might occur as a result of damage of the lingual nerve and a lesion of the salivary canal.

Conclusion

Our case illustrates a rare case of the literature of tongue myositis revealing a dermatomyositis. The interest of this observation is to raise the importance of the histological study of tongue biopsy in case of macroglossia whose cause is not obvious.

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BLASCHKOID LICHEN PLANUS IN AN ADULT KASHMIRI MALE: A RARE PRESENTATION

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Abstract
Lichen planus (LP) is common acquired dermatoses with several morphological forms. Linear lichen planus is frequently seen but cases of zonal/ zosteriform/ dermatomal/ blaschkoid LP are rare. We report a case of blaschkoid LP along with scalp LP in a 42 year old adult Kashmiri male. We report the case to add one more case to the list of this rare form of LP, with the peculiarity in our case of late onset of presentation and coexisting scalp LP, and review the literature to address to the confusion about the various related terms.

Key words: blaschkoid lichen planus; lichen planus; zosteriform lichen planus

Cite this article:
Parvaiz Anwar Rather, Iffat Hassan: Blaschkoid lichen planus in an adult Kashmiri male: a rare presentation. Our Dermatol Online. 2013; 4(2): 179-182.

Introduction

Zosteriform and blaschkoid forms of lichen planus are rare [1]. Both the forms arise either as koebner`s phenomena, wolf`s isotopic phenomena or de novo from normal skin. There is a controversy about the use of terms like zosteriform/ dermatomal LP and blaschkoid LP. We report a case of LP following blaschko lines in an adult Kashmiri male to add one more case to the list of this rare form of LP and review the literature to better understand the terms.

Case Report

A 42 year old male, from urban background, advocate by profession, presented to our out patient department on 28th June 2012, with 6 months duration of voilaceous, moderately itchy skin lesions which started on left shoulder and gradually progressed over 6 months to involve upper limb and also appeared on left trunk, relieved only partially by topical steroid application. He also gave history of 2 year duration of asymptomatic voilaceous-black eruptions on scalp leaving patches of alopecia on healing and having received topical and oral steroids for the scalp lesions. There was nothing significant in the family and drug history. There was no trauma, pain or blistering prior to or during the evolution of the skin eruption. He had no associated co-morbidity like diabetes, hypertension.

General physical examination was normal and nothing abnormal was found on systemic examination. On cutaneous examination, there were multiple unilaterally distributed violaceous, purple, flat papules and plaques of variable sizes, discrete and coalesced, in continuous and interrupted linear pattern as well as in patterns of whorls and wide bands, confined to the left side of body involving anterior and posterior-lateral aspect of arm and forearm, scapular area, anterior and posterior-lateral aspect of trunk, extending over a length of 15-20 cm, with few lesions healed with post inflammatory brownish hyper pigmentation (Fig. 1a, 1b). Some of the lesions were covered with fine adherent scaling and wickham`s striae (Fig. 2a, 2b). Scalp showed multiple violaceous plaques with scarring alopecia and covered with fine scaling, distributed symmetrically over whole scalp and with normal hair texture (Fig. 3a-3c). The oral mucosa, hair and nails were normal. Complete blood count, liver function tests, kidney function tests, urine examination, chest X-Ray, ECG and ultrasound abdomen were normal. Hepatitis B and C serology was negative. A differential diagnosis of lichen planus, lichen striatus, acquired blaschkoid dermatitis was considered and punch biopsy taken from cutaneous as well as scalp lesions.

Histopathological examination from one of the papules on skin under hematoxylin & eosin staining (H & E) showed hyperkeratosis, irregular acanthosis, basal cell degeneration in the epidermis and band like lymphocytic infiltrate in papillary dermis (Fig. 4a, 4b), and that from the scalp lesion with H & E stain showed atrophic epidermal lining with prominent basal pigmented layer, pigment incontinence and chronic inflammatory infiltrate in dermis (Fig. 5a, 5b), both suggesting lichen planus. Direct immune-fluorescence result was negative.

In view of the typical clinical features and histopathology findings, a diagnosis of LP along blaschko lines (Blaschkoid LP) was made and patient was put on oral mini-pulse of steroids with methyl-prednisolone 32mg on 2 consecutive days and also given potent topical steroid clobetasol and oral antihistamines and is doing well.

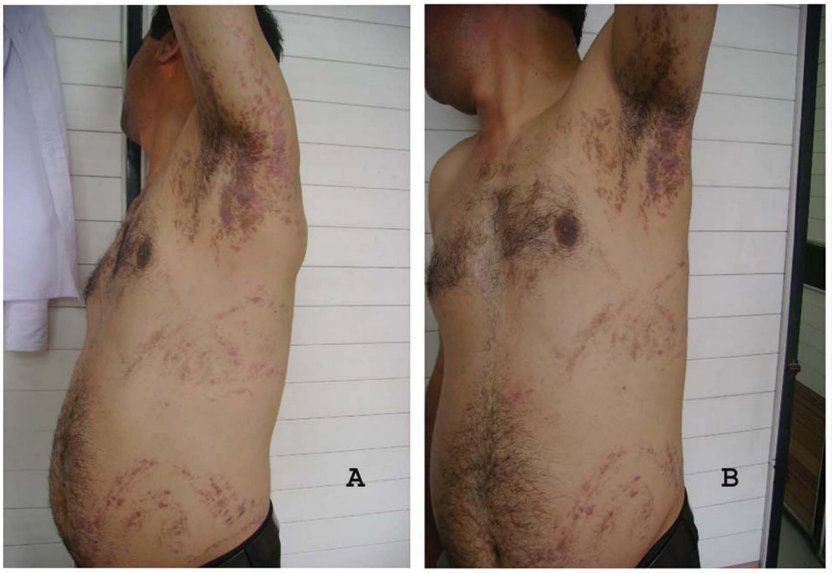

Figure 1a, b. Unilateral purple papules and plaques along blaschko lines with brown post inflammatory pigmentation

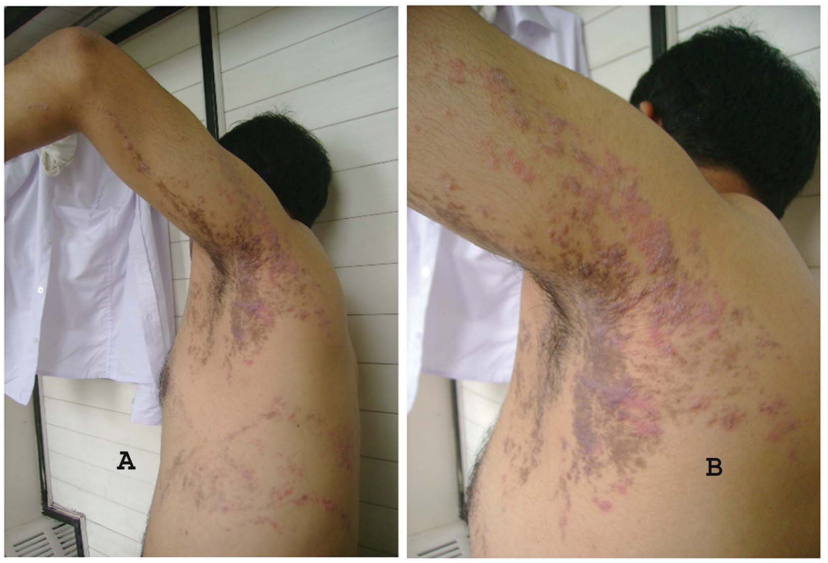

Figure 2a,b. Close up view of the lesions with wickham`s striae

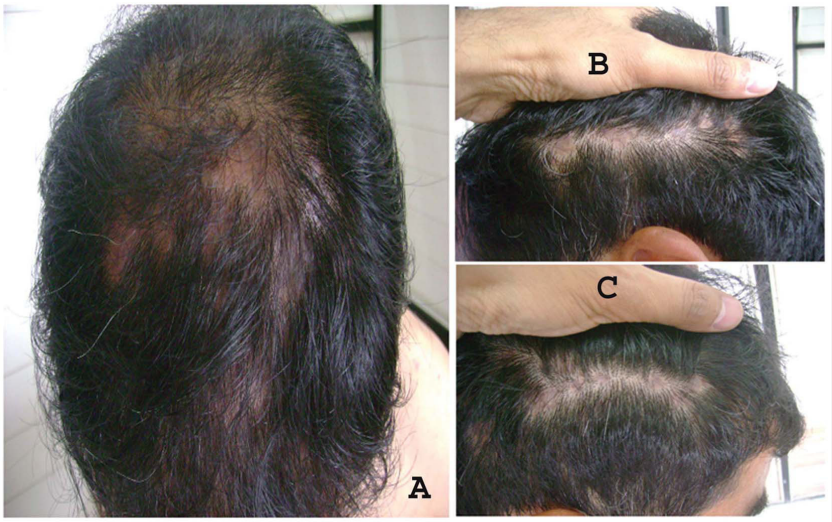

Figure 3a - c. Cicatricial alopecia with underlying brown-purple pigmentation on scalp

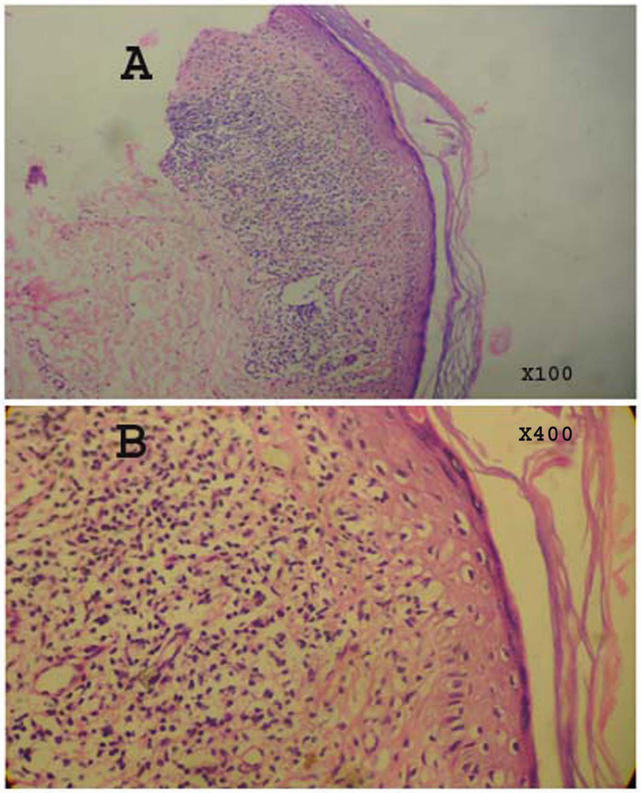

Figure 4a, b. Skin biopsy shows hyperkeratosis, irregular acanthosis, basal degeneration and band like lymphocytic infiltrate in dermis

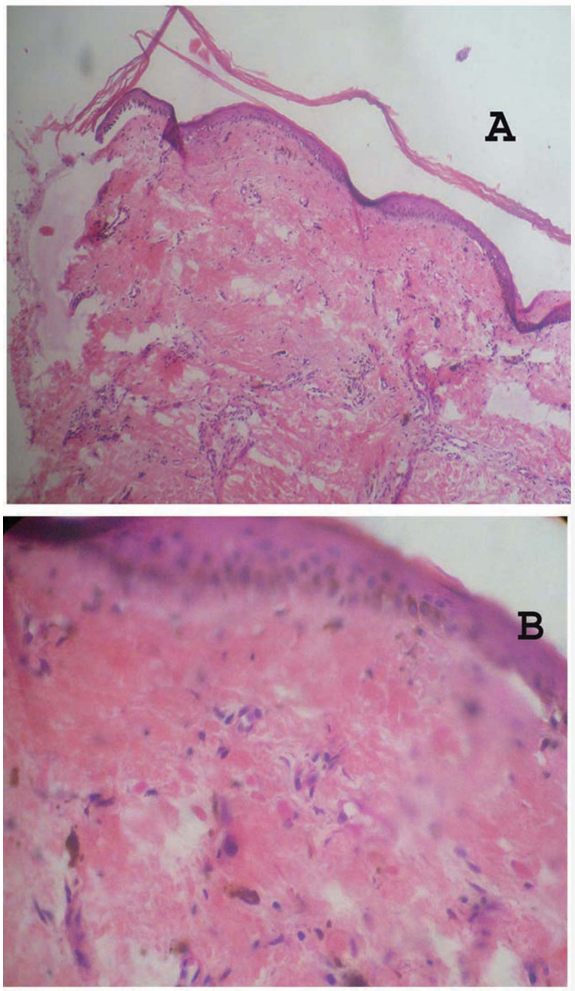

Figure 5a, b. Scalp biopsy shows atrophic epidermas, prominent basal pigment with incontinence and inflammatory infiltrate in dermis

Discussion

Linear form of lichen planus can manifest as zosteriform (dermatomal) or blaschkoid form (along blaschko lines). Dermatome is an area of skin supplied by a spinal cutaneous nerve. Blaschko lines represent a form of "mosaicism", where two or more genetically distinct cell populations are present in an individual derived from a single zygote [2,3]. The lines of blaschko were first described by a German dermatologist, Alfred Blaschko in 1901 [2]. These are distinct from the other known linear patterns of the skin [3]. Blaschko's lines do not correspond to any known nervous, vascular or lymphatic structures [2], but represent developmental growth pattern of the skin [4].

The lines of blaschko may be followed by some X-linked, congenital and inflammatory skin disorders [3]. Blaschko lines are V-shaped on the upper spine, S-shaped on the abdomen, inverted U-shaped from chest area to the upper arm, and perpendicular over the front and back of the lower limbs. They never cross the anterior truncal midline, but run along it.

Zosteriform lichen planus is a rare variant of lichen planus, which shows lichenoid papules forming a broader band that follows the dermatomes. The entity of zosteriform/ dermatomal LP is controversial. Many argue that the term zosteriform lichen planus has been applied inappropriately in cases that actually arise de novo in the lines of blaschko, rather than in true dermatomes [5,6]. Some believe that true zosteriform LP does not exist except in cases arising on the site of healed herpes zoster [7].

Blashkoid LP [8,9] accounts for less than 0.5% of patients with LP [10]. It has been reported more commonly in children than adults. It is very difficult to explain the occurrence of an acquired disorder like LP along the blaschko's lines, which are normally followed by the inherited/ genetic disorders. It seems that there exists a genetic pre-disposition to lichen planus and exposure to an appropriate enviromental or endogenous trigger may lead to the development of lichen planus.

Blashkoid LP or the controversial zosteriform/ dermatomal LP may arise as koebner phenomena because of disseminated disease [10] or at site of previous healed zoster as wolf`s isotopic phenomena [6-9,12-15] or appear de novo [15-17] on previously normal non traumatized skin [18].

With the present available literature, it is difficult to differentiate the two terms with confidence. So it remains unexplored if there are two separate forms of unilateral, de novo lichen planus; one type arising in the lines of blaschko (Blaschkoid LP) [8,9] and the other arising within one or more dermatomes (dermatomal LP). Our patient presented with unilateral linear, curled lesions of LP on normal skin along the blaschko lines, along with scalp LP and the peculiarity of adult onset of presentation, which is rare.

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ZOSTERIFORM LICHEN PLANUS: CASE REPORT OF A RARE VARIANT OF LICHEN PLANUS

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Abstract
Since its original description by Devergie in 1854, several cases of linear lichen planus have been described in the literature, but there have been notably few cases of the more rare zosteriform lichen planus. Zosteriform lichen planus needs to be differentiated from linear lichen planus and other linear dermatoses. We present a case of Zosteriform Lichen planus for its rarity and briefly review literature.

Key words: zosteriform; linear; lichen planus

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Introduction

Lichen planus (LP) has certain clinical variants which may present difficulty in diagnosis especially when the lesions happen to be arranged in a linear fashion. Linear distribution of the disease has been described in less than 1% of patients and need to be differentiated from other linear dermatoses [1]. It may be zosteriform and follows the lines of Blaschko [2,3]. We report a rare case of Zosteriform variant of Lichen planus in a young patient.

Case Report

A 12-year-old boy presented with multiple pruritic, violaceous annular skin lesions over the left side of chest. It started as small violaceous papules which coalesced to form ring lesions. There was no past history of herpes zoster or any other skin lesions. Family history was not significant. Dermatological examination revealed multiple, violaceous annular lesions in T5 dermatomal region over the chest, extending from below the left nipple to the left lateral side of chest, measuring more than 2 cms in diameter (Fig. 1). The margin of the lesion was beaded and the center atrophic. Few flat topped, violaceous, polygonal papules were present on the skin between the annular lesions. No other skin lesions were noted any where in the body.

The histopathologic examination of the lesion showed orthokeratotic hyperkeratosis, variable acanthosis, focal hypergranulosis, hydropic degeneration of basal cell layer and band like inflammatory infiltrate, suggestive of Lichen Planus (Fig. 2).

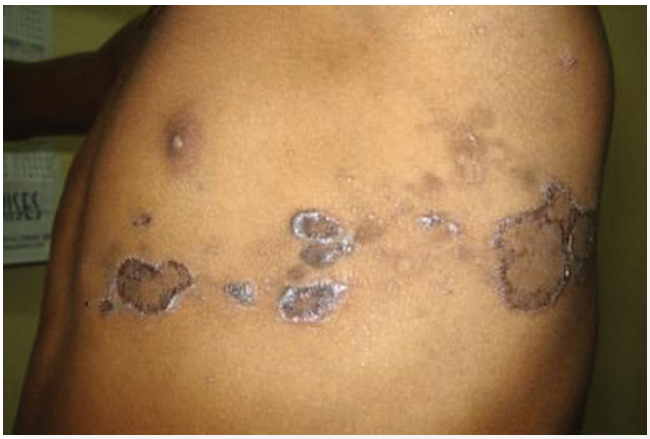

Figure 1. Multiple voilaceous papules and plaques over T5 dermatome

Disuussion

Lichen planus (LP) is an idiopathic inflammatory disease of the skin and mucous membrane. It is characterized by pruritic violaceous papules that favor the extremities.

In addition to the classical appearance, about 20 different variants are described [4]. Linear LP refers to lichen planus with a unilateral linear distribution and may occur at the site of healed zoster [5]. Although it is common in children but is also seen in adults [6]. Zosteriform pattern is a variant of LP that occurs without evidence of herpes zoster, and is extremely rare in occurrence.

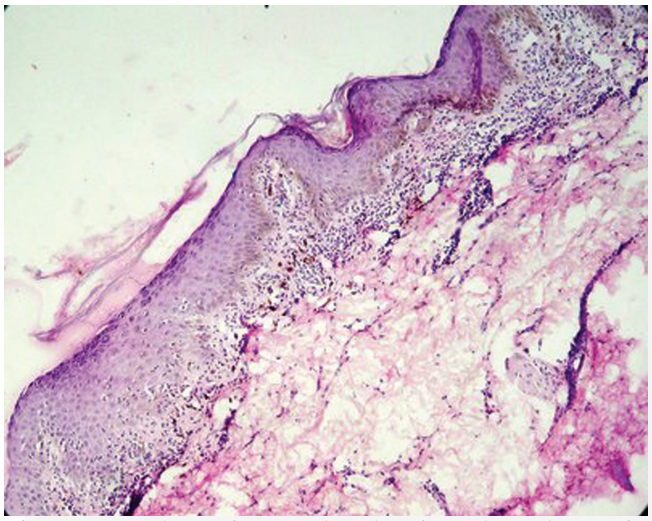

Figure 2. Photomicrograph showing hyperkeratosis, irregular acanthosis, basal cell degeneration and lichenoid dermal infiltrate, H&E, 100X

There is a definite distinction between the linear and the zosteriform type of lichen planus, to which there has not been strict adherence. In the former condition, the papular lesions appear as narrow lines about 1 or 2 cm. wide, which may follow the course of a nerve, of a vein or of a lymphatic vessel or one of Voigt's lines. In the latter the lesions form a band several centimeters wide that follows the course of a peripheral cutaneous nerve and its branches or appears over areas of radicular nerve distribution. Zosteriform LP also needs to be differentiated clinically from zona zoster and other linear dermatoses. It is an extremely pruritic variant of Lichen Planus.

The zosteriform arrangement of lichenoid papules is rare and is interpreted

as a cutaneous reaction possibly triggered by some neural factor [7].

Many disorders occur within an area that is innervated by a particular spinal cutaneous nerve. Such a distribution of lesions has provoked many authors to suggest a theory of neural origin to the linear/zosteriform lichen planus, on the other hand it has been recently suggested that most of the lesions occurring in so-called zosteriform manner do not follow a dermatomal pattern or apparently a nerve segment but are rather along the Blaschko's line [5]. Some authors believe that true zosteriform LP only exists in cases

who have developed lesions on the sites of healed herpes zoster [8].

The histology of linear LP is characteristic and enables distinction from other linear

dermatoses such as lichen striatus, linear nevi and linear psoriasis.

Treatment modalities include topical moderate to high potency corticosteroids, topical salicylic acid, and systemic sedative antihistamines. In unresponsive cases systemic corticosteroids or intralesional corticosteroids can be instituted.

Paediatric aspect of the disease: Children should be allowed to engage in full activities, and to attend school. However, because lichen planus demonstrates the isomorphic response, attempts should be made to minimize trauma to the skin. Children and parents should be informed that lichen planus can be a chronic disorder, and that numerous recurrences may occur over the next several months to years.

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POIKILODERMA OF CIVATTE

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Abstract
Poikiloderma of Civatte (Poikilodermia vascularis et pigmentosa Civatte) is a chronic skin condition which refers to the group of melanodermas. Poikiloderma of Civatte (PC) is characterized by erythema associated with atrophy and pigmentation changes of the skin usually seen on the sun exposed areas such as cheeks and sides of the neck. Chronic exposure to ultraviolet light is considered to be an important etiologic factor. We report a 54 years old female patient presenting with confluent, symmetrical reticular dark-brown patches on her face, predominantly on the cheeks, in the preauricular zone, on the forehead and on the front and lateral parts of the neck. Characteristic of the patches are mottled hyper- and hypopigmentation with numerous telangiectasias and areas of atrophy. A biopsy specimen shows epidermal changes including atrophy of the epidermis and hyperkeratosis. Necrotic keratinocytes are found in the papillary dermis as well as occasional lymphocytes extending into the basal layer. The basal damage is associated with dermal involvement. The dermal infiltrate is predominantly lymphocytic. Multiple melanophages, free pigment and edema are seen in the dermis.

Key words: poikoloderma; sun exposure; histology; treatment

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Uladzimir P. Adaskevich, Maryia A. Katina, Valeryia A. Miadzelets: Poikiloderma of Civatte. Our Dermatol Online. 2013; 4(2): 185-187.

Introduction

Poikiloderma of Civatte (Poikilodermia vascularis et pigmentosa Civatte) is a chronic skin condition which refers to the group of melanodermas. The term „poikiloderma" means a skin change with atrophy (thinning), pigmentary changes (either hyperpigmentation or hypopigmentation) and telangiectasia formation (dilatation of fine blood vessels) [1-3].

Poikiloderma of Civatte (PC) is characterized by erythema associated with atrophy and pigmentation changes of the skin usually seen on the sun exposed areas such as cheeks and sides of the neck. The condition was first described by a French dermatologist Civatte in 1923 [1,2].

PC occurs in females more frequently than in males. Female individuals are most commonly affected in the menopausal period [3]. The incidence of PC is unknown; many patients may have a mild form of the disease and may not ask for medical attention. Fair-skinned people are more prone to the disease, although it may be seen in all skin types [1,3].

Some reports point out to clinical and pathomorophological similarity of poikiloderma of Civatte and Riehl's melanosis. But in the latter case the skin atrophy is less intensive and teleangiectasias are not typical [1,2].

Chronic exposure to ultraviolet light is considered to be an important etiologic factor which is confirmed by the fact that lesions occur on sun-exposed areas. In addition, solar elastosis is a frequent histopathologic finding. Photosensitizing chemicals in perfumes or cosmetics have been implicated in the pathogenesis of poikiloderma of Civatte. Hormonal changes related to menopause or low estrogen levels may also be seen as a possible causative factor. There are some reports suggesting genetic background. The genetically determined predisposition may be expressed in an increased susceptibility of the skin to ultraviolet radiation [1-4]. UV-induced changes of the dermal connective tissue are the predominant histological feature of PC, leading to telangiectasia due to the loss of vascular support. Reticular pigmentation may result from a delayed hypersensitivity reaction to perfume or cosmetic ingredients.

The lesions are usually asymptomatic, but some patients may feel mild burning, itching and increased sensitivity in the affected area. The main clinical sign of the disease is a formation of symmetrical reddish-brown patches on the face, lateral parts of the cheeks and sides of the neck, less commonly in the center of the chest. Poikiloderma of Civatte characteristically spares the shaded area under the chin. Reticulate pigmentation with atrophy and telangiectasia is usually present [1-3].

The first and most important step in the management of PC is to avoid sun exposure. Avoiding perfumes and using proper photoprotection are advocated. It is recommended to use a non-irritating sunscreen with SPF at least 50+.

The treatment of PC must be directed to the simultaneous elimination of both components, the vascular and the pigmented ones [2,9,10]. Recently, Pulsed Dye Lasers and Intense Pulse Light therapy have been used with favourable results. Intense Pulsed Light systems have a wavelength spectrum of 515-1200nm with high-intensity light sources that emit polychromatic, noncoherent light and, thus, are different from lasers. Several treatment courses may be required for complete clearing [3-8]. Use of fractional photothermolysis (laser technology that creates microthermal injury zones in skin) to treat poikiloderma of Civatte has also been described, with promising results [10]. Attempts to correct the disorder using electrosurgery, cryotherapy, and argon laser have been unsuccessful [3,8].

Topical treatment. Mild topical steroid creams, e.g. hydrocortisone valerate, should be applied for 2-4 months twice a day. Topical retinoids, such as cream Retin A, could be beneficial if used for about a year. Hydroquinone-containing preparations may help fade the pigmentation Pigment-lightening products, such as topical tretinoin, glycolic acid and hydroquinone, are most often recommended for combined usage.

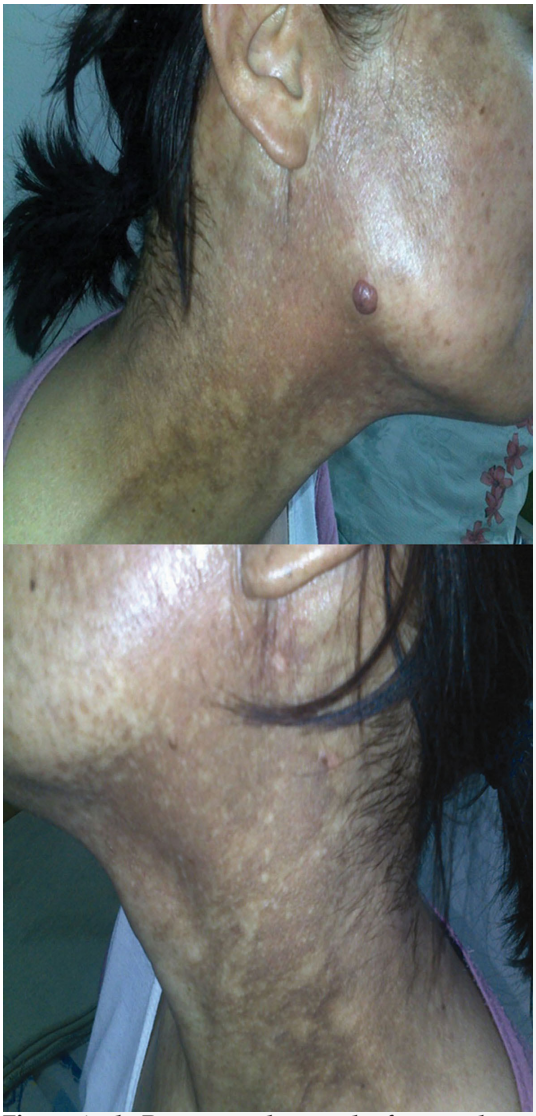

Figure 1a, b. Brown patches on the face, neck

Case Report

We report a 54 years old female patient, admitted to our clinic with the suspected diagnosis «Discoid lupus erythematosus». She had complaints of the skin affection on the face and neck accompanied by a slight tightening of the skin and a tingling sensation.

The first changes of the skin appeared 8 month ago in summer time. The patient noticed deterioration of the skin appearance after sun exposure. After applying of hydrocortisone ointment a slight improvement was seen. The patient had been suffering from euthyroid nodular struma, hysteromyoma for 7 years.

Clinical picture. Confluent, symmetrical reticular dark-brown patches were present on the face, predominantly on the cheeks, in the preauricular zone, on the forehead, on the front and on the lateral parts of the neck. Patches showed mottled hyper- and hypopigmentation with numerous telangiectasias with areas of atrophy (Fig. 1 a, b).

A biopsy specimen shows epidermal changes including atrophy of the epidermis and hyperkeratosis. Necrotic keratinocytes are found in the papillary dermis as well as occasional lymphocytes extending into the basal layer. The basal damage is associated with dermal involvement. The dermal infiltrate is predominantly lymphocytic. Multiple melanophages, free pigment and edema are found in the dermis (Fig. 2).

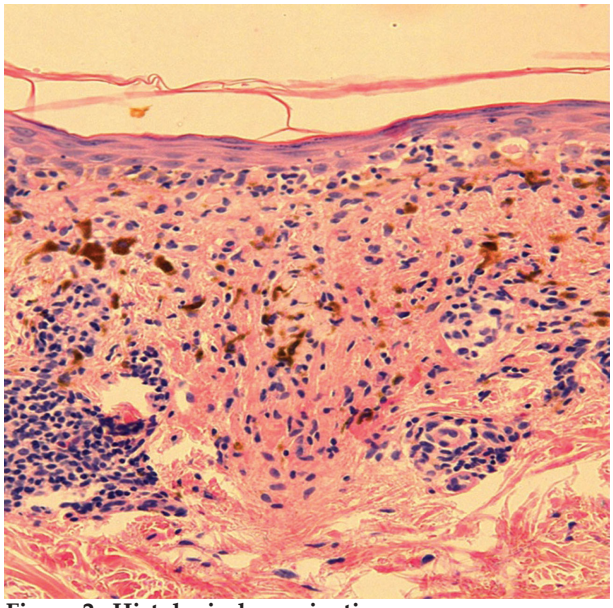

Figure 2. Histological examination

Discussion

Poikiloderma of Civatte (PC) although rather common in countries with lighter skin population and a lot of sunshine (e.g. Greece) is a rare condition in moderate climates of East-European countries like Belarus. Hence, the clinical picture of pigment changes suggestive of PC should be differentiated from other poikiloderma conditions in large-plaque parapsoriasis, lupus erythematosus, dermatomyositis, chronic radiation dermatitis, dyskeratosis congenital and rare syndromes, e.g. Bloom syndrome and Rothmund-Thomson syndrome. PC is classified into erythemato-telangiectatic, pigmented and mixed type depending on predominating clinical feature [3]. In our case it was a mixed type with mottled pigmentary changes and a lot of telangiectasias sparing the submental region and the anterior neck which is highly characteristic of PC. The perimenopausal age of the patient was also typical for this condition.

Treatment of PC remains challenging [4-10]. Ideally it should combine the elimination of vascular and pigment components simultaneously [3]. Various treatment modalities include lasers, depigmenting agents, topical retinoids and chemical peelings [1,3]. Laser therapy is considered to be the most succsessful, though costly and not consistntly effective [3]. Since treatment of PC remains a challenge all means of photoprotection are fundamentally important. Our patient was recommended to use sunscreen with SPF at least 50+ from March till October; to apply pigment-lightening creams: hydroquinone + tretinoin + hydrocortisone from October till March an to visit a gynecologist and an endocrinologist for corresponding examination.

Recommendations to the patient:

- To use sunscreen with SPF at least 50+ from March till October;
- To apply pigment-lightening creams: hydroquinone + tretinoin + hydrocortisone from October till March
- Examination by a gynecologist and an endocrinologist

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POIKILODERMA OF CIVATTE

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This clinical entity is not uncommon in Romania, especially in spring and summer,(but also during winter, for whose who are very fond of sky), most frequently diagnosed in fair-skin women over 40 years old, on a genetic background, after sun exposure (varying from minutes to hours), with a great cosmetic and physiological impact on social life. It is still a problem for dermatologists, not mainly for diagnosis but for therapy.

It is an easy clinical diagnosis, although many other dyspigmentations must be ruled out (see classification).

Classification of facial and neck melanosis [1]:

* Melasma (chloasma)
* Erythema dyschromicum perstans (Ashy dermatosis of Ramirez, erythema chronicum figuratum melanodermicum)
* Lichen planus pigmentosus
* Riehl's melanosis (Pigmented cosmetic/ contact dermatitis)
* Erythromelanosis peribuccale pigmentaire of Brocq
* Poikilodermia Civatte
* Erythromelanosis follicularis of face and neck
* Nevus of Ota
* Miscellaneous causes: Periorbital Melanosis
  Addison's disease
  Exogenous ochronosis
  Post chikungunya pigmentation (reported in India, induced by alpha virus transmitted by Aedes aegypti and Aedes albopictus)
  Acanthosis nigricans

In daily practice a skin biopsy is vary rare necessary for diagnosis, a through endocrinological examination is mandatory, sometimes an allergology test or a very attentive anamnesis are helpful (cosmetics involved).

The mainstay of the treatment is photoprotection, which must be very clearly explained to the patients, the rules regarding the use of sunscreens have to be followed carefully and even so the avoidance of pick hours on sunlight (10 am-4pm) must be strictly accepted.

There are no guidelines for the management of Poikilodermia Civatte so, different, and sometimes combined therapies, have been applied with success in treating this condition, aspect very well presented in the article.

We prefer Hydroquinone and chemical peels (Azelaic acid and Kojic acid) along with laser therapy.

We congratulate the authors for presenting the case, which turned to be an open door to an up-to date overview on Poikilodermia Civatte.

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NASZA DERMATOLOGIA Online
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DOES PITYRIASIS ROSEA KOEBNERISE?

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Abstract
The Koebner phenomenon or isomorphic phenomenon is described in dermatology texts as the production of lesions of the original disease, in clinically uninvolved skin, following trauma. The lesions are located at the site of trauma and evidence of a traumatic causation is the linear arrangement of some of the lesions, such as in the case of lichen planus. Other disorders known to exhibit the Koebner phenomenon include psoriasis and vitiligo. A number of other diseases are associated with the Koebner phenomenon. Pathergy is a phenomenon of pustule production following trauma, which occurs in certain disorders such as Pyoderma gangrenosum and Behçet's disease. In some disorders such as impetigo and verruca vulgaris, inoculation may give the appearance of the Koebner phenomenon.
A case of pityriasis rosea and Koebner phenomenon at the site of routine blood assay is described in this work. This author has not thus far encountered any description of the Koebner reaction in relation to pityriasis rosea in the literature, but, perhaps, with this report, other physicians will be more open to this possibility and actually uncover similar cases.

Key words: pityriasis rosea; Koebner phenomenon; skin disease

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Introduction

The Koebner phenomenon was first described in 1872 by the renowned German dermatologist, Heinrich Koebner [1]. He described it in cases of psoriasis that he had studied and considered it an irritant effect. The Koebner phenomenon or reaction has since been consistently described with vitiligo and lichen planus. It has also be cited in association with a number of other diseases such as lichen sclerosus, sarcoidosis and pityriasis rubra pilaris [1,2]. There appear to be no reports in the literature of this phenomenon being observed in patients with pityriasis rosea.

Case Report (Fig. 1-3)

A 35 year old female presented at our outpatient clinic with an eruption that had developed 2-3 weeks before presentation. It had been preceded by a single lesion on her right flank. The rash was asymptomatic, except for a mild sensation of itch when she thought of the lesions. It had spread gradually to involve her trunk. The patient is a pharmacy assistant by profession.
Her past medical history was negative for medication and for a preceding febrile illness. She had had an appendicectomy at 5years of age and a benign breast nodule at 26 years of age. She had no history of contraceptive use.
On examination, a rash comprised of small, well-defined, erythematous, slightly squamous plaques was seen on the anterior and posterior trunk. Similar lesions were noted (one each) in the cubital fossae. The limbs as well as palms and soles were uninvolved. No regional adenopathy was noted. There were no significant findings on systemic examination.
Upon further questioning, she claimed these cubital lesions appeared about 1 week after needling for routine, work-related, blood testing. The results were normal and she tested VDRL negative.
Since the clinical diagnosis was obvious and the patient also unwilling to consent, no biopsy was taken.
She was placed on observation and asked to return 3 weeks later. At this follow-up visit, the lesions were almost completely absent. She remained asymptomatic and was therefore discharged.

Discussion

Pityriasis rosea is a common, benign, self-limiting dermatosis that affects the trunk and proximal extremities [3]. No treatment is usually required. A viral aetiology has been suggested. The differential diagnosis includes secondary syphilis (herald patch present in our patient), tinea corporis (this patient had squames on the inside of the lesion margin), numular dermatitis (patient's rashes were not round, no vesicles were present), guttate psoriasis (squames were fine rather than coarse in this patient) and pityriasis lichenoides chronica (lesions were not predominantly on extremities in this patient). The mechanism of the production of the Koebner or isomorphic phenomenon is unknown.

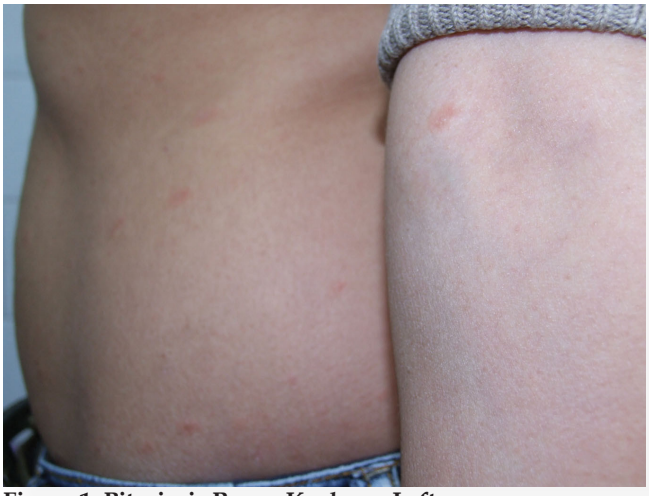

Figure 1. Pityriasis Rosea-Koebner. Left

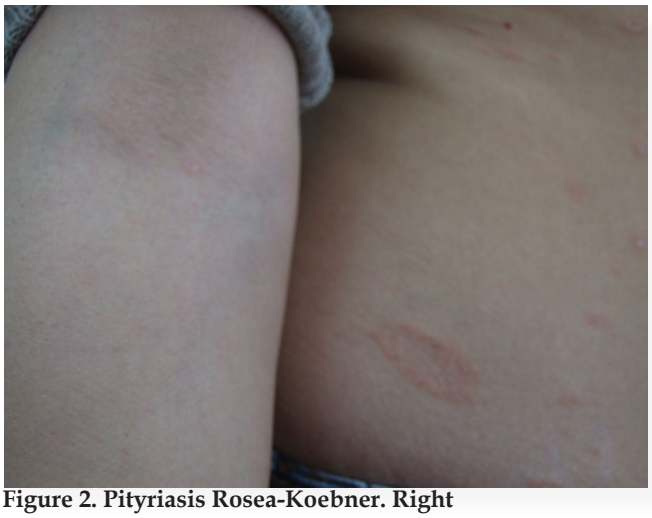

Figure 2. Pityriasis Rosea-Koebner. Right

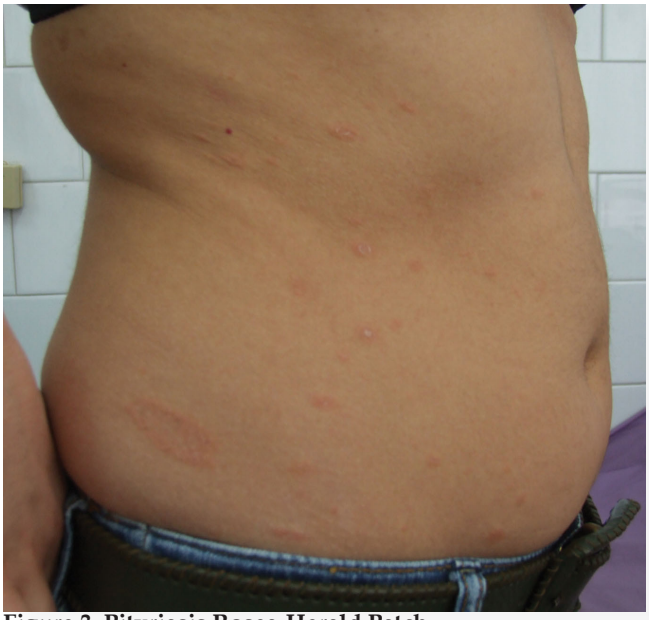

Figure 3. Pityriasis Rosea-Herald Patch

It has been induced most frequently in patients with psoriasis [2], in hopes of better understanding the pathogenesis of this disorder. Indeed, Heinrich Köbner's original experiments were on patients with psoriasis [1]. Time lag for koebnerisation in psoriasis was found to be about 10-20 days, but, in general, for the Koebner phenomenon, it was estimated to vary from 3 days to years [2]. The patient under discussion had a time lag of about 1 week from trauma to onset of koebnerisation.

In psoriasis, sensory neuropeptides may contribute to the development of koebnerisation, thus indicating a neural theory for the causation of this phenomenon [4]. Furthermore upregulation of, as well as increase in, Nerve Growth Factor (NGF) has been noted in psoriatic plaques by some authors [5]. This could, nonetheless, be a conclusion that may not be subject to generalisation, as, in contrast to psoriasis, vitiligo appears to show a higher frequency of the Koebner phenomenon in nonsegmental (47.19%), vs. segmental vitiligo (24.00%), which is hypothesised by some as having a neural aetiology [6]. Pharmacologic inhibition of epidermal growth factor receptor (EGFR) by EGFR inhibitors leading to enhanced and protracted inflammation in the skin is also a putative mechanism of production of the Koebner effect [7].

No mechanism is suggested here for the reaction observed in this patient.

Reverse koebnerisation may also follow injury, although the mechanisms of production may differ [2].

The isotopic phenomenon is the occurrence of a new lesion in the area of skin where another unrelated dermatosis has healed [8]. It is fundamentally different from the isomorphic phenomenon. This is also known as the Wolf's isotopic response [9], although it may have been described 30 years earlier by other authors [8], as Wolf himself acknowledged [9]. Aside from psoriasis, lichen planus and vitiligo, the Koebner phenomenon has been noted in Kaposi sarcoma, Kyrle disease, Darier disease and lichen sclerosis et atrophicus [10].

A case of pityriasis rosea presenting with posttraumatic lesions that suggest the Koebner phenomenon is presented. Although koebnerisation has been associated with a wide variety of disorders [1,2], to the best of our knowledge, it has not been reported in association with pityriasis rosea.

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DOES PITYRIASIS ROSEA KOEBNERISE?

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comment:

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We read with admiration the article by Nwabudike on possible association of pityriasis rosea (PR) and Köebner/ isomorphic response (IR) [1]. A 35-year-old female exhibited PR lesions at bilateral cubital fossae, presumably related to venepuncture. The author believes that there exists no previous report on such association.

We note a typical PR lesion over a superficial vein at the left cubital fossae (Figure 1, [1]). However, the PR lesion at the right cubital fossa was not directly riding on or adjacent to the superficial veins (Figure 2, [1]). A venepuncturer viewing an obvious superficial vein over the radial aspect of the fossa and a Y-shaped thick superficial vein over the ulnar aspect would unlikely puncture at the PR lesion site shown which was near the centreline of the fossa. Should this be the case, IR can be established on one site only – the left arm, which might be coincidental. We therefore cast doubts on whether this patient is genuinely exhibiting IR at the backdrop of venepuncture. However, there exists a possibility that the venepuncturer might have selected a deep vein which was not captured in Figure 2 [1], and we therefore trust the judgements of the author.

We also wish to point out that IR was suspected to occur in a 41-year-old male patient with both PR and secondary syphilis [2]. (However, such might be termed isotopic response in the modern terminology [3,4], as rightly pointed out by the author). IR has also been reported in a patient with inverse-PR [5]. The author may have just inadvertently omitted these reports in his literature search.

Four types of IR have been described – true IR (frequent development of lesions in traumatised and previously uninvolved skin, e.g. psoriasis), pseudo IR (auto-inoculation of microbes, e.g. viral warts and molluscum contagiosum), occasionally-occurring IR (infrequent development, e.g. Darier's disease and erythema multiforme), and questionable IR (limited case reports, e.g. morphoea) [6]. At the current state of knowledge, we believe that PR might be associated with questionable IR only. Should there be further reports, such association might be occasional IR if the pathogen is yet unidentified, or pseudo IR if we finally identify the true culprit(s) in PR, with the latter scenario being highly unlikely at this moment, considering uncertain microbiological aetiology of PR [7].

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NASZA DERMATOLOGIA Online
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IMMUNE RESPONSE IN A CUTANEOUS ALLERGIC DRUG REACTION SECONDARY TO IMIDAPRIL, BENAZAPRIL AND METFORMIN

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Abstract
Introduction: Cutaneous drug reactions may be classified with regard to pathogenesis and clinical morphology. They may be mediated by both immunologic and non-immunologic mechanisms.
Case report: A 56 year old female presented with widespread patches and macules, concentrated on her face, trunk and extremities. The lesions were pruritic, and temporally associated with intake of benzapril hydrochloride, imidapril and metformin.
Methods: Biopsies for hematoxylin and eosin (H&E) examination, as well as for immunohistochemistry (IHC) and direct immunofluorescence (DIF) analysis were performed for diagnostic purposes, and also to evaluate the lesional immune response.
Results: Hematoxylin and eosin staining demonstrated a histologically unremarkable epidermis. Within the dermis, a moderately florid, superficial and deep, perivascular infiltrate of lymphocytes, plasmacytoid lymphocytes, histiocytes and rare eosinophils was identified, consistent with an allergic drug reaction. DIF demonstrated deposits of IgE, Complement/C3 and fibrinogen around dermal blood vessels. IHC demonstrated positive staining with HAM-56 and myeloid/histoid antigen in the cell infiltrate around the upper dermal blood vessels. HLA-ABC was overexpressed around those vessels, as well as around dermal sweat glands. COX-2 demonstrated positive staining in both the epidermis and upper dermis.
Conclusion: Drug reactions are significant causes of skin rashes. In the current case, we were able to identify multiple antigen presenting cells in the area of the main inflammatory process. The immunologic case findings suggest that allergic drug eruptions may represent complex processes. An allergic drug reaction should be suspected whenever dermal, perivascular deposits of fibrinogen, Complement/C3 and other markers such as IgE are identified via DIF.

Key words: HLA-ABC; biomarkers; myeloid-histoid antigen; HAM-56; GFAP; COX-2

Abbreviations and acronyms: Immunohistochemistry (IHC), hematoxylin and eosin (H&E), direct immunofluoresence (DIF)

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Introduction

Cutaneous drug reactions may be classified with respect to pathogenesis and clinical morphology [1-6]. They may be mediated by immunologic and non-immunologic mechanisms. Immunologic reactions involve the host immune response and may be mediated by IgE-dependent, immune complex-initiated, cytotoxic, or cellular immune mechanisms [1,2]. Non-immunologic reactions may occur via activation of effector pathways, overdoses, cumulative toxicity, side effects, interactions with other drugs, metabolic alterations or exacerbation of pre-existing dermatologic conditions [1-6]. Certain cutaneous clinical lesional patterns may be associated with cutaneous drug reactions. These include urticaria, photosensitive eruptions, erythema multiforme, disturbance of pigmentation, morbilliform reactions, fixed drug reactions, erythema nodosum, toxic epidermal necrolysis, vasculitides and bullous reactions [1-7]. In addition, certain drugs cause defined cutaneous syndromes. These include iodides and bromides, hydantoins, corticosteroids, antimalarial agents, gold, cancer chemotherapeutic agents, tetracyclines, thiazides and sulfonamides, nonsteroidal anti-inflammatory agents and coumarin.

Case Report

A 56 year old Caucasian female was evaluated after presenting suddenly with erythematous macules and patches following a one week regimen of imidapril, 5mg once a day with concurrent benzapril and metformin. These medications were prescribed for treatment of her hypertension and Type II diabetes mellitus.

Methods

Our histopathologic studies and hematoxylin and eosin staining were performed as previously described [3-8].

Direct immunofluorescence (DIF) and immunohistochemistry (IHC):

In brief, DIF skin cryosections were prepared, and incubated with multiple fluorochromes as previously reported [3-8]. We utilized a normal skin negative control, obtained from patients undergoing aesthetic plastic surgery. To test the local immune response in lesional skin, we utilized the following markers: antibodies to immunoglobulins A, E, G, and M; Complement/C1q and C3; kappa and lambda light chains, and albumin and fibrinogen. All of these antibodies were either fluorescein isothiocyantate (FITC) or Texas red conjugated for the DIF testing, and obtained from Dako (Carpinteria, California, USA). We also utilized Cy3 conjugated monoclonal anti-glial fibrillary acidic protein (GFAP) antibody from Sigma (Saint Louis, Missouri, USA). We studied the following markers via immunohistochemistry: Complement/C5b-9/MAC, HLA-ABC, monoclonal mouse anti-human myeloid/histiocyte antigen and COX-2, all also from Dako; and HAM-56 antibody from Cell Marque Corporation (Rocklin, California, USA). Our IHC studies were performed as previously described [3-8]. The IRB consent was obtained.

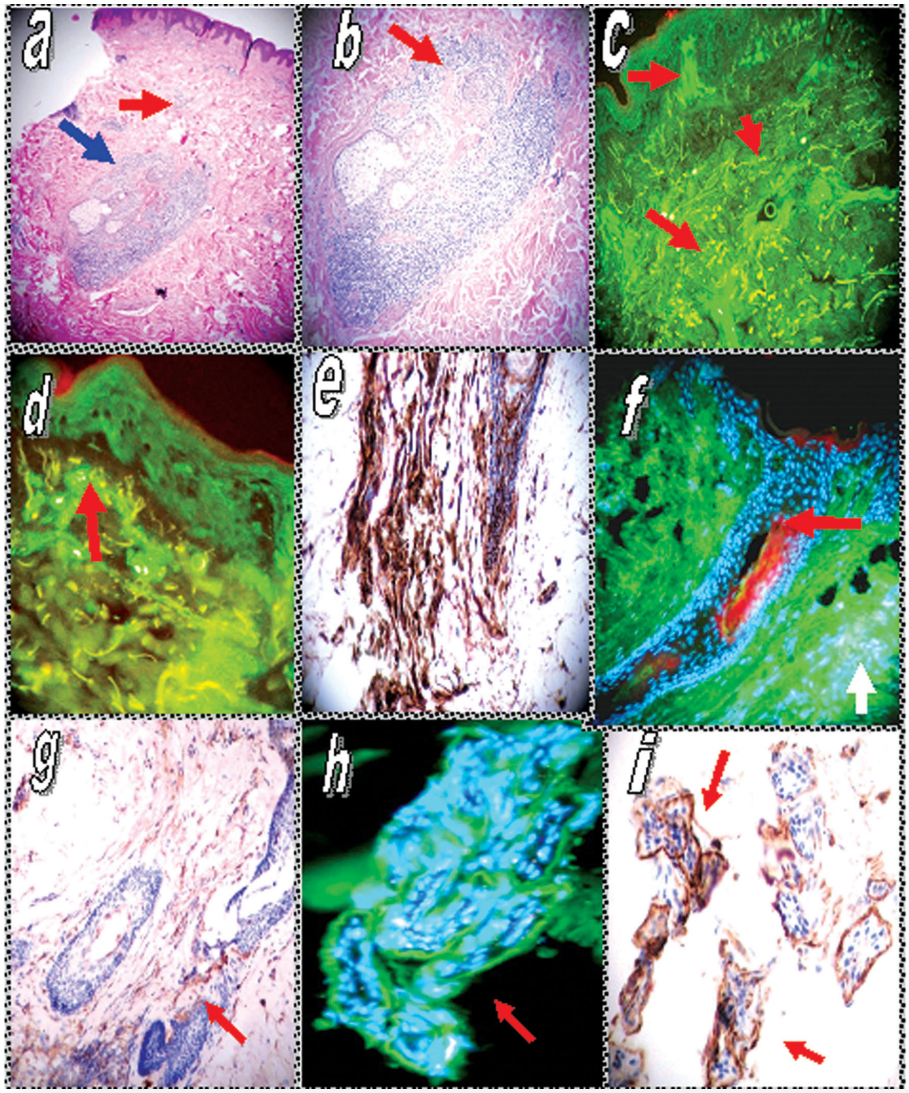

Figure 1. a and **b.** H&E staining at 100X and 200X respectively, showing a mixed inflammatory infiltrate along the upper and intermediate neurovascular plexuses of the skin(red arrows), as well as a strong infiltrate of lymphocytes, plasmacytoid lymphocytes, histiocytes and eosinophils near dermal hair follicles and sebaceous glands (blue arrow). **c and d.** At 100X and 400X magnification, respectively. DIF demonstrating positive staining with FITC conjugated IgE in the upper and intermediate neurovascular plexuses of the dermis (green staining; red arrows). **e.** HLA-ABC positive IHC staining around the hair follicular unit and blood vessels around this structure. **f.** DIF, demonstrating anti-human FITC conjugated kappa light chain antibody with positive staining around hair follicle areas (green staining; white arrow). Also note positive Texas red conjugated Complement/C3 staining inside the hair follicle (red staining; red arrow). **g.** Complement C5b-9/MAC positive IHC staining around the hair follicles and sweat glands(brown staining; red arrow). **h.** FITC conjugated Complement/C5b-9/MAC positive DIF staining around the hair follicles and eccrine glands(green staining; red arrow) **i.** IHC, demonstrating anti-human kappa light chain positive staining around dermal sweat glands(green staining; red arrows).

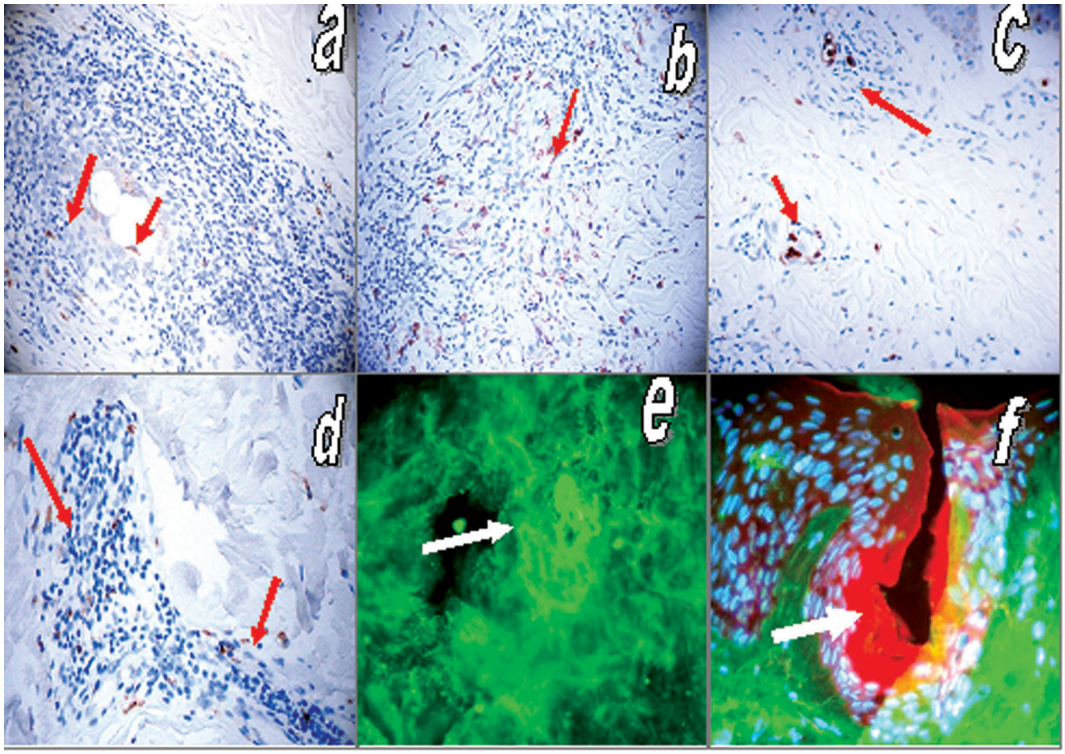

Figure 2. a and b. Positive IHC staining in the cell infiltrate with myeloid/histiocyte antigen (brown staining; red arrows). **c and d.** Positive IHC staining with HAM-56 (brown staining; red arrows). **e**. Positive DIF staining with FITC conjugated Complement/C3 to dermal blood vessels (green staining; white arrow). **f.** Positive Texas red conjugated Complement/C3 DIF staining in the isthmus of the hair follicle (red staining; white arrow).

Results

Examination of the H&E tissue sections demonstrated a histologically unremarkable epidermis. Within the dermis, a moderately florid, superficial and deep, perivascular infiltrate of lymphocytes, plasmacytoid lymphocytes and histiocytes was identified. Neutrophils and eosinophils were rare. Mild, deep dermal eccrine gland inflammation was also noted (Fig. 1). No dermal mucin deposition was seen. On DIF review, FITC conjugated anti-human IgE and Complement/C3 antibodies were positive around dermal blood vessels, especially those within the upper and intermediate dermal plexuses. Anti-human kappa light chain, Complement/C3, C1q and fibrinogen FITC conjugated antibody staining was positive within dermal eccrine sweat glands (Fig. 1, 2). On IHC review, the Complement/C5b-9/MAC complex antibody stained positive around dermal blood vessels, hair follicles and eccrine sweat glands (Fig. 1, 2). IHC also demonstrated positive staining via HAM-56 and myeloid/histoid antibodies in the cell infiltrate around the upper dermal blood vessels. HLA-ABC was overexpressed around those vessels, as well as around dermal sweat glands. COX-2 was positive in both the epidermis and upper dermis.

Discussion

Allergic reactions include 1) mild clinical events such as pruritus; 2) moderate events, including generalized skin eruptions and gastrointestinal and respiratory symptoms, and 3) severe reactions such as anaphylaxis with cardiovascular complications; these reactions represent common clinical challenges [1-6]. Allergic reactions may develop to inhaled substances, food and food additives, and foreign substances (blood, latex, etc.). Many medications are documented causes of anaphylactic reactions, asthma, and generalized urticaria or angioedema [1-6]. Moreover, multiple skin reactions are induced by drugs via immune complexes, complement mediated reactions, direct histamine liberation and modulators of arachidonic acid metabolism. Notably, we found strong expression of COX-2 in both the epidermis and upper dermis. Finally, insect venom allergies may manifest with pain, disseminated exanthems and angioedema [1-9].

The discovery of new associations between drug toxicities and specific HLA alleles has been facilitated by the use of DNA-based molecular techniques and the introduction of high-resolution HLA typing, which have replaced serologic typing in this field of study [10,11]. Drug toxicity/HLA associations have been best documented for immunologically mediated reactions, such as drug hypersensitivity reactions associated with the use of abacavir, and severe cutaneous adverse drug reactions, such as Stevens-Johnson syndrome and toxic epidermal necrolysis induced by carbamazepine and allopurinol use, respectively. The testing of HLA-ABC screening for the early diagnoses of potential drug reactions may thus be of interest in dermatologic practice for selected patients [10,11].

In our results, we found multiple antigen presenting cells present within the inflammatory reaction around dermal blood vessels, hair follicles and sweat glands. Other authors have reported similar findings [11]. Other authors also reported that following neurotoxicity screening, a gliosis reaction represents a hallmark of many types of nervous system injury [12].

Using a battery of neurotoxic agents, the authors showed that overexpression of the astroglial protein glial fibrillary acidic protein (GFAP) [12], could represent a skin biomarker of drug neurotoxicity. Qualitative and quantitative analysis of GFAP has shown this biomarker to be a sensitive and specific indicator of neurotoxic conditions. In our study, we found that GFAP was overexpressed and seemed to be associated with the hair follicular isthmus.

In summary, we conclude that the in situ immune response to our selected drug eruption is complex; more cases are needed to understand its pathogenesis. Given the aging of the population in some countries, older patients are often prescribed multiple medications for multiple clinical issues. The vendors of metformin clearly state that either the doctors and/or the pharmacist should avoid using metformin simultaneously with nonprescription medications such us vitamins, nutritional supplements, and herbal products. They also recommend avoiding the following medications:

1) acetazolamide (Diamox);
2) amiloride (Midamor, in Moduretic);
3) angiotensin-converting enzyme (ACE) inhibitors such as benazepril (Lotensin), captopril (Capoten), enalapril (Vasotec), fosinopril (Monopril), lisinopril (Prinivil, Zestril), moexipril (Univasc), perindopril (Aceon), quinapril (Accupril), ramipril (Altace), and trandolapril (Mavik);
4) beta-blockers such as atenolol (Tenormin), labetalol (Normodyne), metoprolol (Lopressor, Toprol XL), nadolol (Corgard), and propranolol (Inderal);
5) calcium channel blockers such as amlodipine (Norvasc), diltiazem (Cardizem, Dilacor, Tiazac, others), felodipine (Plendil), isradipine (DynaCirc), nicardipine (Cardene), nifedipine (Adalat, Procardia), nimodipine (Nimotop), nisoldipine (Sular), and verapamil (Calan, Isoptin, Verelan);
6) cimetidine (Tagamet);
7) digoxin (Lanoxin);
8) other diuretics, including furosemide (Lasix);
9) androgen and estrogen hormone replacement therapy;
10) insulin and other medications for diabetes;
11) isoniazid;
12) medications for asthma and colds;
13) medications for mental illness and nausea;
14) medications for thyroid disease;
15) morphine (MS Contin, others);
16) niacin;
17) oral contraceptives;
18) other oral steroids such as dexamethasone (Decadron, Dexone), methylprednisolone (Medrol), and prednisone (Deltasone);
19) phenytoin (Dilantin, Phenytek);
20) procainamide (Procanbid);
21) quinidine;
22) quinine;
23) ranitidine (Zantac);
24) topiramate (Topamax);
25) triamterene (Dyazide, Maxzide, others);
26) trimethoprim (Primsol);
27) vancomycin (Vancocin);
28) zonisamide (Zonegran) [13].

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RECURRENT POSTCOITAL FIXED DRUG ERUPTION CAUSED BY CO-TRIMOXAZOLE MIMICKING A SEXUALLY INDUCED DISEASE

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Abstract
We report a case of a woman which had in 6-months three episodes of a recurrent postcoital skin eruption, each lasting for a few weeks. It seemed like a sexually induced eruption. She admitted to take only her permanent therapy that could not be connected to her skin signs. Thanks to her Health Insurance Card with the digital record of all the drugs she had received in the last two years it was possible to find out that she was intermittently taking co-trimoxazole in order to prevent an after intercourse urinary bladder infection. A good evidence of the patient's medication has a key role in the diagnosis of skin adverse drug reactions. Fixed drug eruption is a common adverse drug reaction and everyone prescribing a long term antibiotic prophylaxis should be aware of it.

Key words: postcoital fixed drug eruption; co-trimoxazole; therapy digital recording

Cite this article:
Marko Vok: Recurrent postcoital fixed drug eruption caused by co-trimoxazole mimicking a sexually induced disease. Our Dermatol Online. 2013; 4(2): 196-198.

Introduction

Fixed drug eruption (FDE) is a common adverse drug reaction. Co-trimoxazole, a combination of sulphamethoxazole and trimethoprim, is one of the most frequent causative agents. Co-trimoxazole induced FDE is often located on the male genital [1,2]. It can be also caused during the sexual intercourse by genital fluids of the partner taking the drug systemically or locally [3,4]. We report a case of a recurrent FDE appearing after the intercourse, although not induced by it.

Case Report

A 41-year-old woman reported three episodes of skin and oral mucous eruption from October 2011 till March 2012 each time following a sexual intercourse with her husband. Multiple acute lesions developed the day after the intercourse as sharply marginate, round or oval, burning erythema multiforme-like plaques with a 1 to 5 cm diameter, lasting for about a week and followed by some weeks lasting pigmentations. Multiple lesions were on the trunks, limbs and sometimes on the oral mucous (Fig. 1-4).
She regularly takes levothyroxin and a combination of drospirenon and ethynilestradiol tablets. She denied taking other medicaments. Thanks to the Health Insurance Card System we could see on her card the digital record of all the drugs she acquired in the last two years (Fig. 5). So, we saw that her general practitioner had prescribed her two boxes of co-trimoxazole in April 2011. After we had warned her, she remembered that she used to take a half or whole tablet of co-trimoxazole after the intercourse in order to prevent a urinary bladder infection to which she was prone. Partly she attributed it to her husband insulin dependent diabetes mellitus. He regularly takes insulin and metformin. In the past our patient had already taken co-trimoxazole tablets several times, but she has started to experience problems only since October 2011. She was not quite sure if she took a tablet after every intercourse, but mostly she did.
In our opinion the diagnosis of FDE to co-trimoxazole was clear-cut. We recommended her to avoid it. In the ten months period after she had stopped taking co-trimoxazole the FDE did not recur.

Discussion

The diagnosis of FDE is considered as easy when the presentation and the course of the disease are typical. FDE usually appears as a solitary or multiple, well circumscribed, erythematous-violaceous oedematous plaques and in a regression period into some weeks lasting pigmentations. The lesions usually recur 30 minutes to 8 hours after the exposure to the causative drug at the same sites of the previous eruption.

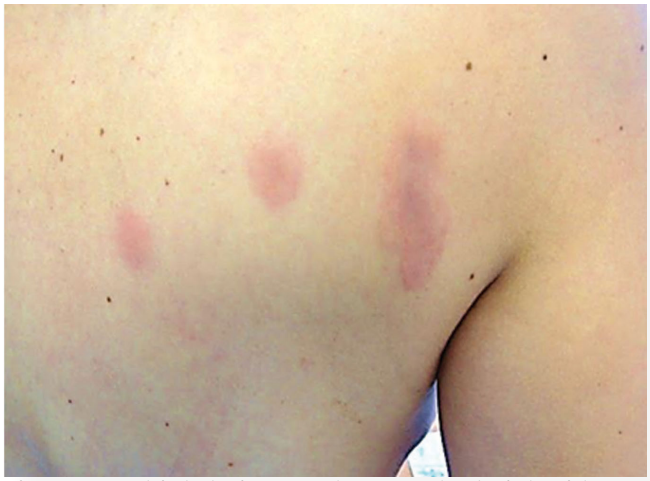

Figure 1. Multiple lesions on the upper back right side

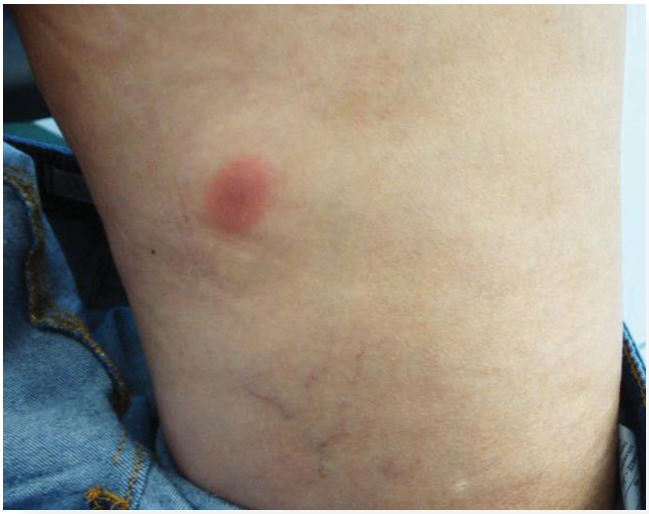

Figure 2. Lesion on the left thigh

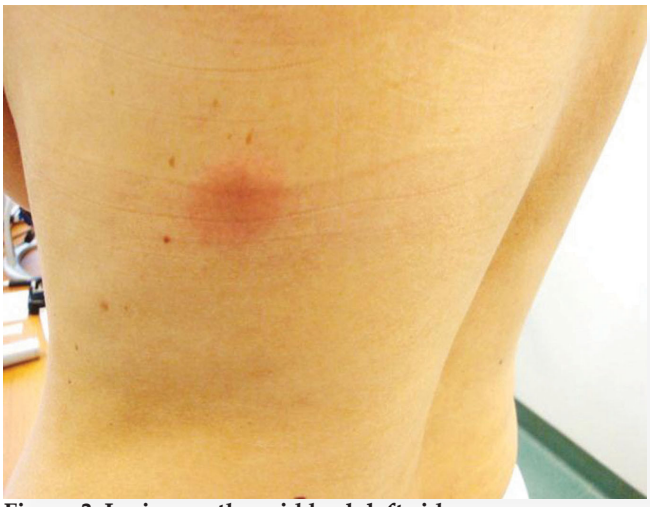

Figure 3. Lesion on the mid back left side

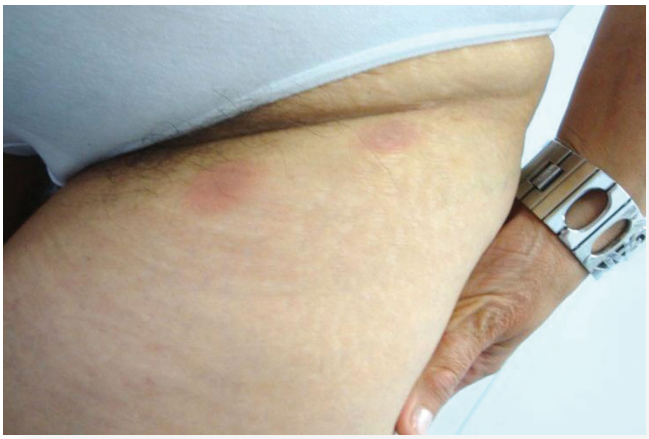

Figure 4. Two lesions on the left groin

Figure 5. Sample of the Slovenian Digital Health Insurance Card

The previously involved sites do not flare with each exposure, which is known as the refractory period. The duration of this period varies from a few weeks to several months. It seems that FDE develops more frequently to intermittently receiving drugs than to those taking permanently. The sensitization period varies from some weeks to several years [5]. There are some reports of spontaneous or induced desensitization to the causative drug in cases of FDE. The exact mechanism of tolerance induction is still poorly understood [6,7].

Our reported case of FDE to co-trimoxazole is a quite typical case for its clinical picture, course, latency period from intake to appearance, refractory period, several years lasting sensitization period and for being caused by an intermittently taken drug.

In the pathogenesis of FDE epidermal $CD8^+$T cells resident in the FDE lesions have a major role. FDE is considered a T cell mediated delayed drug hypersensitivity reaction with important specificities in the effector cells involved [8].

Oral challenge is considered the most reliable method for establishing the causative drug in FDE. It is usually performed by administering a single dose of the suspected drug, starting at one-tenth of the therapeutic dose [5]. In some rare cases it should be kept in mind that the oral challenge may induce a severe reaction.

Patch testing represents a simple and safe screening test especially when multiple drugs are suspected. A positive patch test reaction is usually a strong evidence of a causative drug. A negative patch test does not exclude a possible hypersensitivity.

The patch test should be always done in the site of a previous lesion at least two weeks after its disappearance (refractory period). Persistent lack of reactivity to some drugs is an important limitation. One of the reasons of the false negative patch test result could be the impaired penetration of the specific drug through the stratum corneum, the other that the patient may not be sensitized to the original drug but to its metabolites [5,9].

Patch testing with co-trimoxazole in FDE is not yet standardized. The available commercial hapten of co-trimoxazole is a 10% dilution in petrolatum. But it seems that petrolatum is not a suitable vehicle and that dimethylsulfoxide is more appropriate. Özkaya-Bayazit et al. reported of 25 positive reactions (20 to sulphamethoxazole and 5 to trimethoprim) in 27 patients with co-trimoxazole FDE patch tested with different concentrations of sulphametoxazole and trimethoprim diluted in dimethylsulfoxide, while all patients had negative results when they used petrolatum as a vehicle [10].

In our patient we did not perform any diagnostic test because there was no need for that. In fact, the patient had done the challenge test herself the evening before the visit by having sex with her husband and the usual taking of a tablet of co-trimoxazole after the intercourse. Only that she attributed the skin symptoms to the intercourse and not to the drug.

Conclusions

Our case shows the importance of the information of drugs taken by patients. For different reasons they often omit to mention all the drugs that they take. In these cases an accompanying electronic evidence of the prescribed drugs could be crucial to solve the problem of an adverse drug reaction. Our case also shows how misleading can be the history of the disease. In our patient it was typical for a sexually induced skin eruption. In fact it was the consequence of a relatively common after intercourse antibiotic prophylaxis of urinary bladder infections. The possibility of a cutaneous adverse drug reaction should be considered by everyone prescribing a long term intermittent antibiotic prophylaxis.

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SPECIFIC CUTANEOUS HISTOLOGIC AND IMMUNOLOGIC FEATURES IN A CASE OF EARLY *LUPUS ERYTHEMATOSUS SCARRING ALOPECIA*

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Abstract
Introduction: Immunoreactants detected by direct immunofluorescence (DIF) in the skin of patients with lupus erythematosus represent an important tool in the diagnosis of this disorder.
Case report: A 46 year old African American female presented complaining of hair loss and scarring in her scalp.
Methods: Biopsies for hematoxylin and eosin (H&E) examination, as well as for direct immunofluorescence (DIF) were performed.
Results: The histologic features were representative of early lupus erythematosus. DIF demonstrated immune deposits of several immunoglobulins and complement, primarily around skin appendageal structures(hair follicles and sweat glands). Deposits of immunoglobulin D were seen in several areas of the epidermis.
Conclusion: In lupus erythematosus, evaluation of immune reactions against cutaneous appendageal structures may be crucial in differentiating this disorder from other autoimmune and non-autoimmune diseases.

Key words: discoid lupus erythematosus (DLE); scarring alopecia; direct immunofluorescence(DIF); skin appendices; lichen planopilaris (LPP)

Abbreviations and acronyms: Hematoxylin and eosin (H&E), direct immunofluoresence (DIF), discoid lupus erythematosus (DLE), pseudopelade of Brocq (PB).

Cite this article:
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Introduction

Pseudopelade of Brocq is a progressive, scarring alopecia characterized by early alopecic patches localized in the scalp, that then coalesce into larger, irregular plaques with polycyclic borders [1]. Pseudopelade of Brocq can be considered either the final atrophic stage of multiple scarring disorders such as lichen planopilaris (LPP) and discoid lupus erythematosus (DLE), ie, secondary PB or, alternatively, a discrete nosologic disease (primary PB) [1].

Case Report

PA 46 year old African American female was evaluated for hair loss and scarring in her scalp. The patient reported a family history of lupus erythematosus. Physical examination confirmed a scarring alopecia in patches, with focal desquamation, erythema and hyperpigmentation. Skin biopsies were obtained for hematoxylin and eosin (H&E) review, and for direct immunofluorescence. Laboratory data demonstrated a normal complete blood count (CBC) and differential analysis, and a normal erythrocyte sedimentation rate. Antiphospholipid antibody testing was negative; serum electrolytes, blood urea nitrogen, creatinine, and liver function tests, as well as urinalysis and chest radiographs were within normal limits. The antinuclear antibody (ANA) titer was normal. Specific ANA screening yielded negative results for anti-Smith, anti-double stranded DNA (dsDNA), and anti-histone antibodies. Tests for anti-ribonuclease antigen (RNAse), extractable nuclear antigen (ENA), small nuclear antigen(sn), ribonucleoproteins (RNPs), and U1 and U2 complexes were negative, as was testing for anti-SS-A (anti-Ro) and anti-SS-B (anti-La). Levels of Complement/C3 and C4 were within normal limits. Perinuclear anti-neutrophil cytoplasmic antibody testing was negative. However, both histologic and direct immunofluorescence (DIF) findings were representative of early lupus erythematosus.

Materials and Methods

Hematoxylin and eosin staining was performed as previously described [2-7].

Direct immunofluorescence (DIF):

In brief, skin cryosections were prepared, and incubated with multiple fluorochromes as previously reported [2-8]. We utilized normal skin as a negative control from patients going under aesthetic plastic surgery. To test the immune response in lesional skin, we utilized the following markers: antibodies to immunoglobulins A, G, D, E and M; IgG3 and IgG4; Complement/C1q and C3; kappa light chains, lambda light chains, fibrinogen and albumin. All antibodies were fluorescein isothiocyanate (FITC) conjugated, and all obtained from Dako (Carpinteria, California, USA).

Results

Examination of the H&E tissue sections demonstrates no significant epidermal follicular plugging. A mild, interface infiltrate of lymphocytes and histiocytes was noted. Within the dermis, a mild, superficial and deep, perivascular and periadnexal infiltrate of lymphocytes, histiocytes and plasma cells was observed. Occasional neutrophils are present within the infiltrate. Eosinophils were rare. Increased dermal mucin was not appreciated. Minimal, perifollicular dermal scarring was noted, approximating ten (10) per cent of the biopsy area. The histologic features were representative of early lupus erythematosus. The Verhoeff elastin special stain confirmed the extent of dermal scarring (Fig. 1). The PAS special stain displayed positive staining around the skin appendageal structures, and revealed no fungal organisms (Fig. 1).

Direct immunofluorescence (DIF): DIF demonstrated the following results: IgG (+, focal granular epidermal stratum spinosum, and dermal perivascular and periadnexal); IgG3 (-); IgG4 (+, focal granular epidermal stratum spinosum, and dermal perivascular); IgA (+, focal granular deep dermal perivascular); IgM (+, focal granular dermal perivascular, also in superficial epidermal free nerves and periadnexal); IgD (+, focal granular epidermal stratum spinosum cytoplasmic); IgE (-); Complement/C1q (-); Complement/C3 (+, Focal granular epidermal straum spinosum, and dermal perivascular); Kappa light chains (++, Focal granular epidermal stratum spinosum, and dermal perivascular and periadnexal); Lambda light chains (+, focal granular epidermal stratum spinosum); Albumin (++, focal granular dermal perivascular) and fibrinogen (++, focal granular epidermal stratum spinosum, and focal dermal perivascular and periadnexal). (Fig. 1). Since the H&E biopsy demonstrated an early scarring alopecia compatible with lupus erythematosus and given the DIF results, the patient was prescribed oral prednisone, clobetasol gel, and sun protection.

Discussion

Pseudopelade of Brocq is a type of scarring alopecia of the scalp associated with a peculiar clinical presentation and evolution. Many authorities do not consider pseudopelade of Brocq a purely autonomous nosologic entity, because in 66.6% of patients it represents the end stage of other inflammatory chronic diseases such as lichen planopilaris and discoid lupus erythematosus. Primary cicatricial alopecias result from inflammatory destruction of the hair follicle, followed by its replacement by a fibrotic area [9]. It is often difficult to clinically differentiate between psueodopelade of Brocq, lichen planopilaris and discoid lupus erythematosus. Thus, histopathologic and immunopathologic studies such DIF are recommended in the workup of these disorders; overall, the appropriate diagnosis depends on clinicopathologic correlations Primary cicatricial alopecias are further subclassified as neutrophilic, lymphocytic and mixed types. Each of these groups contain specific disorders, including folliculitis decalvans, dissecting folliculitis of the scalp, erosive pustulosis of the scalp, keloidal acne of the nape, frontal fibrosing alopecia, lichen planopilaris and lupus erythematosus [9]. In our case, DIF reactivity against dermal skin appendices assisted in establishing a diagnosis of early lupus erythematosus.

Prompt diagnosis and treatment are needed in scarring lupus erythematosus to contain the hair loss, scarring and emotional distress that often accompany these sequelae [10]. The dermatologic nursing staff may facilitate the diagnostic and treatment process, and through educational and other supportive measures exert a positive impact on the patient's overall medical course [10].

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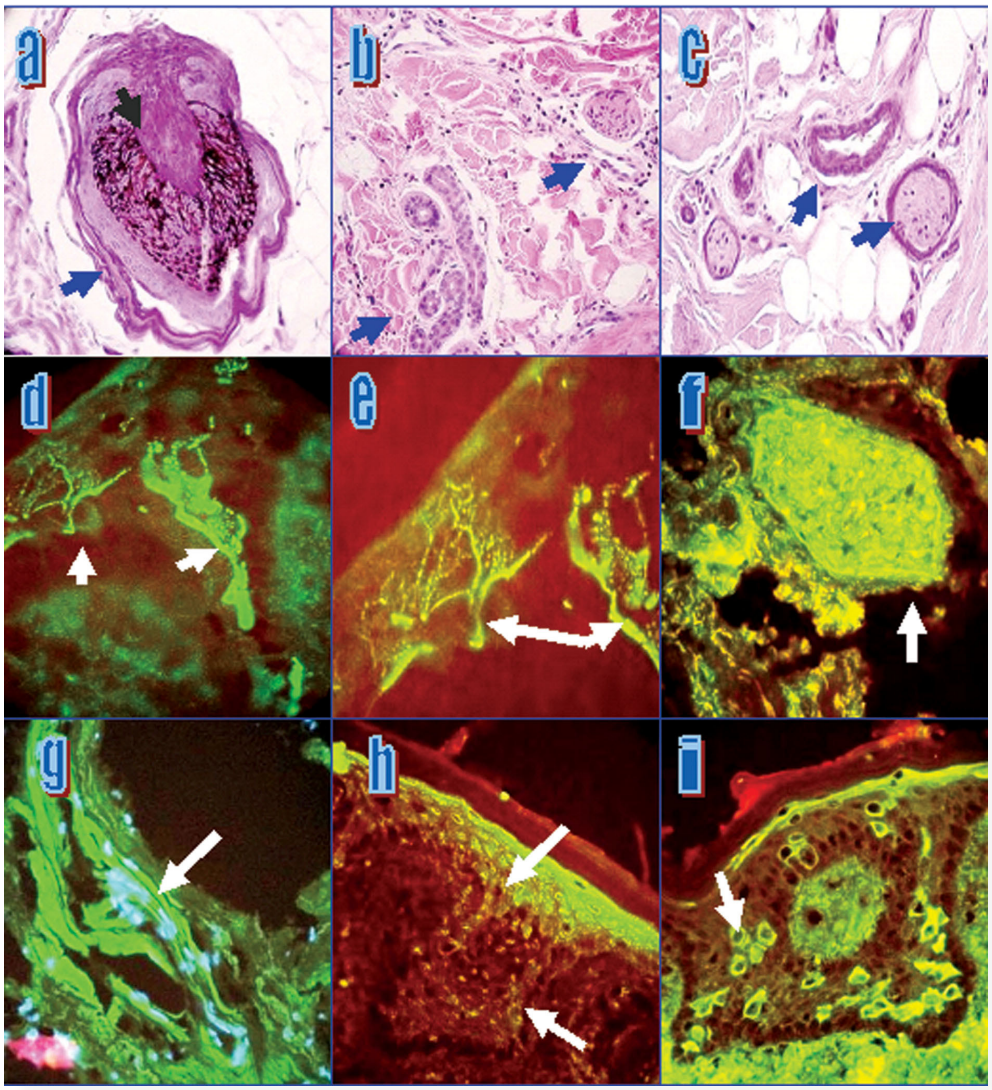

Figure 1. a PAS positive accentuation in a hair follicle outer root sheath (dark pink staining; blue arrow) and in the follicular papilla border area (green arrow) (400x). **b.** H&E staining, showing a mild inflammatory infiltrate around an eccrine gland coil and a neurovascular package (blue arrows) (100x). **c.** PAS positive accentuation around a dermal blood vessel and nerve (200x). **d and e.** DIF using FITC conjugated anti-human IgM antibody, and demonstrating positive staining against superficial, thin nerves entering the epidermis and in the upper dermal neurovascular plexus (green staining; white arrows). **f.** Positive DIF staining against a sebaceous gland, utilizing FITC conjugated anti-human fibrinogen antibody (green-yellow staining; white arrow). **g.** Positive DIF staining against a deep dermal blood vessel, utilizing FITC conjugated anti-human lambda light chains antibody (green staining; white arrow). **h.** Positive DIF staining with FITC conjugated anti-human IgD antibodies against the upper and central epidermal stratum spinosum layer in an anti-nuclear and/or perinuclear keratinocytic staining pattern (white arrows). **i.** Positive DIF FITC conjugated anti-human lambda light chains staining, in an epidermal stratum spinosum perinuclear and pericytoplasmic pattern (white arrow).

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STRICT ANATOMICAL CO EXISTENCE AND COLOCALIZATION OF VITILIGO AND PSORIASIS – A RARE ENTITY

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Abstract
The coexistence of psoriasis and vitiligo is rare. We describe a case report of a 58 year old female patient who developed typical psoraiatic plaques covering completely or partly the vitiliginous areas of her skin. Her psoriasis was strictly limited to the vitiliginous patches with no involvement of the normal skin. Strict anatomical coexistence of both diseases is extremely rare and suggests a casual mechanism, possibly due to a koebner phenomenon but genetic and environmental factors may also be involved.

Key words: autoimmune; colocalization; koebner phenomenon; patch; psoriasis; vitiligo

Cite this article:
Neerja Puri, Asha Puri: Strict anatomical co existence and colocalization of vitiligo and psoriasis – a rare entity. Our Dermatol Online. 2013; 4(2): 202-204.

Introduction

The occurrence of psoriasis in patients with vitiligo has not been often described [1-3]. Vitiligo and psoriasis are common conditions with a prevalence of approximately 1% and 3% respectively [4,5] and may be present in the same person. Patients with vitiligo and psoriasis may have the koebner phenomenon [3]. In 1982, Koransky and Roenig described the association of vitiligo and psoriasis to be rare [6,7]. The increased incidence of presumably autoimmune diseases in patients with vitiligo and psoriasis is an evidence of the autoimmune origin of these two conditions [8].

Case Report

A 58 year female reported to the department of dermatology with depigmented patch over axilla, neck, breast and groins, arms, forearm, elbows, hands, fingers, legs & thighs since 6 years. Patient noticed erythematous plaques with thick scaling over extensors and scalp since 2 years. Cutaneous examination revealed well defined and erythematous papules and plaques with silvery scales over arm, forearm, elbows, hands (Fig. 1), fingers, legs and thighs. There was mild pruritis present over lesions. There was co-habitation of psoriatic lesions over vitiligo patches. The PASI score of the patient was 12. Auspitz sign was positive.

A clinical diagnosis of coexistent vitiligo and psoriasis was made. On cutaneous examination, there were present thick plaque on the wrist over a depigmented patch measuring 8cm x 6cm in diameter. The plaque had thick silvery scaling. The depigmented patches were present over the tips of fingers, over central part of buttocks (10cm x 8cm) measuring 10cm x 8cm, over both axilla (right axilla 7cm x 6cm and left axilla 8cm x 5cm) and groins and breast (right breast 5cm x 3cm and left breast 4cm x 4cm). The nail showed pitting, beaus lines and longitudinal striations. All the investigations of the patient were within normal limits except the ESR which was 48 mm 1st hour. A clinical diagnosis of psoriasis in association with vitiligo was made.

Two skin biopsies of the patient were taken from the localized lesion. The skin biopsy taken from left elbow showed neutrophilic crust and parakeratosis. Epidermis showed loss of granular layer and elongation of rete ridges with suprapapillary thinning of the epidermis (Fig. 2). Dilated capillaries and papillary dermal oedema were seen. Lymphohistiocytic infiltrate was noted.

So, the biopsy from left elbow was consistent with psoriasis. The second biopsy was taken at the site of depigmented patch over the left elbow. The histopathological findings were as follows: Architecturally normal epidermis showed intact basal layer. Mild lymphohistiocytic infiltrate was present in the papillary dermis. There was an absenceof melanocytes in the basal cell layer confirmed by special staining (Fig. 3). There was inflammation at the dermoepidermal junction. The clinical features were consistent with colocalized vitiligo and psoriasis.

The patient was put on melanocyl 0.6 mg/kg body weight on alternate days, but patient complained of increased photosensitivity with aggravation of lesions after a few weeks of treatment. Subsequently, the patient was put on methotrexate 0.2 mg/kg body weight. The response started appearing within two weeks of treatment.

The psoriatic lesions cleared upto 6 weeks after treatment with methotrexate (with a drop of PASI score to 3.2), but the repigmentation in vitiligo lesions took a longer time. After 12 weeks of treatment, the vitiligo patches showed mild repigmentation (Fig. 4). The patient is still on treatment for vitiligo and is on a regular follow up.

Discussion

In our case, the patient first presented with vitiligo and later psoriasis developed at the site of vitiligo. Papdavid et al. [3] stated that in cases of psoriasis limited to areas of vitiligo, their coexistence resulted from koebner phenomenon. Several studies indicated a polygenic model for vitiligo [9] and psoriasis [10,11].

Many susceptibility loci of psoriasis [12] and vitiligo [13,14] have been mapped and the subsequently locus for vitiligo. AISI, in chromosome IP 31, is situated close to the susceptibility locus for psoriasis PSORS7 [13].

Nevertheless, the possibility of loci being identical is minimized by the low prevalence of psoriasis in patients with vitiligo.

In 1989, Menter et al reported the first possible case of psoriasis guttate restricted to areas of vitiligo. In 1998, Dhar and Malaks described the first likely case of vitiligo associated with psoriasis in a pediatric patient, a nine year old boy [15].

The hypothesis that generalized vitiligo [16] is an autoimmune process is based principally on its association with other presumably immunologic disorders and demonstration of antimelanocyte antibody. Co-habitation of two disorders which possess a prominent immunological component in their pathogenesis may offer a clue as their causation [17].

To conclude, we report the co-existence of vitiligo and psoriasis with few colocalized lesions. Many pigment cell biologists and dermatologists have concluded that vitiligo is an autoimmune disease and psoriasis, the T cell mediated skin disease [18], may be associated. Whether their interpretation of the association is valid is uncertain. This requires further insight into their pathogenesis, as we believe that psoriasis and vitiligo are spectra of diseases with varied etiology for varied clinical presentation [19].

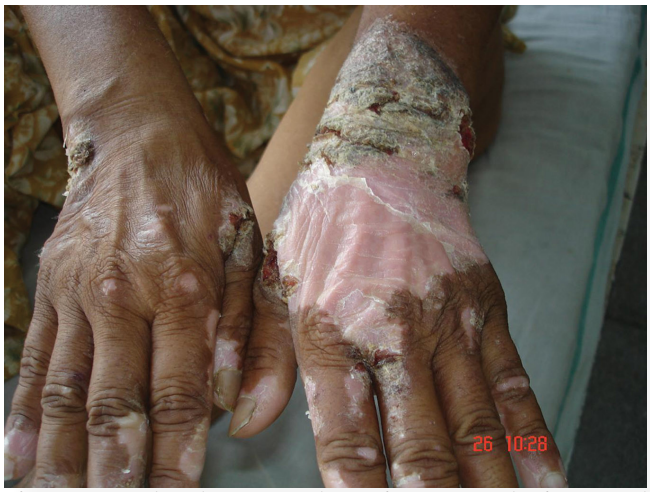

Figure 1. Scaly plaque on the wrist over a depigmented patch before treatment

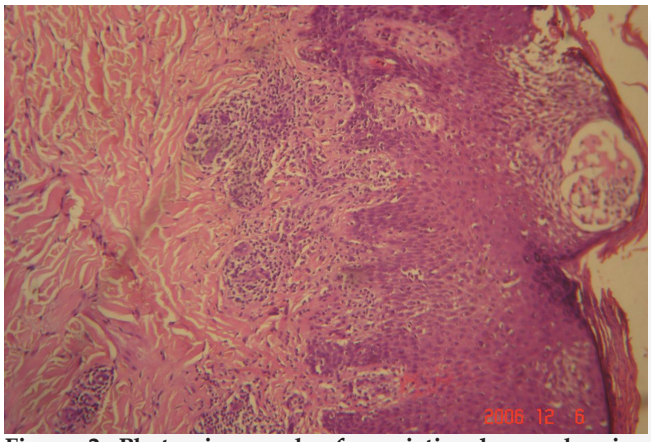

Figure 2. Photomicrograph of psoriatic plaque showing micromunro abscess H & E stain

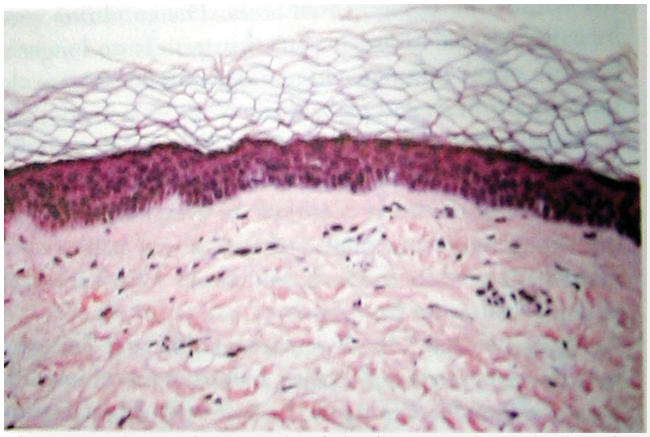

Figure 3. Photomicrograph of depigmented patch showing absence of melanocytes in the basal layer

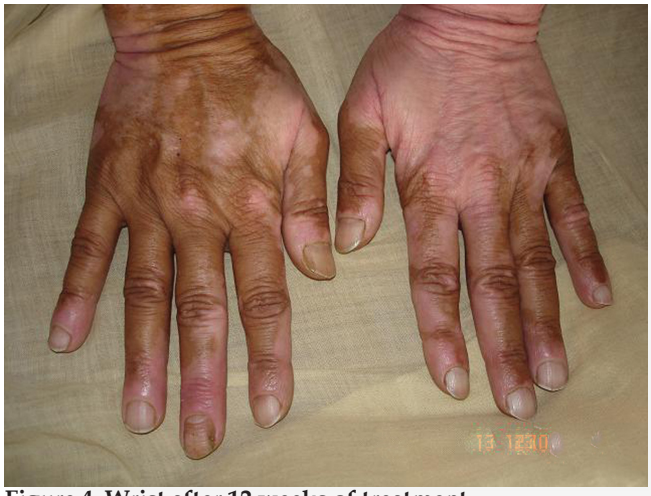

Figure 4. Wrist after 12 weeks of treatment

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INFANTILE PSORIASIS TREATED SUCCESSFULLY WITH TOPICAL CALCIPOTRIENE

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None

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Abstract
Infantile psoriasis is a benign disease. Systemic features are rare and spontaneous remission occurs. There is a hazard of viral infection particularly in steroid treated children. Psoriasis in infancy is often more therapeutically challenging than atopic and seborrheic dermatitis. We report a case of nine month old infant treated with topical calcipotriene for infantile psoriasis who experienced greater benefit than he had with standard corticosteroid medications.

Key words: psoriasis; infantile; calcipotriene; scaling; childhood; histopathology

Cite this article:
Neerja Puri: Infantile psoriasis treated successfully with topical calcipotriene. Our Dermatol Online. 2013; 4(2): 205-207.

Introduction

Psoriasis is a common inherited papulosquamous dermatosis that may be a diagnostic dilemma, particularly in infants and children [1]. The treatment of children with psoriasis should be handled with caution and tailored according to the child's age, as well as to the extent, distribution, and type of psoriasis. Childhood psoriasis is a disease with manifold clinical presentations which can make the correct diagnosis sometimes difficult. Infantile variety of psoriasis may sometimes be confused as sebopsoriasis, i.e., in between stage of seborrhic dermatitis and psoriasis where lesions are mainly confined to scalp, eyebrow and behind the ear but on histopathology typical spongiform pustules are absent. Infantile variety of psoriasis is rare (about 1-2% of pediatric psoriasis) and only two cases of congenital variety have been reported [2]. The clinical manifestations of psoriasis in a child are generally similar to those in an adult. However, the condition often takes on atypical forms in children which can lead to diagnostic problems. Certain childhood dermatoses which involve the buttocks, eyelids and scalp strongly resemble psoriasis [3].

Case Report

A nine month old boy reported with complaint of generalized scaling for last two months. He was referred from periphery with a diagnosis of extensive seborrhic dermatitis and was treated with some topical medication containing salicylic acid and corticosreroids. Initially, mother noticed a circular greyish scaly lesion about 3 cm in diameter over parietal region of scalp. The scales were large and loose. The lesion was non discharging and scalp hair were normal. The lesion then gradually spread to involve the entire scalp and quickly affected whole of the body including palms and soles (Fig. 1). In some lesions over abdomen (Fig. 2), pustules were found under the scales and on removal of scales pin point bleeding was noticed (Auspitz sign). During the course of illness there was spontaneous remission of some lesions which was followed by recurrence. No history of similar lesions in other family members was present. The baby was well nourished with normal motor and mental milestones. Birth history was uneventful and baby was exclusively breast fed and immunized. Skin biopsy showed parakeratosis with psoriasiform hyperplasia of epidermal lining in one area of the upper epidermal layer and Munromicroabscess (Fig. 3). There was exocytosis with spongiosis wherein acute inflammatory cells were found. The patient was put on topical calcipotriene. 0005% for a period of 4-6 weeks. Laboratory testing for calcium metabolism was normal during the course of therapy. Remarkable improvement was seen after six weeks of treatment with clearance of scales and decrease of erythema.

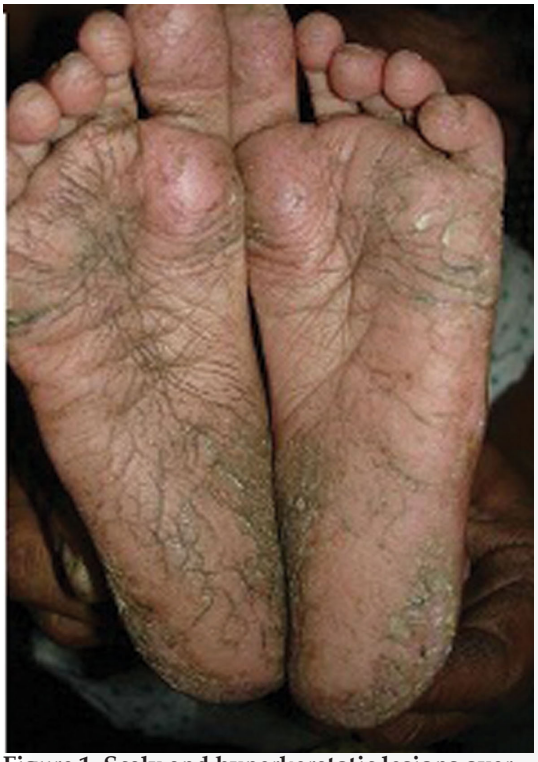

Figure 1. Scaly and hyperkeratotic lesions over the soles

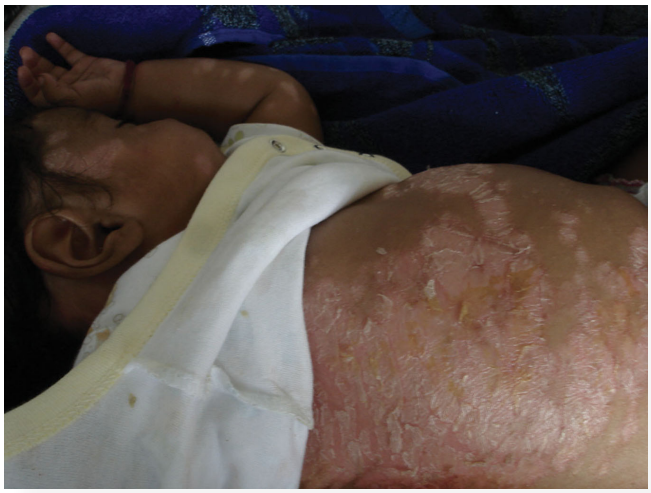

Figure 2. Erythematous scaly plaques over the abdomen of a nine month old child

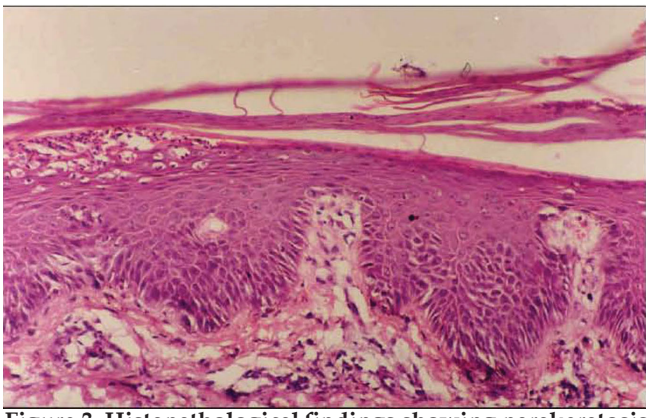

Figure 3. Histapathological findings showing parakeratosis with Munromicroabscess with spongiosis H&E stain 100X

Discussion

The onset of psoriasis is observed before the age of 10 years in 15% of all patients. The clinical pattern often takes on a peculiar form [4]. Psoriasis guttata--or less frequently nummular psoriasis--is the initial phase during childhood. It is very difficult to establish a diagnosis on the basis of incipient features when childhood psoriasis is located on the head, palms, soles, or on the fingers, toes and nails [5]. Intertriginous or flexural psoriasis, psoriasis spinulosa and oral psoriasis is also described. Infantile psoriasis resembles adult psoriasis and causes itching in 30% of cases. However, it has some specific clinical features. The Koebner phenomenon, whereby psoriatic lesions appear on areas of the skin that have been traumatised or irritated, is particularly common in children [6].

The fact that a child has suffered with psoriasis from his or her earliest years is not in itself an unfavourable prognostic factor. Similarly, the onset of severe psoriasis during childhood does not mean that the child will continue to suffer from severe psoriasis as an adult. However, given the chronic nature of psoriasis, it is highly likely that the child will continue to suffer from flares of psoriasis punctuated by periods of remission for the rest of his or her life. Psoriasis can appear very early in life, but is rarely present from birth. However, babies are subject to a particular form of psoriasis called napkin psoriasis. This is a dermatosis with lesions chiefly present on the buttocks due to irritation of the skin by urine and stools. These lesions are not obviously psoriatic, and this type of psoriasis poses diagnostic problems. It is difficult to be sure if the baby has psoriasis or simply a dermatosis on the buttocks which resembles psoriasis. In older children, psoriasis has to be distinguished from a seborrheic dermatosis resulting in lesions in the skin folds and on the buttocks and scalp. The generalized nature of psoriasis and the intensity of inflammation often reduce the efficacy of topical corticosteroids. Furthermore, involvement of intertriginous skin and the presence of scalp disease limit the potency of the topical steroids that can be prescribed. Guttate psoriasis is particularly common in childhood. It is characterised by the sudden appearance of small, red, scaly lesions particularly on the trunk, arms and legs.

The lesions in children are located in virtually the same places as in adults. However, whereas facial lesions are rare in adults (present in 5.6% of cases), they are common in children (present in 30% of cases). Lesions are found on the forehead and cheeks, which become very red, and sometimes involve the eyelids and ears. Facial lesions have profound consequences on the patient's ability to form relationships.

The type of treatment prescribed by the dermatologist should not only be appropriate to the clinical form of psoriasis but also take into account the wishes of the child (if he or she is old enough to express them) and of the parents. The dermatologist, parents and child should work together to find the treatment which best suits the child. The benefits and risks of a treatment should be weighed even more carefully than for adults, especially as regards systemic treatments. Because of the toxicity of certain drugs, there are fewer treatments available for children than adults. Infantile psoriasis is commonly treated sequentially, with treatment altered every three months (it is treated the same way in adults). Lesions are less visible during summer because of exposure to the sun. Treatment should be recommenced when new eruptions appear, and continued until lesions turn white. Most importantly, the skin must be moisturised, usually by taking baths with emollients and using moisturising creams. Cortisone-based ointments are highly effective on psoriasis lesions [7]. The dermatologist may suggest a systemic treatment when the psoriasis is very widespread or severe and when it is having a significant impact on the patient's life.

Systemic treatments have significant side effects and their advantages and disadvantages should be carefully balanced with the child and his or her family. They are usually prescribed for a very short period of time and their use must be monitored. PUVA therapy is generally used only on children over 15 years as it leads to an increased risk of cancer. Retinoids are used for treating pustular and erythrodermic psoriasis and psoriatic rheumatism [8,9]. Children must be particularly carefully monitored to make sure that retinoids are not inhibiting their growth. Calcipotriene is very effective over the intertriginous areas where corticosteroids cant be prescribed [10]. Moreover the potent corticosteroids can be hazardous in children.

Conclusions

We conclude that calcipotriene can be a safe and effective therapy for psoriasis in early infancy.

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OCULOCUTANEOUS ALBINISM COMPLICATED WITH AN ULCERATED PLAQUE

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Abstract
A 32-year-old male with a history of albinism and farmer by occupation presented with an ulcerated plaque on the right wrist. The patient had light eyes, hair, and skin. Physical examination showed extensive photodamage. A skin biopsy specimen from the plaque revealed a well-differentiated squamous-cell carcinoma. Wide surgical excision was done. The most common types of oculocutaneous albinism (OCA), OCA 1 and OCA 2, are autosomal recessive disorders of pigmentation that commonly affect the skin, hair and eyes. Photodamage and skin cancers plague patients with albinism. Albinos face a myriad of social and medical issues. Importance of photoprotection, skin cancer surveillance and treatment has been stressed upon in this report.

Key words: albinism; photoprotection; melanin; squamous cell carcinoma

Cite this article:
Belliappa Pemmanda Raju, Umashankar Nagaraju, Leena Raveendra, Vivekananda, Priya Kootelu Sundar, Lokanatha Keshavalu: Oculocutaneous albinism complicated with an ulcerated plaque. Our Dermatol Online. 2013; 4(2): 208-211.

Introduction

Albinism is a genetically inherited disorder characterized by hypopigmentation of the skin, hair and eyes due to a reduced or lack of cutaneous melanin pigment production [1]. Generally, there are two principal types of albinism, oculocutaneous, affecting the eyes, skin and hair, and ocular affecting the eyes only [1,2]. The mode of inheritance of albinism is thought to vary, depending on the type. The oculocutaneous type is considered autosomal recessive, and the ocular variant sex-linked [1-3]. Ocular problems faced by albinos are nystagmus, strabismus, photophobia, foveal hypoplasia and decreased visual acuity. The cutaneous problems seen with oculocutaneous albinism include sunburns, blisters, centro-facial lentiginosis, ephelides, solar elastosis, solar keratosis, basal cell carcinomas and squamous cell carcinomas. Squamous cell carcinoma has been reported to be the commonest skin malignancy seen in albinos [4,5].
Albinos are at an increased risk of developing skin malignancies due to the absence of melanin, which is a photo protective pigment, protecting the skin from the harmful effects of ultraviolet radiation [6]. Hence, a regular examination for early detection and treatment of these malignancies would increase their life expectancy to a great extent. We report here a case of oculocutaneous albinism with well-differentiated squamous cell carcinoma in a farmer and also review oculocutaneous albinism with emphasis on treatment and preventive aspects.

Case Report

A 32- year- old male, who was a known case of Oculocutaneous Albinism, presented with an ulcerated lesion over the right forearm. It started as a small wound which arised from normal looking skin and gradually increased to the present form over a period of six months. The patient was a farmer who had occupational sun exposure with no apparent photoprotection for the past fifteen years. History of photosensitivity and photophobia was present. He was born out of a non-consanguineous marriage. He had an elder sibling who was unaffected, but had an affected first-degree relative.
On physical examination, generalized depigmented skin with white hairs, brownish freckles and telangiectasia were seen on his body (Fig. 1 - 3). Ocular Examination revealed photophobia, nystagmus and decreased visual acquity.
Ulcerated plaque measuring 4cm x 3 cm with crusting with rolled out edges was seen on the right wrist (Fig. 4a, b). It was fixed to the underlying tissues. Regional lymphadenopathy was absent. General physical and systemic examination was normal.

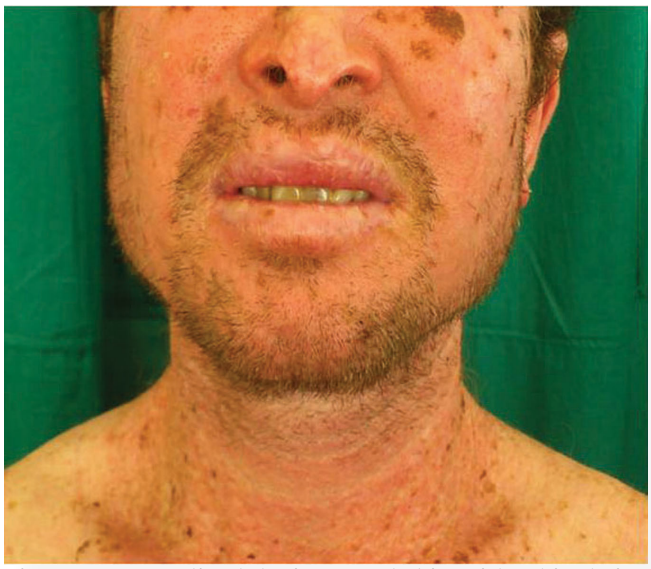

Figure 1. Generalized depigmented skin with white hairs, brownish freckles and telangiectasia

On the basis of these clinical findings, a differential diagnosis of Oculocutaneous Albinism with Squamous Cell Carcinoma (SCC)/ Basal Cell Carcinoma (BCC)/ Malignant Melanoma (MM) was made.

Routine haematological and biochemical investigations were normal. Chest X-Ray and Ultrasound Abdomen did not reveal any abnormalities. Biopsy from the ulcerated plaque showed features of well differentiated squamous cell carcinoma (Fig. 5). Wide surgical excision was done in this patient and counselled regarding photoprotection. He is under long term follow-up for recurrences.

Discussion

Oculocutaneous albinism (OCA) is a group of four autosomal recessive disorders caused by either a complete lack or a reduction of melanin biosynthesis in the melanocytes resulting in hypopigmentation of the hair, skin and eyes. Reduction of melanin in the eyes results in reduced visual acuity caused by foveal hypoplasia and misrouting of the optic nerve fibres.

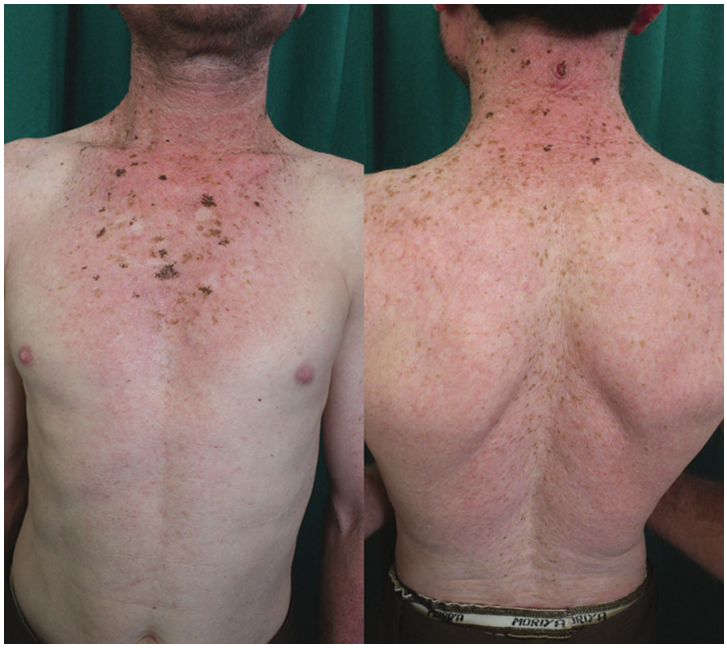

Figure 2 and 3. Depigmentation with brownish freckles and telangiectasia over the chest and back

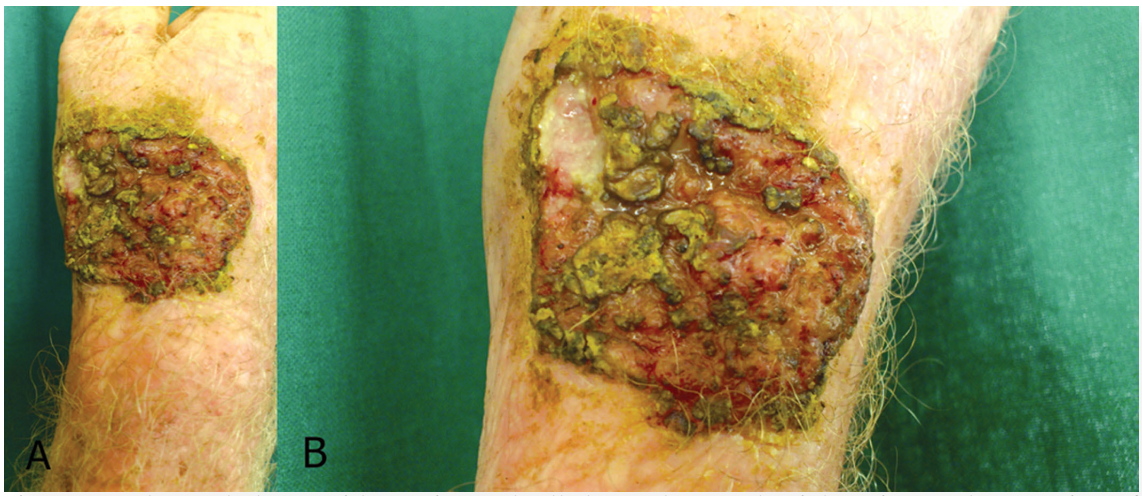

Figure 4A. Ulcerated plaque with crusting and rolled out edges on the right wrist. B. Close-up view of the ulcerated plaque

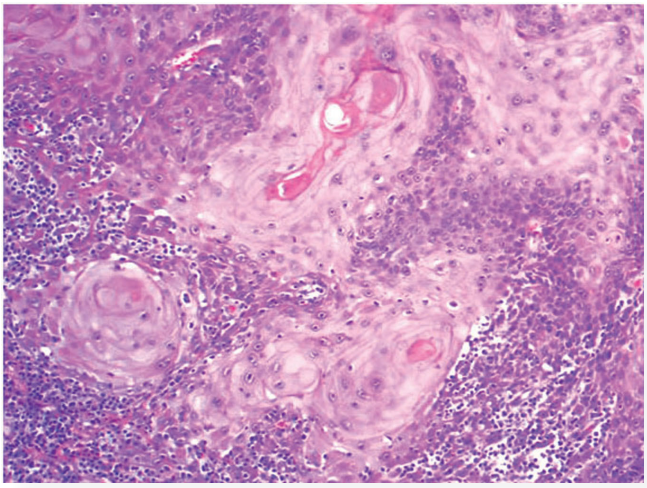

Figure 5. Invasive neoplastic cells with keratin pearl formation (H&E, 10x)

The clinical spectrum of OCA varies, with OCA1A being the most severe type characterized by a complete lack of melanin production throughout life, while the milder forms OCA1B, OCA2, OCA3 and OCA4 show some pigment accumulation over time. The different types of OCA are caused by mutations in different genes but the clinical phenotype is not always distinguishable, making molecular diagnosis a useful tool and essential for genetic counseling [7].

Albinism can affect people of all ethnic backgrounds and has been extensively studied. Approximately one in 17,000 people have one of the types of albinism [8]. This suggests that about 1 in 70 people carry a gene for OCA [7]. Prevalence of the different forms of albinism varies considerably worldwide, partly explained by the different founder mutations in different genes and the fact that it can be difficult clinically to distinguish between the different subtypes of albinism among the large normal spectrum of pigmentation. OCA2 is the most prevalent form worldwide [9].

OCA1 is caused by mutations in the tyrosinase gene on chromosome 11q14.3 [10]. Mutations completely abolishing tyrosinase activity result in OCA1A, while mutations rendering some enzyme activity result in OCA1B allowing some accumulation of melanin pigment over time. Mutations in the OCA2 gene (formerly known as the P-gene) cause the OCA2 phenotype [11].

All types of OCA and ocular albinism (OA) have similar ocular findings, including various degrees of congenital nystagmus, hypopigmentation of iris leading to iris translucency, reduced pigmentation of the retinal pigment epithelium, foveal hypoplasia, reduced visual acuity usually in the range 20/60 to 20/400 and refractive errors, and sometimes a degree of color vision impairment [8,12]. Photophobia may be prominent. A characteristic finding is misrouting of the optic nerves, consisting in an excessive crossing of the fibres in the optic chiasma, which can result in strabismus and reduced stereoscopic vision [13]. Absence of misrouting excludes the diagnosis of albinism.

The degree of skin and hair hypopigmentation varies with the type of albinism but is in general reduced [12]. In OCA1A the hair, eyelashes and eyebrows are white, and the skin is white and does not tan. Irises are light blue to almost pink, and fully translucent. Pigment does not develop and amelanotic nevi may be present. The symptoms do not vary with age or race. Visual acuity is 1/10 or less, and photophobia is intense. In OCA1B, the hair and skin may develop some pigment with time (after 1 to 3 years), and blue irises may change to green/brown. Visual acuity is 2/10. This phenotype was previously known as yellow albinism. In OCA2, the amount of cutaneous pigment may vary, and newborn nearly always have pigmented hair. Nevi and ephelids are common. Iris color varies and the pink eyes seen in OCA1A are usually absent. Visual acuity is usually better than in OCA1, and can reach 3/10 [7].

The diagnosis of OCA is based on clinical findings of hypopigmentation of the skin and hair, in addition to the characteristic ocular symptoms. However, due to the clinical overlap between the OCA subtypes, molecular diagnosis is necessary in order to establish the gene defect and thus the OCA subtype. Molecular genetic testing is based on mutational analysis of the genes, by standard screening methods such as denaturing high performance liquid chromatography (DHPLC) or single stranded conformational polymorphism (SSCP), followed by DNA sequencing [7].

The parents of an affected child are obligate carriers, the recurrence risk for another affected child is 25%, and healthy sibs are at 67% risk of being carriers. Offspring of an affected person are obligate carriers. Carriers are asymptomatic. Prenatal diagnosis can be done on DNA extracted from chorion villus sampling (CVS) at 10–12 weeks gestation or on DNA extracted from cultured amniocytes [7]. Prenatal diagnosis has been performed on skin biopsies from the fetus [14].

Skin cancers are generally commoner in the middle aged and elderly. In albinos however these cancers are known to present earlier [15-17]. Most important risk factor for the development of SCC is environmental exposure to ultraviolet light, as evidenced by increased incidence in sunnier climates, lower rates in dark skin & majority arising over sun-exposed skin.

Melanin is a photo protective pigment, protecting the skin from the harmful effects of ultraviolet radiation. Its deficiency in people with albinism predisposes them to the harmful effects of ultraviolet radiation exposure, resulting in issues such as photophobia, decreased visual acuity, extreme sun sensitivity, and skin cancers [18]. High levels of exposure to ultraviolet radiation increase the risk of all three major forms of skin cancer and are responsible for the anatomical site distribution [19]. No use of protection for the skin increased the risk of skin cancer in these patients. Kromberg et al. reported that 23.4% of albinos developed skin cancer out of 111 albinos studied in South Africa [5].

The head and the neck is the site most commonly affected and squamous cell carcinoma has been reported to be the commonest skin malignancy seen in albinos [5,20]. Sqamous cell carcinoma arising from actinic keratoses has been reported from India [21]. In the African albino, the risk of developing these malignancies in comparison to the general population has been reported to be as high as up to 1000 fold [22].

Surgery has been reported to be the mainstay of treatment of the majority of skin cancers in albinos [22,23]. Adequate surgical resection is most important to prevent local recurrence. Good results can be obtained with radical surgery and optimal surgical margins along with reconstructive procedure when needed [24].

From available reports, skin cancers in albinos are preventable [21,22]. There is therefore a need for early institution of skin protective measures in these patients which include protective clothing, sun-screening agents, indoor occupations, and early detection and treatment of skin cancers. For this to be effective: the public should receive education on early institution of preventive measures, register all albinos early in life and educate them to prevent the damaging effect of the sun (protective clothing, sun-screening agents and indoor occupations). Regular examination of all albinos for early detection and treatment of the various malignant lesions to which they are prone deserves to be included in the current anti-cancer campaign, to which the medical world is committed [21]. Dermatologists should maintain a high index of suspicion in this vulnerable population.

Conclusions

Albinism and solar radiation are risk factors for skin cancer. Early implementation of public education strategies on skin cancer prevention is an important long term goal which should improve the outcome in these albinos. Providing free annual skin check up would improve early detection and treatment, hence reducing the morbidity and mortality of skin cancers in these patients.

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MILIA-LIKE IDIOPATHIC CALCINOSIS CUTIS OF THE MEDIAL CANTHUS

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Abstract
Calcinosis cutis is a term used to describe a group of disorders in which calcium deposits form in the skin and may be classified as dystrophic, metastatic, idiopathic or iatrogenic calcification, and calciphylaxis. Idiopathic calcinosis cutis occurs without any underlying tissue damage or metabolic disorder. In this paper, the authors report a new case of idiopathic calcinosis involving the medial canthus of the left eye that was mistaken for milia. An 18-year-old previously healthy male patient, presented with an asymptomatic whitish solitary tumour of the medial canthus of the left eye. The patient had no systemic or trauma history, and the serum levels of calcium and phosphorous were normal. An excisional biopsy was performed and histopathologic examination revealed subepidermal calcinosis. Calcinosis cutis is a rare condition that should be included in the differential diagnosis of a benign-appearing lesion of the face. While it can occur in patients with a history of inflammation, trauma, or hypercalcemia, its etiology can also be idiopathic.

Key words: idiopathic; calcinosis cutis; medial canthus

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Introduction

Calcinosis cutis is a rare disease characterized by the deposition of insoluble calcium salts in cutaneous tissue [1]. Idiopathic calcinosis cutis occurs in the absence of tissue injury or systemic metabolic effect. No causative factor is identifiable and calcification is most commonly localized to one general area. Idiopathic calcification of normal skin has been described mainly in scrotum, penis, vulva and breast but rarely in the face. In this paper, the authors report a new case of idiopathic calcinosis involving the medial canthus of the left eye which was mistaken for milia.

Case Report

An 18-year-old previously healthy male patient, presented with an indolent lesion of the medial canthus of the left eye of one year duration. The patient had no systemic or trauma history. Laboratory data showed no abnormalities. Serum calcium and phosphorous levels were within normal range. On examination, there was a 2mm hard, whitish nodule involving the medial canthus of the left eye with oozing of central whitish material. The suspected clinical diagnosis was milia. The contralateral eye was unremarkable. An excisional biopsy of the nodule was performed. Histopathologic examination, demonstrated the presence of massive amorphous basophilic-stained calcification deposits beneath the epidermis, with occasional foreign body giant cells around the calcific masses and acanthosis of the overlying epithelium (Fig. 1-4). The final pathological diagnosis was idiopathic calcinosis cutis of the medial canthus. The patient has been followed on an outpatient basis without specific findings over 3 months of follow-up.

Discussion

Calcinosis cutis is separated into five subtypes: dystrophic, metastatic, idiopathic, iatrogenic calcification, and calciphylaxis [2]. Dystrophic calcification appears as a result of local tissue damage with normal calcium and phosphate levels in serum [1,2]. Metastatic calcification is characterized by an abnormal calcium and/or phosphate metabolism, leading to the precipitation of calcium in cutaneous and subcutaneous tissue. Skin calcification in iatrogenic calcinosis cutis is a side effect of therapy. Calciphylaxis presents with small vessel calcification mainly affecting blood vessels of the dermis or subcutaneous fat.

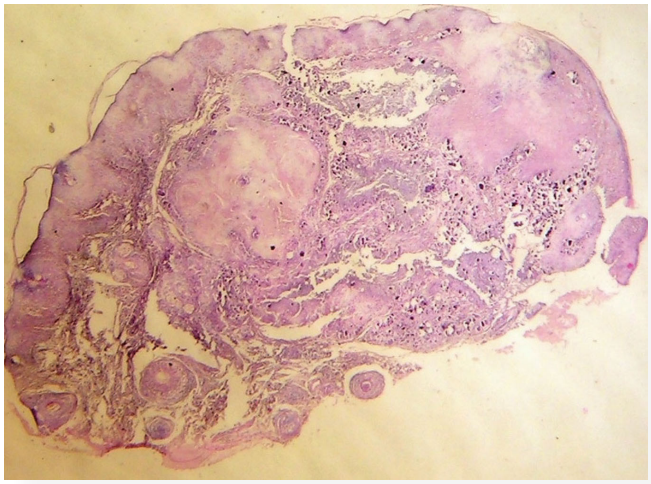

Figure 1. Histological section showing massive amorphous basophilic-stained calcification deposits beneath the epidermis. (H & E, original magnification x 10)

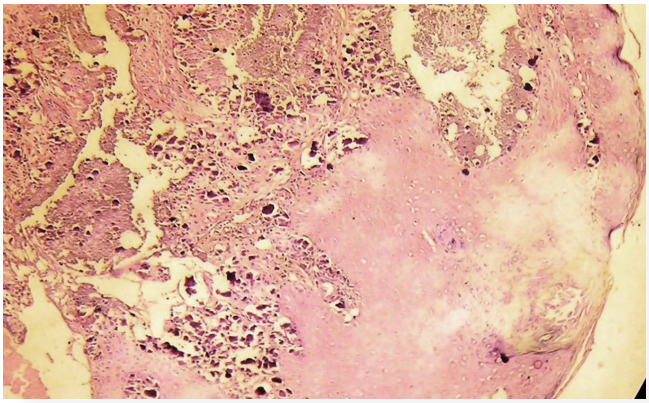

Figure 2. Granular basophilic deposits in the dermis with acanthosis of the overlying epidermis. (H & E, original magnification x 25)

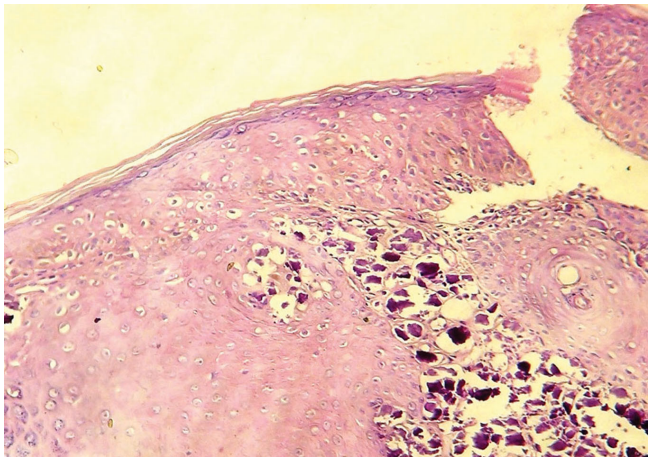

Figure 3. Amorphous basophilic calcification deposits in the papillary dermis. (H & E, original magnification x 40)

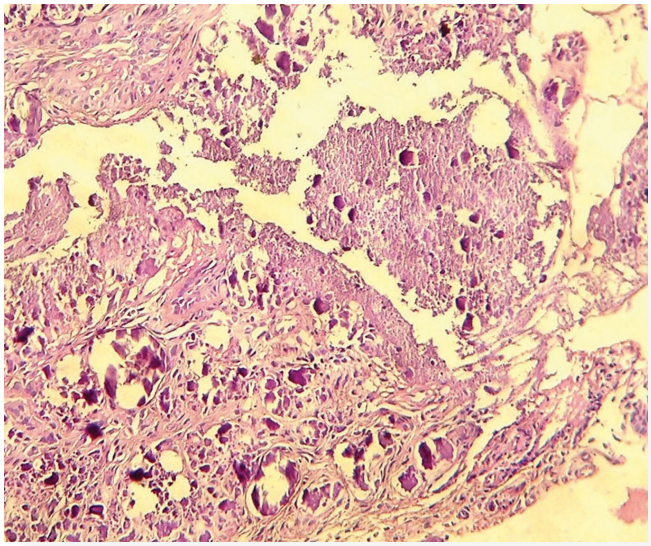

Figure 4. Massive amorphous basophilic-stained calcification deposits. (H & E, original magnification x 40)

Idiopathic calcification occurs without any underlying tissue damage or metabolic disorder. The calcification is most commonly localized to one general area, but a case of unusually widespread calcinosis cutis has been reported [1]. Idiopathic calcinosis cutis comprises tumoral calcinosis, subepidermal calcified nodules, and scrotal calcinosis. Subepidermal calcified nodules occur on the head and extremities, mainly as solitary, hard, white yellowish papules of 3 mm to 11 mm. The disorder usually occurs in children and can even be present at birth [3,4]. Some investigators suggest that they represent calcified adnexal structures [5-7]. Idiopathic calcinosis has also been reported in patients with Down Syndrome in association with syringomas where lesions are found on the hands, forearms, and thighs [8]. In our case, dystrophic, metastatic and iatrogenic calcinosis cutis were ruled out, respectively, by the lack of history of trauma, no preceding pathologic lesions at the sites of the nodular lesion, normal serum calcium and phosphorus levels and absence of history of parenteral therapy. The pathogenic mechanism of idiopathic calcinosis cutis is unknown. Some advocate an active role of the increased sweat duct calcium levels in the development of these lesions [8]. Therefore, the high concentration of sweat glands in the groin and pubic areas may play a role in the development of calcium deposits at these sites [1,8]. Calcinosis cutis has been mistaken clinically for molluscum contagiosum, milia, verruca, and xanthoma [6]. In our case, calcinosis cutis was clinically mistaken for milia. Histopathologically, calcium deposits stain dark blue with hematoxylin eosin and black with von Kossa stain. Fine granules of calcium can be observed in the dermis and large, irregular calcium masses occur in the subcutaneous tissue [9]. A foreign body reaction with inflammation and fibrosis may be seen around larger calcified deposits [9]. Because calcinosis cutis is rare, there is a notable lack of controlled clinical trials on its treatment. The efficacy of calcinosis treatment has only been reported in single cases or small case series. No treatment has been generally accepted as standard therapy, although various treatments have been reported to be beneficial. Small calcified deposits or larger localized lesions can be successfully treated by surgical intervention. Disseminated, extended calcinosis often requires systemic treatment. The effect of surgery can be evaluated within days or weeks, whereas systemic therapy may need months of treatment before improvement is seen [1].

In summary, a case of idiopathic calcinosis of the medial canthus is reported along with pathological findings. Calcinosis cutis is a rare condition that should be included in the differential diagnosis of a benign - appearing lesion of the face. While it can occur in patients with a history of inflammation, trauma, or hypercalcemia, its etiology can also be idiopathic.

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RECURRENT ECCRINE HIDRADENOMA OF THE BREAST IN A MALE PATIENT: PROBLEMS IN DIFFERENTIAL DIAGNOSIS

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Abstract
Introduction: Hidradenoma is an uncommon usually benign tumor of the skin that grows slowly.
Case report: We describe a case of a 39 patient with a breast mass. Physical examination revealed a solitary, well-circumscribed tumor, measuring 1 cm by 0.7 cm. No other skin abnormalities were found. A total surgical excision was performed and histologic examination concluded to an eccrine hidradenoma with clear cells.
Conclusion: Here we discuss problems in the differentiate this tumor, mainly in this not common location, from a breast primary (ductal carcinoma or adenomyoepitelioma), from a metastatic clear cell carcinoma and from other types of skin tumors. Moreover, this patient presented with a recurrence of the tumor in the same location, suggesting a locally aggressive form of this neoplasia; few reports in the literature are described as at low malignant potential, but definite criteria for this diagnosis are not well defined.

Key words: eccrine hidradenoma; breast; clear cell; recurrence

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Maria Orsaria, Laura Mariuzzi: Recurrent eccrine hidradenoma of the breast in a male patient: problems in differential diagnosis. Our Dermatol Online. 2013; 4(2): 215-217.

Introduction

Hidradenoma is a benign adnexal neoplasm, mostly dermal located, historically considered eccrine, but with evidences suggesting also an apocrine differentiation [1]. This neoplasia presents most often in young adults, and appears to be slightly more common in women than in men. Common locations are head, neck and limbs [2]. The histologic appearance put in the differential diagnosis other skin neoplasms and other tumors depending on the location [3-6]. These tumors are usually benign but they can have rarely low malignant potential, and they should be surgically removed with safety margins, because they have a high local recurrence rate and a potential of malignant transformation [7].

Case Report

A 39-year-old man presented with a recurrent nodule of the left outer upper left breast quadrant, superficially located. The lesion was asymptomatic. He reported a previous history of a excised breast mass in the same location. No clinical-pathological report of the prior resection was available. Physical examination revealed a solitary, well-circumscribed tumor, measuring 1 cm by 0.7 cm. No other skin abnormalities were found. The tumor was excised and submitted for histological examination.

Tissues were fixed in buffered formalin, paraffin embedded and routinely processed for histological diagnosis. For immunohistochemistry, the Dako REAL™ EnVision™ Detection System, Peroxidase/DAB+, Rabbit/Mouse Code K5007 method was used. The antisera employed are listed in Table I, together with their source, dilution and antigen retrieval method.

Results

The histopathological result of a needle core biopsy showed a tumor composed of solid sheets of clear cells with an abundant vascularization; the subsequent excisional biopsy (Fig. 1) revealed a lobulated masses in the dermis with focal extension into the subcutaneous fat, without connection with the above skin, composed of two cell types (Fig. 2 a, b): a population of cuboidal cells with eosinophilic cytoplasm and round to oval nucleus with conspicuous nucleolus; elsewhere it consisted of cells with clear cytoplasm containing large glycogen deposits and with a small eccentrically located nucleus. No mitosis were found. Focally, duct-like structures were present, lined by cuboidal cells resulting in perivascular pseudorosettes. The tumor lobules were surrounded by a desmoplastic stroma. No breast ductules were found in proximity of the tumor lobules.

Antibody	Clone, source	Dilution
ER	1D5, DAKO	1:100
PR	636, DAKO	1:100
GCDFP-15	23A3, DAKO	1:40
Mammaglobin	304-1A5, DAKO	1:100
CK19	RCK108, DAKO	1:50
High molecular weight Cytokeratin	34βE12, DAKO	1:50
CK5/6	D5/16B4, DAKO	1:50
Cytokeratin 7	OV-TL 12/30, DAKO	1:100
Vimentin	V9, DAKO	1:300
RCC	SPM314, DAKO	1:50
Calponin	CALP-1, DAKO	1:50
CD10	56C6, DAKO	1:50
P63	4A4, DAKO	1:100
AR	AR441, DAKO	1:100
S100	Polyclonal, DAKO	1:1000

Table I. Antibodies employed for immunohistochemistry

ER Estrogen receptor; PR Progesterone receptor; GCDFP15 Gross cystic disease fluid protein-15; RCC Renal cell carcinoma marker; AR Androgen receptor. DAKO, Glostrup, Denmark

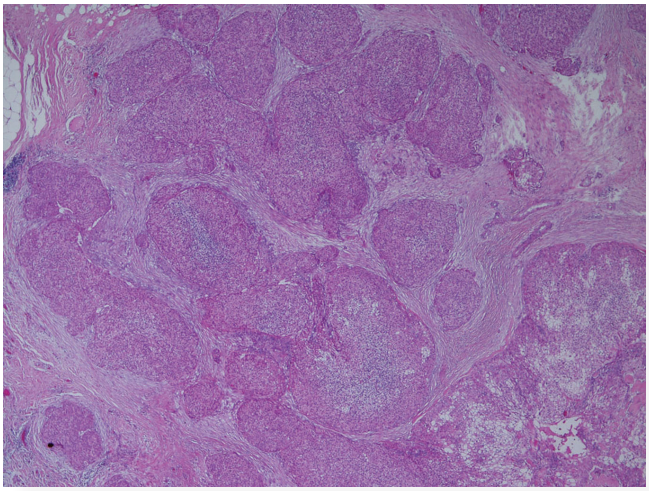

Figure 1. Excisional biopsy showing a lobulated tumor in the dermis with focal extension into the subcutaneous fat, without connection with the above skin (haematoxylin--eosin; original magnification x 20)

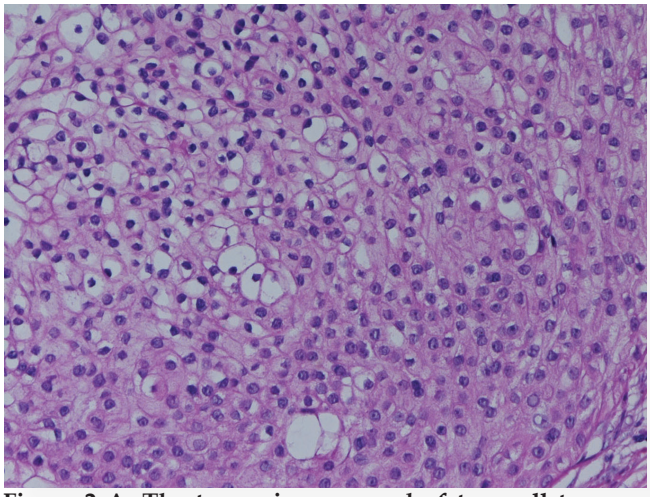

Figure 2 A. The tumor is composed of two cell types: a population of cuboidal cells with eosinophilic cytoplasm and round to oval nucleus with conspicuous nucleolus; elsewhere it consisted of cells with clear cytoplasm containing large glycogen deposits and with a small eccentrically located nucleus (haematoxylin-eosin; original magnification x 200)

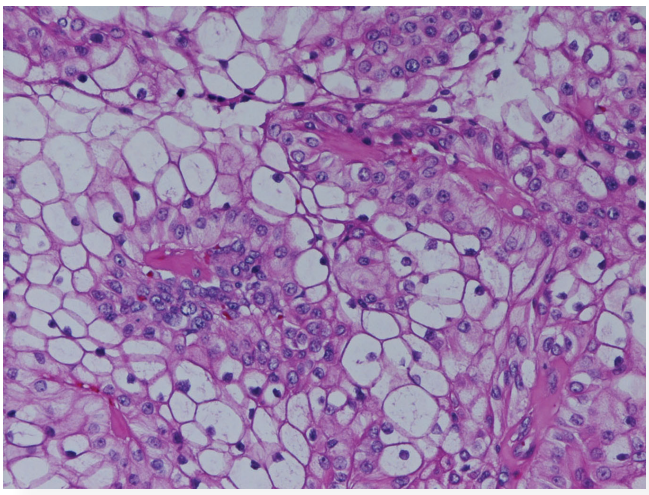

Figure 2 B. Focally, duct-like structures were present, lined by cuboidal cells resulting in perivascular pseudorosettes (haematoxylin-eosin; original magnification x 200)

Discussion

The breast location of this tumor and its particular histological findings include in the differential diagnosis a primary breast ductal carcinoma [3], a metastatic clear cell carcinoma [4], an adenomyoepithelioma [5] and primary skin tumors with follicular differentiation, sebaceous differentiation, or sweat gland differentiation [6]. Against a diagnosis of a primary breast ductal carcinoma, there weren't ductular structures around the tumor, cytological atypia was lacking and the neoplastic proliferation had a biphasic cellular population [3]; moreover, mammaglobin, GCDFP15, ER and PR immunohistochemical staining were negative, whereas in a well differentiated breast ductal carcinoma they would expect to be positive. A diagnosis of a metastatic clear cell carcinoma, mainly a renal cell carcinoma, was considered, but immunohistochemistry revealed that the neoplastic cells were positive for CK7, CK19, CK34βE12 and negative for CD10, vimentin and RCC (renal cell carcinoma). A diagnosis of adenomyoepitelioma was ruled out by the negative immunostaining for myoepithelial antibodies (calponin, CD10 and S100). The tumor showed only positivity for CK5/6, p63, PAS and negativity for PAS-D; even if these markers are quite nonspecific they were not in contrast with a diagnosis of eccrine hidradenoma; the morphology of the tumor along with the negativity for androgen receptor excluded a sebocytic differentiation; moreover, the tumor had no connection with the above skin, excluding a tumor with follicular differentiation.

The clinical and radiological suspicious in this case was of a primitive breast carcinoma; the difficulty in the diagnosis of this tumor was to differentiate if the neoplasia was a skin primary or a breast one, in particular a clear cell hidradenoma, a very rare breast tumor that share the histological features of sweat gland tumors with only 18 cases reported in the literature [5], but the lacking of normal breast ductules around the tumor lobules let us to think that this case originated from the skin.

Eccrine hidradenoma is an usually benign, slowly growing, asymptomatic, solid or cystic sweat gland tumour that occurs on the head, neck and limbs; the breast location is unusual, even if cases located in the trunk and in the breast were reported [5,7], and the male gender is less common [8]. When a tumor with features as ours occurs in the breast it is worthwhile to keep in mind in the differential diagnosis a skin eccrine hidradenoma, the diagnostic clue is the lobulated architecture with the two-cell pattern of proliferation, composed of polygonal cells with distinct cell border and clear cytoplasm, and dark cuboidal cells lining the duct structure [5].

Eccrine hidradenoma shows rarely low malignant potential and the histopathologic criteria of an aggressive behavior are not well defined [9-11]. There are few reports in the literature highlighting that the "malignant" form of this tumor is very rare; all these cases were characterized by a significant rate of locoregional recurrence and some patients developed distant metastatic spread [12]. Hidradenoma with malignant potential is usually found in the scalp, face or anterior surface of the trunk.

Malignant clear cell hidradenoma usually develops de novo, not arising from a benign form, and invades the dermis and subcutaneous tissue; it might share significant histopathological features with its benign form. The mitotic index may not be representative and helpful in the differential diagnosis, that often it's impossible to make. Therefore, the diagnosis of malignancy through standard pathological examination may be extremely difficult [9].

This patient had a history of prior resection of a neoplasia in the same location of the left breast, probably the same lesion that recurred; therefore, this would qualify this tumor as having a possible malignant biological potential and a wide re-excision and careful follow-up is therefore advisable for these worrisome lesions that show an increased risk of recurrence [11].

Conclusions

It is important to consider eccrine hidradenoma with clear cells as a rare differential diagnosis of cutaneous tumors and, if it arise in the breast region, of primary breast carcinoma with histologic features of sweat gland tumors. Moreover, it is clear, even if it is a very rare event, that this entity could have an aggressive clinical behavior with local recurrences and also metastatic disease in the regional lymphatics, therefore in these "malignant" cases a wide excision should be warranted.

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CUTANEOUS NODULE ON THE FACE: ADAMANTINOID TRICHOBLASTOMA - A RARE, UNIQUE TUMOR

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Abstract
Adamantinoid trichoblastoma is a rare adnexal tumor which clinically masquerades as various benign and malignant lesions. Less than 50 cases have been documented so far. In this report we discuss the clinicopathological features of this rare and fascinating tumor with a brief review of literature.

Key words: cutaneous lymphadenoma; adamantinoid trichoblastoma; tumor infiltrating lymphocytes

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Lakshmi Rao, Vidya Monappa, Mohammed Musheb: Cutaneous nodule on the face: Adamantinoid trichoblastoma - a rare, unique tumor. Our Dermatol Online. 2013; 4(2): 218-220.

Introduction

Adamantinoid trichoblastoma (AT) is an uncommon benign skin adnexal (follicular) neoplasm with a prominent lymphocytic infiltrate and an adamantinoid appearance (similar to dental adamantinoma). It was originally described as "lymphoepithelial tumor of the skin" by Santa Cruz and Barr in 1987 and was later renamed as "cutaneous lymphadenoma" (CL) in 1991 [1]. Currently many authors believe it to be a variant of trichoblastoma, a benign follicular tumor with both epithelial and mesenchymal components. It is a rare tumor with fewer than 50 cases reported in the world literature.

Case Report

A 42 year old lady presented with a firm, nodular, slowly progressive swelling in the face of 1 year duration. Clinically it had the appearance of a keloid /neurofibroma. Surgical excision of the lesion was performed and received in our laboratory for histopathological study.

Pathological findings:
Grossly, the excised skin covered tissue measured 1.3 x 0.8 x 0.5 cm and showed grey white areas on cut section. Sections showed thinned out epidermis overlying an unencapsulated, well circumscribed dermal tumor composed of irregular islands of epithelial nests with palisading basaloid cells at the periphery. Central areas within the islands showed large polygonal cells with clear cytoplasm with a dense lymphocytic infiltrate, edema, histiocytes, giant cells, focal ductal and follicular differentiation along with Reed Sternberg like cells. Surrounding stroma showed dense fibrosis with lymphocytic infiltrates (Fig. 1-4).
Histopathological diagnosis of *Nodular Adamantinoid trichoblastoma (Cutaneous lymphadenoma)* was made.

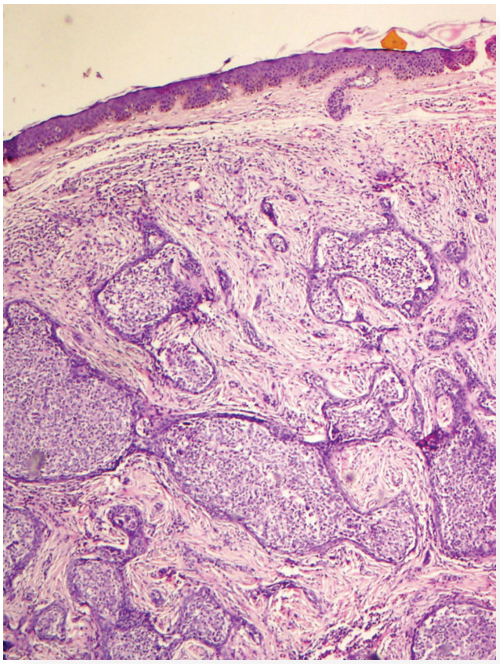

Figure 1. Section shows thinned out epidermis overlying a tumor composed of irregular epithelial nests with peripheral palisading basaloid cells. H&E, 100X

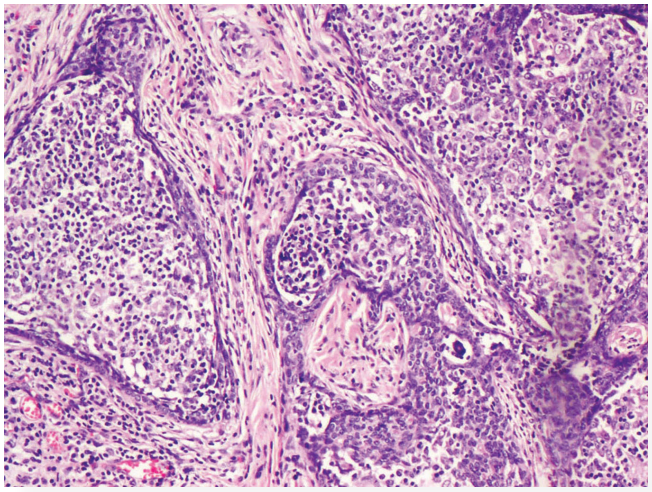

Figure 2. The epithelial nests showing lymphocytic infiltrates surrounded by desmoplastic stroma. H&E, 200X

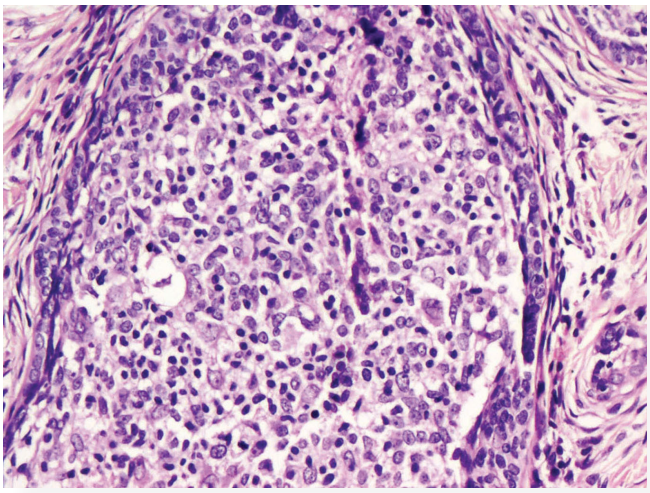

Figure 3. Higher power view of the epithelial islands showing cells with clear cytoplasm, lymphocytes and histiocytes. H&E 400X

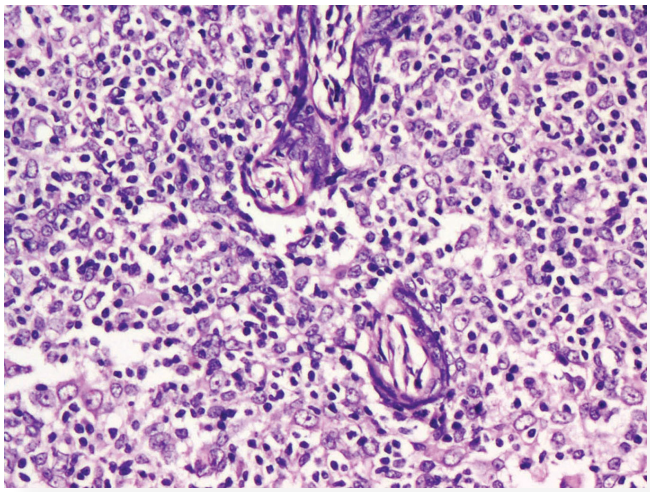

Figure 4. Higher power view of the epithelial nests shows scattered large Reed Sternberg- like cells. H&E, 400X

Discussion

AT/CL is an uncommon skin adnexal tumor usually presenting as an asymptomatic, small, dome-shaped, flesh-colored papule or nodule, typically in the head and neck region. They are commonly seen in young to middle aged adults with a predominance of male patients. Unlike other skin adnexal tumors, the description of these tumors is limited to random case reports [2]. Less than 50 such cases have been reported in the world literature.

Possibly because of their rarity, ATs are generally misinterpreted clinically as basal cell carcinoma, adnexal tumor, nevus or dermatofibroma [2]. The present case was clinically thought to be a keloid/neurofibroma.

Microscopic examination demonstrates a triphasic tumor composed of cell nests with palisading basaloid cells at the periphery (epithelial component), lymphoid infiltrate and desmoplastic stroma (mesenchymal component). There can also be large cells that resemble Reed Sternberg cells within the lobules. Focal ductal, follicular and sebaceous differentiation, central keratinization and stromal mucinosis have been described [3]. The tumor is unencapsulated and generally well circumscribed, but may have infiltrating outlines making way for tumor recurrence [2].

The histological differential diagnosis includes - clear cell Basal cell carcinoma, clear cell syringoma, trichoepithelioma and malignant lymphoepithelioma - like carcinoma. However, a lymphoid cell infiltrate within the tumor lobules is not prominent in such tumors [4]. In younger patients it has to be differentiated from dermal thymus, which is an aberrant location of thymic tissue in the skin, due to defective migration, and presents as linear symmetric ulcerated scar-like lesions in the neck in patients with other facial - branchial abnormalities [2].

Several proposals have been made regarding the possible histogenesis of this tumor. Santa Cruz et al and Santa Cruz and Barr originally suggested immature pilosebaceous differentiation [5,6]. Others proposed an eccrine origin suggesting the term “lymphotrophic solid syringoma” and “lymphotrophic eccrine benign tumor” [7,8]. Dahill and Seywright [9] reported a case of synchronous occurrence of cutaneous lymphadenoma and syringoid eccrine carcinoma in a single patient providing additional evidence to support eccrine differentiation. Fillipo et al [10], argued that AT/CL is not a distinct entity but may represent a basal cell carcinoma with possibly pilar or eccrine differentiation. Mc Niff et al [11], evaluated the immunohistochemical staining patterns of CK20 (merkel cells), bcl2 (epithelial), S100 and CD1a (langerhans cells) and CD34 (stromal cells) in CL, trichoblastomas and nodular basal cell carcinomas. They observed similar staining patterns for the first three markers in CL and trichoblastomas (importantly peripheral staining of bcl2 in tumor lobules) and opined that CLs are in-fact a variant of trichoblastomas. Most recently, these tumors have been classified as an adamantinoid variant of trichoblastoma, considering its resemblance to dental adamantinoma, a benign epithelial and mesenchymal tumor of oral cavity [2,12,13]. The marked lymphocytic infiltrate in these tumors is thought to be a result of either defective lymphocyte–epithelial interaction or an exuberant host response to the tumor cells [14,15].

A benign clinical course has been described in the literature for these tumors [1-15]. Surgical excision of the tumor is the definitive treatment. However the tendency to local infiltration may result in tumor recurrence in cases of incomplete removal. To conclude, AT is a rare benign adnexal tumor which can clinically masquerade as a variety of benign and malignant lesions.

Histologically, it is a triphasic tumor with epithelial islands, lymphocytic infiltrates and desmoplastic stroma. AT has been called by different names in the past which include, lymphoepithelial tumor of the skin, lymphotrophic solid syringoma, lymphotrophic eccrine benign tumor and cutaneous lymphadenoma. The term AT describes the close relationship of this tumor with trichoblastoma and its peculiar admantinoid appearance in tissue sections, but it does overlook the prominent intra-tumoral lymphocytic infiltrate, which is a characteristic feature of this tumor. The authors thus feel that 'Lymphotropic AT' would be a better terminology to describe this rare tumor.

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NASZA DERMATOLOGIA Online
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SYRINGOCYSTADENOCARCINOMA PAPILLIFERUM: A CASE REPORT OF A RARE SKIN ADNEXAL TUMOUR

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Abstract
Syringocystadenocarcinoma papilliferum (SCACP), a rare skin adnexal carcinoma with apocrine differentiation is the malignant counterpart of syringocystadenoma papilliferum (SCAAP). It usually occurs in the head and neck region of elderly individuals. We describe a 46 year old south Indian female with a lesion in the scalp. Morphologically the tumour had the characteristic features of SCAAP along with frank invasion into deep dermis and malignant cytologic features. Immunohistochemically, the tumour cells stained strongly with cytokeratin (CK) 7, carcinoembryonic antigen (CEA), gross cystic disease fluid protein-15 (GCDFP-15), We made a diagnosis of SCACP in the patient, and a wide excision with skin grafting was performed on the patient.

Key words: syringocystadenocarcinoma papilliferum; adnexal neoplasm; apocrine differentiation

Cite this article:
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Introduction

Syringocystadenocarcinoma papilliferum (SCACP) is a rare skin adnexal carcinoma, considered to be a malignant counterpart of syringocystadenoma papilliferum (SCAAP) by the World Health Organization [1,2]. Adnexal tumours are skin tumours whose differentiation is towards one or more of the cutaneous adnexal structures (apocrine, follicular, and sebaceous) of the skin. SCACP is a malignant adnexal neoplasm that shows predominantly apocrine differentiation [3]. It is an extremely rare cutaneous neoplasm with only 13 reported cases, to our knowledge until 2012, since it was first reported in 1980 [2,4]. Most of the SCACP's is preceded by long standing SCAAP. The malignant tumour has a predilection for the head and neck region and commonly affects older people in the 5th to 7th decade [3,4].
To our knowledge, only one case of SCACP has been reported from India [4]. Herein we report a case of SCACP occurring on the right scalp of 46 year old South Indian female patient.

Case Report

A 48 year old south Indian woman presented with an ulcerative growth over the scalp for duration of 1 year (Fig. 1). Since the clinical diagnosis was in favour of squamous cell carcinoma, a needle core biopsy was performed.
Histopathologic examination revealed cystic invaginations and papillomatous downgrowths extending up to the deep dermis. The cystic invaginations and papillomatous downgrowths were lined by luminal high columnar cells, with few showing decapitation secretion and outer cuboidal cells (Fig. 2a). The cells show moderate to severe atypia with increased mitotic activity (Fig. 2b). There was a dense infiltration of abundant plasma cells and lymphocytes in the stroma of the tumour. In few loci, frank invasion of the stroma by the tumour was seen. Based on these histopathological findings, the diagnosis of SCACP was made.
Wide excisional surgery with split skin graft was performed to remove the tumour. There were no signs of lymph node involvement ultrasonographically and clinically. The patient had no clinical or radiographic evidence of any primary tumour elsewhere.
Gross pathology of the whole specimen revealed a skin ellipse with a large asymmetric ulcerative growth measuring 6 * 5 * 2 cms. The growth was poorly circumscribed and seen extending up to the dermis. The histopathological examination of the whole growth revealed the same, with stromal invasion being prominent.

Immunohistochemically, the tumour cells stained strongly with cytokeratin (CK) 7, carcinoembryonic antigen (CEA), gross cystic disease fluid protein-15 (GCDFP-15), which further provided support to the apocrine nature of the neoplasm (Fig. 3a, b, c). The margins were found to be free of tumour.

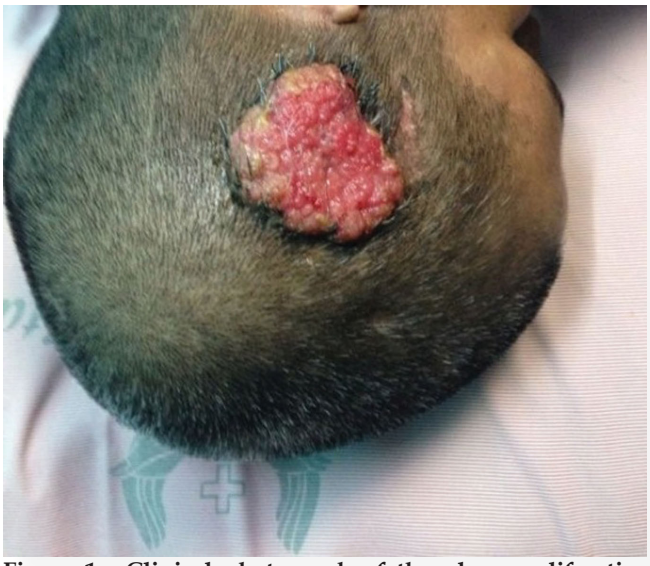

Figure 1. Clinical photograph of the ulceroproliferative growth over the right scalp

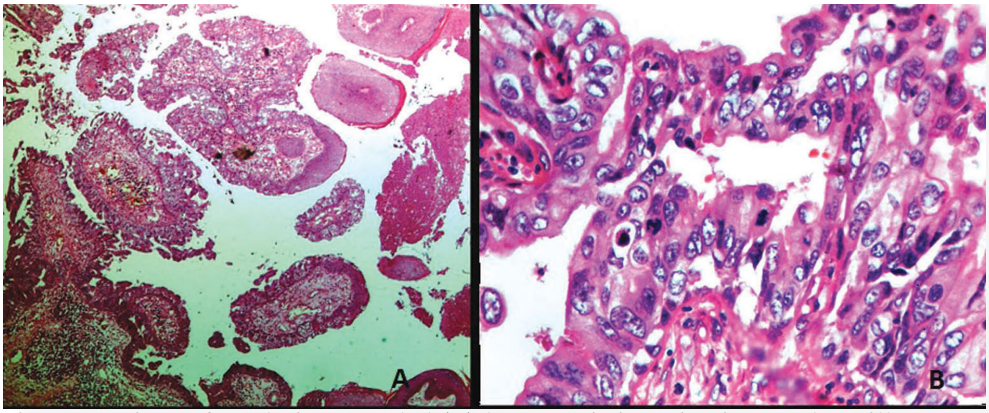

Figure 2. Biopsy from lesion revealed (A) large cystic invaginations and papillomatous downgrowths lined by inner columnar and outer cuboidal cells (Haematoxylin and Eosin X 10); (B) tumour cells with moderate to severe atypia with increased mitotic activity (Haematoxylin and Eosin X 40)

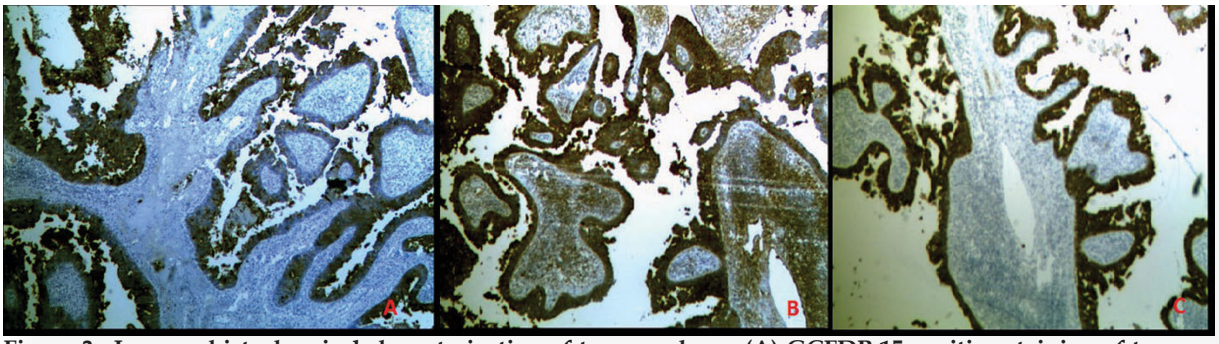

Figure 3. Immunohistochemical characterization of tumour shows (A) GCFDP-15 positive staining of tumour cells; (B) CEA positive staining of tumour cells; (C) CK7 positive staining of tumour cells

Discussion

SCACP is one of the cutaneous adnexal neoplasm, and is a rare neoplasm showing apocrine differentiation [3]. The apocrine histiogenesis is supported by decapitation of the luminal surface of tall columnar cells, continuity of the tumour to pilo-sebaceous units and presence of apocrine glands in the underlying tissue [3]. It is considered to be a malignant counterpart of the SCAAP. Clinically, the long standing lesion suddenly begins to enlarge in size with bleeding, crusting and ulceration. It is most commonly seen in the head and neck region of elderly individual with no gender predilection [1,3].

Morphologically, SCACP resembles SCAAP with cystic papillomatous invaginations connected to the skin surface by funnel shaped structures lined by infundibular epithelium. The upper part of the cystic invaginations are lined by keratinizing squamous epithelium while the lower part and papillary projections are lined by inner columnar cells with decapitation and outer cuboidal cells. This epithelial transition from keratinizing squamous epithelium to glandular lining recapitulates the physiologic relationship of the apocrine gland to the hair follicle. In healthy skin, apocrine gland arises at the follicular infundibulum, characterized by a gradual transition from stratified squamous epithelium at the skin surface to the bi-layered ductal structures in the dermis [1,3]. Stroma of the tumour contains a dense inflammatory infiltrate of plasma cells and lymphocytes mirroring the attraction of plasma cells by glands of the normal secretory immune system [5]. SCACP differs from SCAAP and SCAAP IN SITU by infiltration of tumour cells into deep dermis or subcutaneous fat. SCACP and SCAAP IN SITU differ from SCAAP by cytological features of tumour cells characterized by higher nuclear cytoplasmic ratio, nuclear irregularity, coarse chromatin and increased mitotic activity [1,3,6]. Though there is no definitive immunohistochemical profile for a SCACP, the strong expression of CK7, CEA and GCFDP-15 supports the apocrine differentiation of the neoplasm [1]. It is well established that SCAAP shows positivity to CEA, CK7 and epithelial membrane antigen (EMA) in the luminal cells. However only 2 out of the 4 SCACP's on which GCFDP-15 expression was evaluated, showed positive expression. Though there is not many published literature on the immunohistochemical characterization of SCACP, it is assumed that SCACP mirrors the immunohistochemical profile of SCAAP [7].

Our case was identified as SCACP since it showed the characteristic histology along with infiltration into deep dermis and presence of malignant cells. Also, the tumour strongly expressed apocrine differentiation markers CK7, CEA and GCFDP-15.

Though a rare neoplasm, the entity should be recognized correctly as it may affect patient treatment and prognosis. Similar to other rare adnexal tumours, the literature reveals a good prognosis for SCACP patients treated only by surgical excision [8-10].

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Department of Histopathology, SRL Mumbai for performing the immunohistochemical characterization of the tumour.

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A COLUMELLAR DEFORMITY CAUSED BY A CONGENITAL SCHWANNOMA

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Abstract
Schwannoma is a benign neoplasm arising from Schwann cells of the peripheral nerve. It very rarely located in the nasal tip. We report two cases of congenital schwannoma of the nasal columella and discuss the surgical approach of such tumor.
Case report: Case 1: An 18 year-old female was referred to us for a very slow growing tumor of the columella. The deformity had been present since the birth. The patient underwent an excision of its tumor using an open rhinoplasty approach. The histological examination revealed a schwannoma. No recurrence was found within 2 years of follow up.
Case 2: A 4 month male baby presenting a congenital tumor of his columella. He underwent an excision using open rhinoplasty approach. The histological examination showed a plexiform schwannoma.
Discussion: Schwannoma of the nasal tip is a benign tumor that gradually causes aesthetic and functional disorders. Congenital schwannoma of columella is an extremely rare clinical situation. Its diagnosis and treatment can pose certain challenges. The treatment is surgical excision and histological analysis of the specimen. Open rhinoplasty approach provided a good surgical exploration and a good cosmetic result on this nasal tip tumor.

Key words: nose neoplasms; neurilemmoma; rhinoplasty

Cite this article:
Mohamed El Bouihi, Saad Lahmiti, Souad Aimadeddine, Ahmed Zaroual, Saad Fawzi, N. Mansouri Hattab: A columellar deformity caused by a congenital schwannoma. Our Dermatol Online. 2013; 4(2): 224-225.

Introduction

Schwannoma is very rare in the nasal tip [1] and to the best of our knowledge congenital schwannoma of the nasal columella has never been reported. The diagnosis of such rare tumor is challenging and even if it is a benign neoplasm excision biopsy is essential to exclude sinister pathology [2]. We report a case of columellar schannoma.

Case Report

Case 1:

A 18 year-old female, came to our service for a slow growing deformity of her columellar. The deformity had been present from the birth (Fig. 1).
Physical examination revealed a well-defined elastic tumor in of the anterior part of the columella measuring 1cm. The overlying skin was thin.
The patient underwent an excision of its tumor using an open rhinoplasty approach and a "V" shaped incision (Fig. 2). A superficial subcutaneous undermining allowed tumor removal. The medial crus of greater alar cartilage were slightly distorted. No nerve of origin was seen during the disssection. The limits of the excision were free of tumor, and histologic analysis of the tumor showed the characteristics of a schwannoma. The patient remains having no recurrence within 2 years of postoperative follow up.

Case 2:

A 4 month-old baby was referred to us for a congenital rapidly growing tumor of the columella (Fig. 3). On examination he presents an oval firm, mobile, globular swelling of the columella measuring 15 mm by 5 mm. The tumor had extended to the upper part of the philtrum and had caused a partial obstruction of the right nostril. Excision of the tumor was performed using an open rhinoplasty approach. Pathological examination revealed the diagnosis of plexifom schwannoma.

Discussion

Developmental midline nasal masses in children are rare, with a reported annual incidence of one in every 20,000–40,000 live births [2]. They result from a failure of embryologic separation of neuroectodermal and ectodermal tissues during the development of the nose and frontobasal region. Many such lesions may include an intracranial extension or connection. Their differential diagnosis includes abscesses, hemangiomas, fibromas, lipomas, granulomas, and mucoceles.

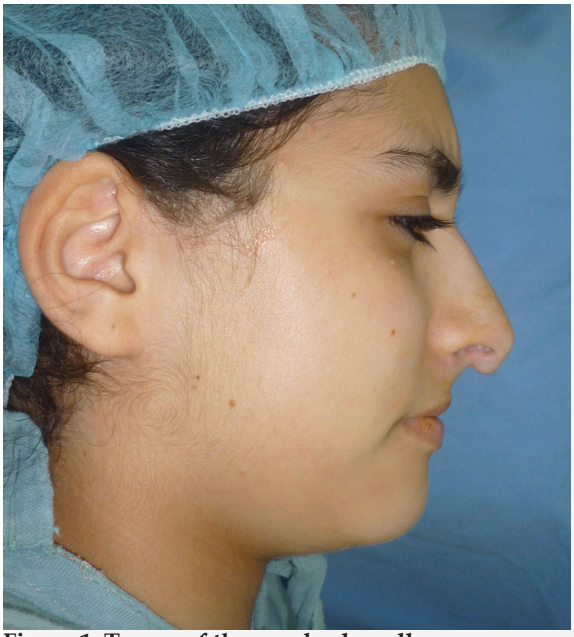

Figure 1. Tumor of the nasal columella

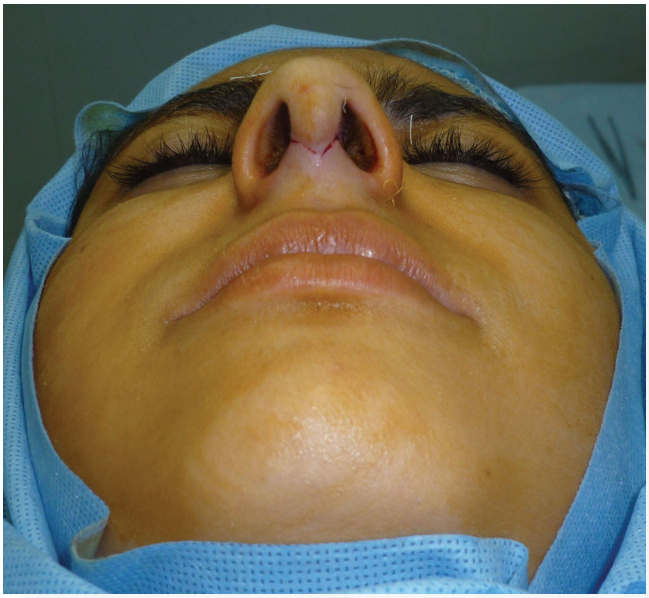

Figure 2. A "V" incision and open rhinoplasty approach to excise the tumor

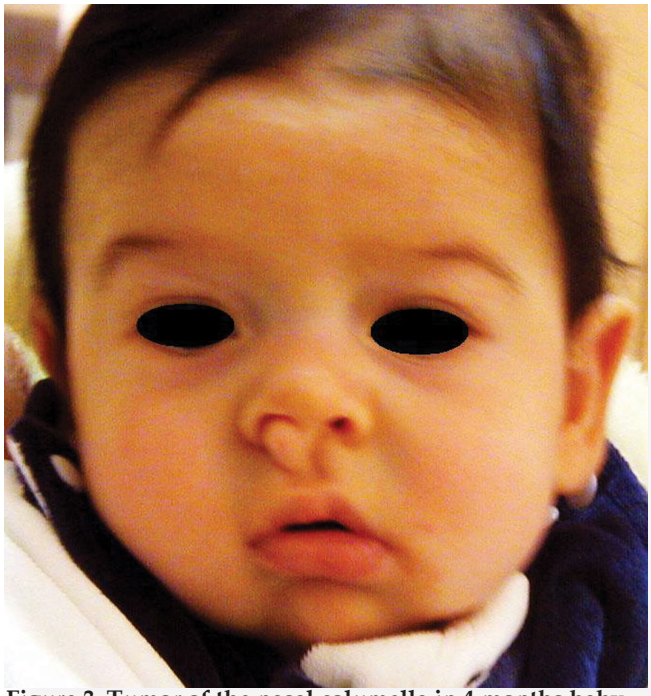

Figure 3. Tumor of the nasal columella in 4 months baby.

Schwannomas are slightly less common than neurofibromas, but, like the latter, constitute about 5% of all benign soft-tissue tumors. Nasal tip location of schwannoma was first described by Bingham et al [3]. Seven cases of nasal tip Schwannoma have been reported in the literature and none of involving the only columella.

Schwannoma is a common tumor that can develop at any age. It grows slowly, expanding and applying pressure to the surronding tissues, causing aesthtic and sensory problems [4]. Even if nasal tip schwannoma is a benign tumor it should be removed as soon as possible to avoid cartilage deformities. Radiologic examinations can assist in its diagnosis [5]. Magnetic resonance imaging (MRI) is especially useful for the diagnosis and can helps also to evaluate the local effect of the tumor on the surrounding structures.

The open rhinoplasty approach is ideal for excision of the tumour in the nasal tip [6-8]. Schwannomas have a true capsule composed of epineurium, which allows their successful surgical resection [9]. The increased exposure makes it easier to perform certain technical manoeuvres for resection, and gives the surgeon the ability to diagnose the deformity of the osseocartilaginous framework.

Recurring tumors are rare (2%) when removal is complete, and are particularly associated with neurofibromatosis [10].

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NASZA DERMATOLOGIA Online
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HAND, FOOT AND MOUTH DISEASE IN NORTHEASTERN PART OF ROMANIA IN 2012

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Abstract
Hand-foot-mouth disease (HFMD) is an acute viral infection that occurs usually among children in summer. This paper reports a high incidence of HFMD in children and adults, occurred in summer-autumn 2012 in the northeastern part of Romania. We present a few cases with some atypical clinical manifestations.

Key words: Enteroviruses; Hand, Foot and Mouth disease; Romania

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Introduction

Hand-foot-mouth disease (HFMD) is an acute viral infection that occurs usually among children in summer [1]. The diagnosis is based on clinical manifestations of oval vesicles on the hands and feet and painful oral mucosa ulcerations. It is a self-healing disease in great majority of cases, but sometimes with severe evolution and neurological complications [2].
Pediatricians and dermatologists should be aware of atypical manifestations.

Case Report

Case 1.

We were asked to see a 3 year-old boy hospitalised in Pediatric Department, with very high fever, nausea, diarhhea, abdominal pains, malaise, who was diagnosed with acute pharyngitis, with no good clinical evolution under antibiotics Claritromicin orally). It was the first days of June 2012 and among the first cases of HFMD (Fig. 1, 2).

Case 2.

A female student of 23 years old, was incapable of walking for many days, by a lof of small vesicles and erythema on the feet; she has also high fever, malaise, diarhhea and headache. In the fifth day she developed also a few vesicles on the palms, no oro-pharyngeal involvment (Fig. 3).

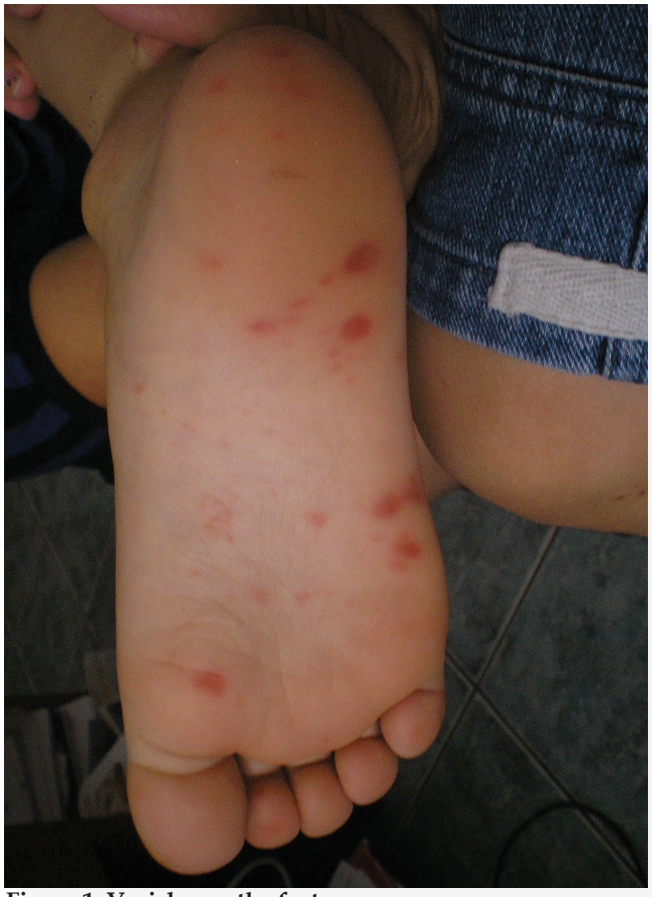

Figure 1. Vesicles on the feet

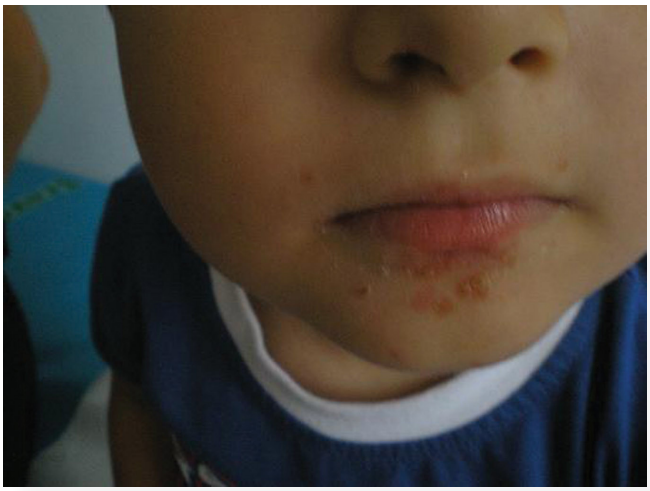

Figure 2. Vesicles and crusts around the mouth

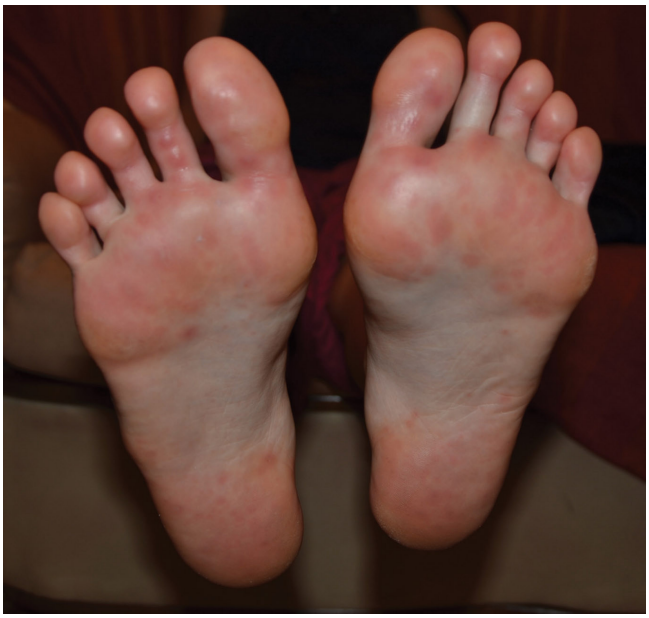

Figure 3. Slight erythema, small papules on the feet

Case 3.

A case of HFMD diagnosed in a child by the classical clinical aspect, but who also presented small papules and vesicles and intense pruritus over the elbows (Fig. 4 a-d).

Case 4.

A 32-year-old female patient was referred to our department because of round vesicles on the hands, slight erythema and pruritus, headache and fever (38,5 Celsius degrees). She was in good general state, no systemic complaints, no complications and the usual laboratory parameters were within normal limits. The fever lasted for 24 hours, no new vesicles were noted and a full recovery was obtained in the next 72 hours (Fig. 5).

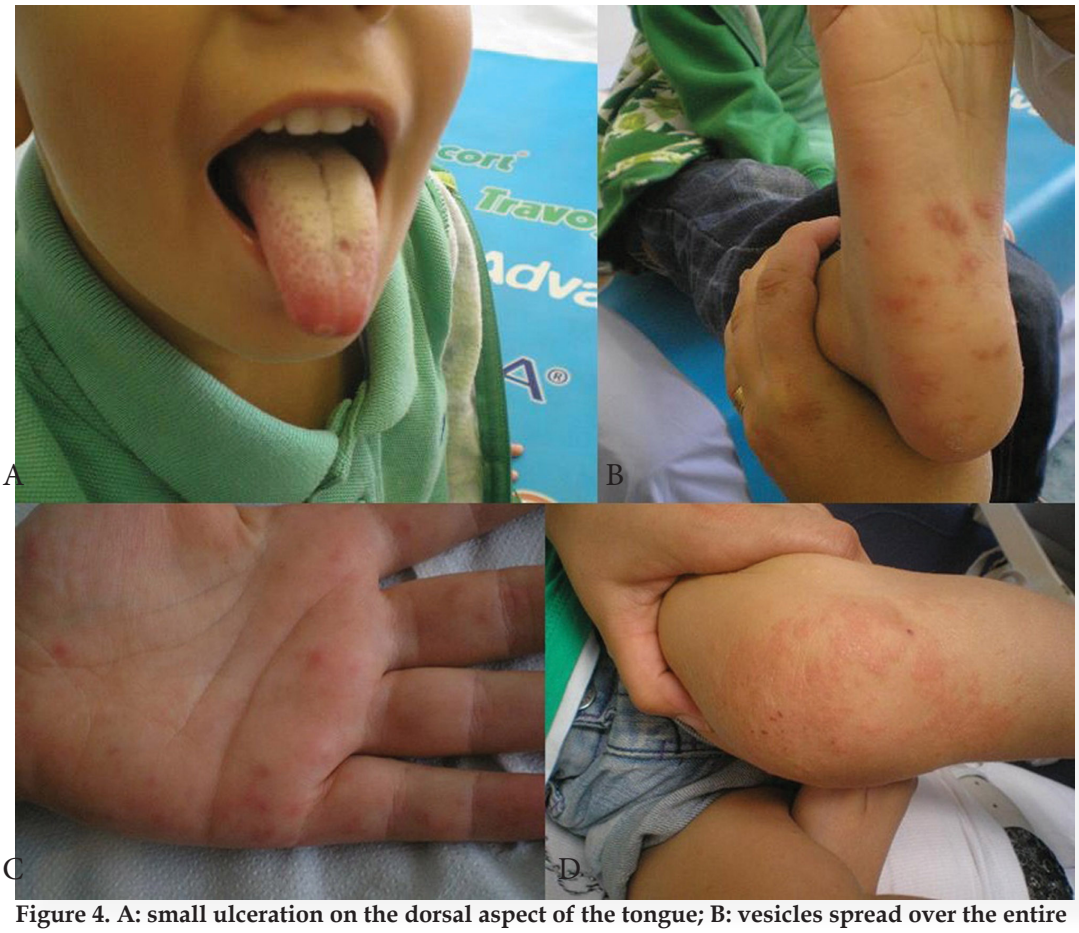

Figure 4. A: small ulceration on the dorsal aspect of the tongue; B: vesicles spread over the entire plantar face of the feet; C: small erythematous vesicles and papules on the left hand; D: cluster of papul-vesicles around the elbow, excoriations due to pruritus

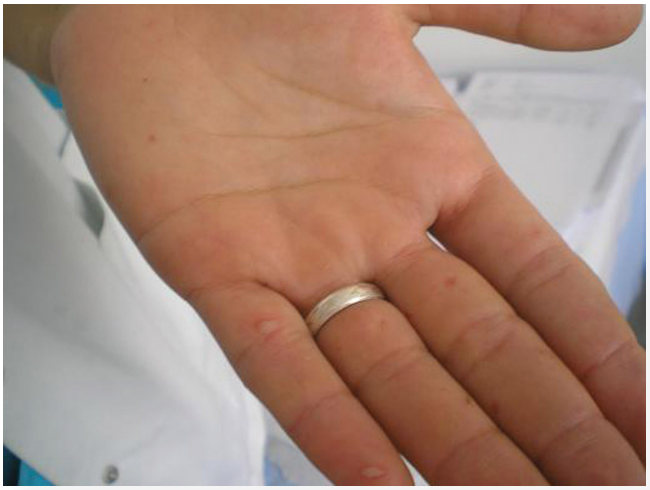

Figure 5. Round vesicles on the left hand

Case 5.

A 5 year old boy was adressed to us for an erythematous papulo-vesicle eruption distributed in the ano-genital area, on the limbs and a few papules scattered on the trunck; no oral manifestations, no systemic complaints, no fever but a long-lasting evolution (almost two weeks with full recovery) (Fig. 6 a-d).

Discussion

Enteroviruses are single-stranded, positive-sense RNA viruses in the Picornaviridae family. There are more than 100 human enterovirus serotypes, including 3 poliovirus serotypes, 23 coxsackievirus A (CA) serotypes, 6 coxsackievirus B (CB) serotypes, 31 echovirus serotypes, and 39 numbered enterovirus serotypes (EV68-71, EV73-102, EV104-107, and EV109).

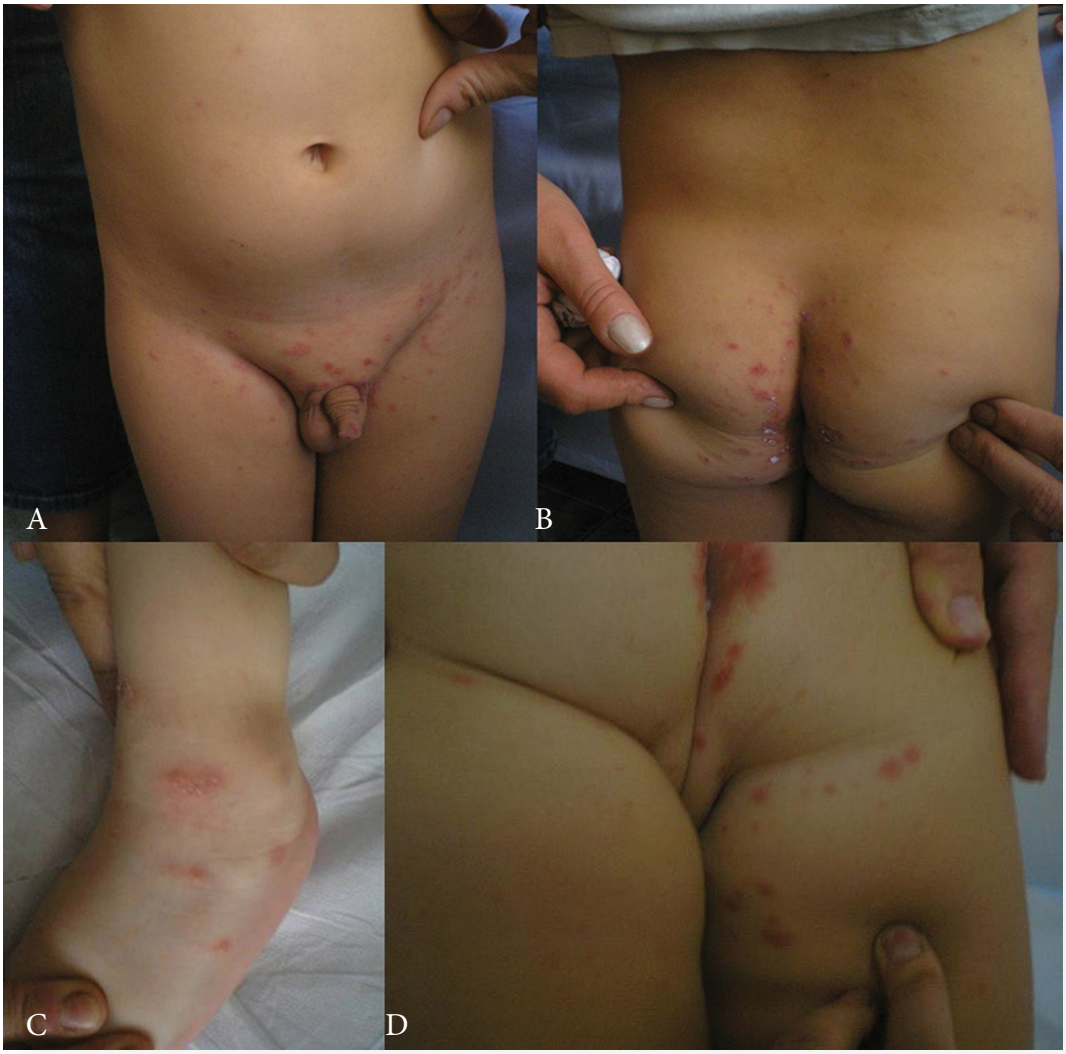

Figure 6. A: vesicles distributed on the genital area; B: perianal lesions; C: grouped vesicles on the dorsal face of left foot; D: the perianal lesions observed 48 hours after the admission

Human enterovirus 71 belongs to the Human enterovirus A species of the genus Enterovirus of the family Picornaviridae and is a major causative agent of hand, foot and mouth disease (HFMD) (usually in children aged <5 years). EV71 was first isolated in 1969.

Coxsackievirus A16 (CA16) was the first viral agent isolated from patients with HFMD. Later CA4, CA5, CA6, CA9, and CA10 as well as Coxsackievirus B (CB) were also found as etiologic agents for HFMD [3]. Recently, EV71 caused life-threatening outbreaks of hand-foot-mouth disease (HFMD) with neurological complications in Asian children (aseptic meningitis, acute flaccid paralysis and encephalitis) 1 and for this reason development of EV71 vaccines is a national priority in some Asian countries (Japan, China) [3,4].

Outbreaks have occurred recently in the Asia-Pacific region: Malaysia (2000-2003), Taiwan (1998-2005), Singapore (2000), Brunei (2006), Thailand (2008-2009), Korea (2008-2009), and Hong Kong (2008) [5].

The viruses implicate in HMFD spread by fecal-oral and respiratory routes,the contamination of other family members commonly occurs and the reinfection within the same family was recently described.

There is usually a prodrome consisting of low-grade or even high fever (especially in small children), anorexia, sore mouth, and malaise. Children younger than 5 years are most commonly affected. Oral lesions occur chiefly on the anterior buccal mucosa and tongue, where the vesicles transform rapidly in superficial ulcers with erythematous bordersand sometimes very painful. The lesions on the palms and soles are papules or vesicles on a surrounding zone of erythema. Less commonly, the dorsal or lateral surfaces of the hands and feet may also been affected [6]. Involvement of the buttocks is common in small children. The eruptions are nonpruritic and usually resolve without crusting.

Onycomadesis is considered a more severe form of Beau lines. The definition of onycomadesis is: the separation of nail plate from nail bed starting at the proximal end and resulting in shedding of the nail. Onycomadesis is a silent sign of HFMD that can appear a few weeks after the viral infection [7,8].

The viral determination from stool, pharynx and vesicle is the mainstay of the diagnosis, although,because of the high price of these methods (not to mention the newest RT-PCR), the diagnosis is established in most cases by clinical grounds. Enteroviruses persist 1-4 weeks in the naso-pharynx and 1-18 weeks in the stool [9,10]

We confrunted during summer-autumn 2012 with many cases, especially in children,but not rare in adults too. It was quite unusual to see so many different and interesting cases in a part of Europe (north-eastern of Romania) where we were used to diagnose one or two cases per year, or even in a few years. We were teaching our residents about this disease using pictures from books or from articles published in the last years, mostly in Asia.

All our cases were not severe, with no complications, a 6-10 days evolution, high contagiosity among children; the diagnosis was only clinical, no virology tests were performed; with different and sometimes atypical picture, with all the laboratory parameters within normal limits.

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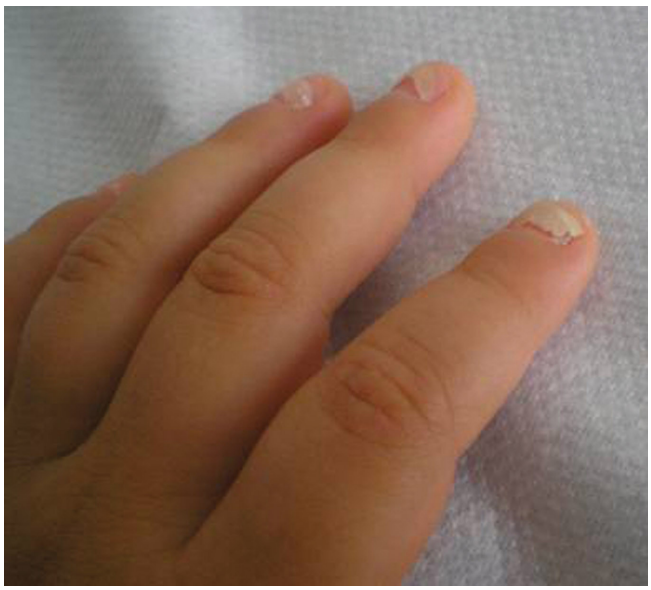

Figure 7. Onycomadesis in a small child (proximal separation of the nail plate from the nail bed with the shedding of the nail).

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CLAVES PARA EL DIAGNÓSTICO DERMATOPATOLÓGICO DE LOS QUISTES CUTÁNEOS

DERMATOPATHOLOGICAL CLUES FOR THE DIAGNOSIS OF SKIN CYSTS

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Resumen
Los quistes cutáneos son patologías frecuentes en la práctica dermatológica. Tienen etiologías variadas, y aunque la mayoría son adquiridos también pueden representar un fenómeno determinado genéticamente. Pueden ser esporádicos o aparecer de forma familiar. Son clínicamente fáciles de diagnosticar como "quistes", pero el diagnóstico de certeza es estrictamente anatomo-patológico, ya que los mismos se nombran por el tipo de epitelio que los reviste, los elementos que se observan en sus paredes y en su interior. Los pacientes con quistes consultan por preocupaciones cosméticas o debido a las molestias de la irritación mecánica o inflamación del quiste. Las lesiones pueden ser proliferantes e incluso pueden desarrollarse tumores a partir del epitelio.
Hacemos una breve revisión de los quistes y pseudoquistes cutáneos más importantes y presentamos tres casos clínicos.

Abstract
Skin cysts are common conditions in dermatology practice. They have varied etiologies, and while most are acquired they may also represent a genetically determined phenomenon. They can be sporadic or familial. They are easily diagnosed clinically as „cysts", but the definitive diagnosis is strictly pathological, since they are named for the type of epithelial lining, the elements seen in the walls and the elements inside. Patients with cysts consult for cosmetic concerns and for inconvenience caused by mechanical irritation or inflammation of the cyst. The lesions may proliferate and even tumors may develop from the epithelial coating.
We briefly review the most important skin cysts and pseudocysts and present three clinical cases.

Palabras clave: quiste; pseudoquiste; quiste epidérmico; quiste triquilemal; esteatocistoma
Key words: cyst; pseudocyst epidermal cyst; trichilemmal cyst; steatocystoma

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Introducción

Se define como quiste a una cavidad de contenido líquido o semi-sólido revestida por epitelio [1]. Su apariencia clínica no es específica por lo que su diagnóstico de certeza es estrictamente anatomo-patológico y se los designa según el tipo de epitelio que tapiza sus paredes.
En general los quistes cutáneos se hallan revestidos por un solo tipo de epitelio, el cual suele ser en la mayoría de los casos de tipo escamoso estratificado. Existen algunos casos en que se pueden presentar más de un tipo de epitelio de revestimiento y en ese caso se denominan híbridos.
Se diferencian de los pseudoquistes en que éstas son lesiones que forman cavidades que carecen de revestimiento epitelial.
Un seno es un trayecto tapizado por epitelio o recubierto por tejido de granulación, de origen malformativo o inflamatorio, y una hendidura es pequeña depresión en la superficie epidérmica.

Los quistes cutáneos se originan de las diversas porciones de los anexos, ya sean las unidades pilosebáceas o las glándulas sudoríparas.
En un breve repaso de la anatomía de los anejos cutáneos, la unidad pilosebácea recordemos se divide en tres porciones:
1. Infundibular: entre el ostium folicular y la glándula sebácea. De esta porción derivan el quiste epidérmico, de milium, folicular pigmentado y de pelo velloso.
2. Istmica: entre la glándula sebácea y la inserción del músculo piloerector. De esta porción derivan el quiste triquilemal y el esteatocistoma.
3. Inferior: entre la inserción del músculo piloerector y la papila dérmica del pelo.
De las porciones secretora y excretora de las glándulas sudoríparas ecrinas y apocrinas se originan los hidrocistomas.
Solo en muy escasas ocasiones los quistes se originan a causa de alteraciones del desarrollo.

Clasificación

En base al epitelio de revestimiento pueden dividirse en (Tabl. I):

Hallazgos histopatológicos en las formas más frecuentes de quistes cutáneos

Con revestimiento epitelial:

1. Tapizados por epitelio escamoso estratificado
   - o No contienen estructuras en su pared:
     - Epidérmico.
     - Pilar.
     - Millium.
     - Híbrido.
     - Pelo velloso.
     - Folicular pigmentado.
   - o Contienen estructuras en su pared:
     - Esteatocistomas.
     - Tímicos.
     - Dermoides.

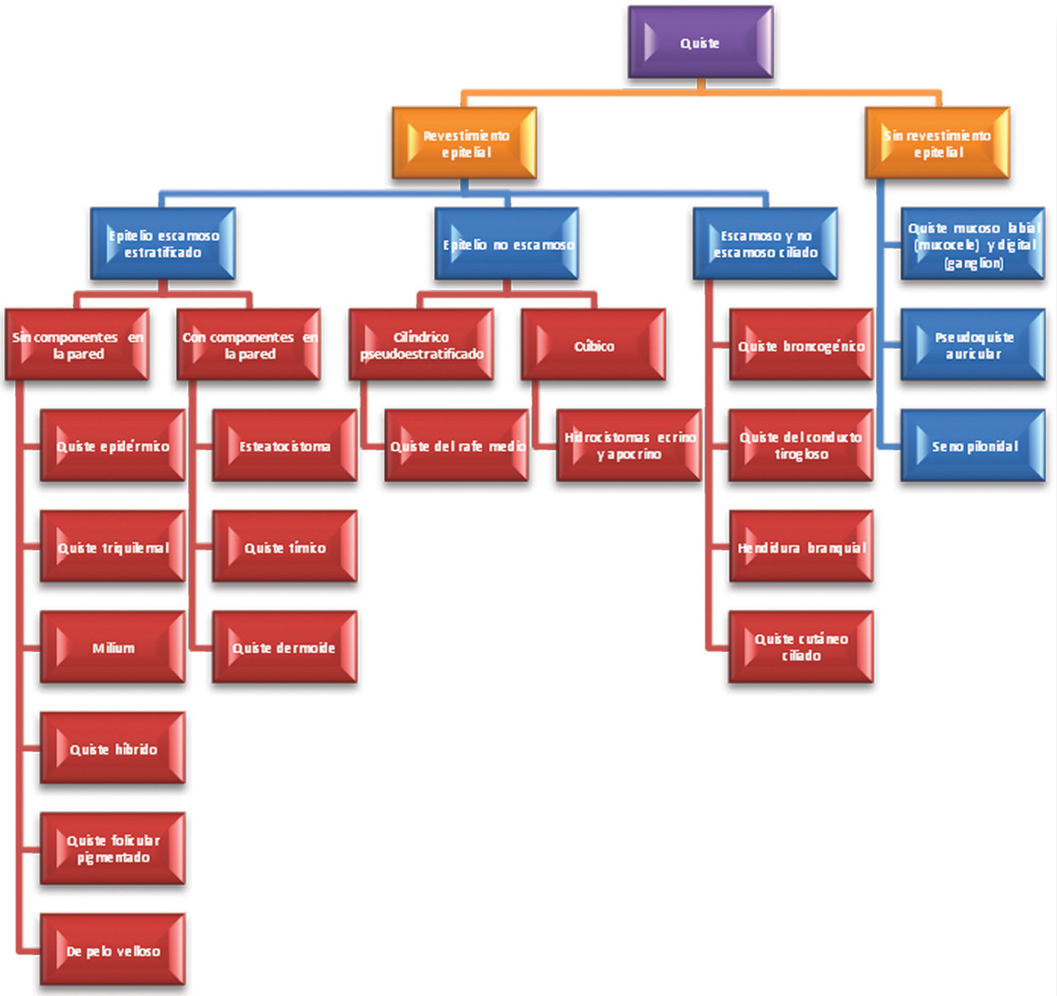

Tabla I. Clasificación de los quistes y pseudoquistes cutáneos en base a su epitelio de revestimiento
Table I. Classification of cysts and pseudocysts based on their lining epithelium

Quiste epidérmico o infundibular (QE):

Llamado también "de inclusión". Se trata de una formación quística originada de la porción infundibular de la unidad pilosebácea. Son los más comunes (80%). Asientan sobre todo en cara, cuello, torso, hasta en palmas y plantas. Piel acra: implantación post trauma. Piel no acra: inflamación del folículo. Formas múltiples en escroto posteriormente se calcifican «calcinosis escrotal» [2].

Epitelio: escamoso estratificado con presencia de capa granulosa.

Pared: desprovista de elementos.

Contenido: queratina laminada ortoqueratósica.

Ruptura: proceso inflamatorio desde agudo supurativo hasta crónico granulomatoso con células gigantes (Fig. 1).

Milium:

Se trata de una formación quística originada de la porción infundibular de la unidad pilosebácea en su forma primaria o del infundíbulo, conductos ecrinos, etc. en su forma secundaria. Es un quiste epidérmico (infundibular) en miniatura, localizado en la porción superficial de la dermis, pudiendo manifestarse de forma primaria o secundaria a trauma, biopsia previa, quemadura, dermoabrasión o acompañando patologías ampollosas como el penfigoide ampolloso o la porfiria cutánea tarda [3].

Epitelio: escamoso estratificado con presencia de capa granulosa.

Pared: desprovista de elementos.

Contenido: queratina laminada ortoqueratósica.

Ruptura: proceso inflamatorio desde agudo supurativo hasta crónico granulomatoso con células gigantes.

Los quistes de milium secundarios se rodean de tejido conectivo cicatricial como un hallazgo bastante constante.

Quiste pilar:

Llamado también triquilemal o sebáceo. El 2º en frecuencia

(10-15%) detrás del quiste epidérmico o infundibular. 90% asientan en cuero cabelludo (Fig. 2). Tienen un claro predominio femenino. Son solitarios, aunque se describen formas múltiples. Los casos hereditarios están vinculados al cromosoma 3p24-p21.2.10. Se trata de una formación quística originada del istmo. Asienta en dermis reticular media a profunda y tejido celular subcutáneo.
Epitelio: escamoso estratificado de 3-4 capas de espesor con queratinización abrupta sin interposición de capa granulosa. Células más cercanas a la cavidad pálidas.
Pared: desprovista de elementos.
Contenido: queratina homogénea y compacta. Ocasionales hendiduras de colesterol en la queratina.
Ruptura: reacción granulomatosa de cuerpo extraño, calcificación (25%) y osificación.
Áreas con granulosa («quíste híbrido») [4].

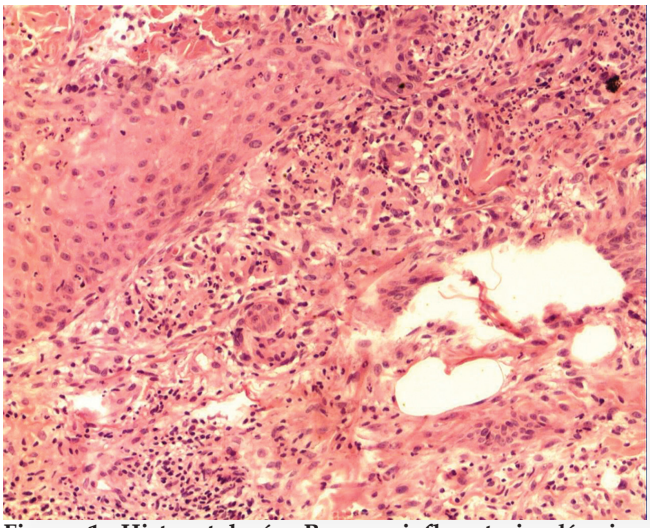

Figura 1. Histopatología. Proceso inflamatorio dérmico, crónico granulomatoso con células gigantes multinucleadas de tipo cuerpo extraño a material querático, secundario a ruptura de quiste epidérmico. Hay focos de supuración.
Figure 1. Histopathology. Dermal chronic granulomatous inflammatory process, with multinucleated giant cells of foreign body type, secondary to the ruptured of an epidermal cyst. Foci of suppuration.

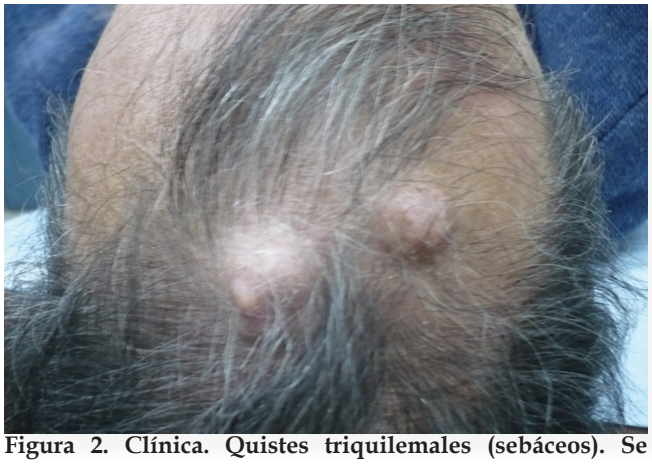

Figura 2. Clínica. Quistes triquilemales (sebáceos). Se observan dos grandes lesiones que asientan en cuero cabelludo.
Figure 2. Clinic. Trichilemmal (sebaceous) cysts. Two large lesions on the scalp.

Quiste de pelo velloso:
Pueden ser múltiples con herencia autosómico dominante (quistes del pelo velloso eruptivos) o solitarios no hereditarios. No hereditarios: aparición abrupta en la 2ª-3ª décadas de la vida. Adultos. Casos familiares: edades más tempranas incluso nacimiento. Lesiones pequeñas, pigmentadas. Asientan en tórax anterior, abdomen y extremidades. Se sitúan en dermis media o superior [5-7].
Epitelio: escamoso estratificado con capa granulosa (igual al QE ya que también se forman del infundíbulo).
Pared: desprovista de elementos.
Contenido: Presencia de múltiples pelos cortados transversalmente (no pigmentados) en la cavidad mezclados con queratina.

Esteatocistoma:
Lesiones solitarias no heredadas (simple) o lesiones múltiples heredadas (AD). Forma hereditaria: lesiones numerosas, pequeñas, blancas o amarillentas en axilas, ingle o pecho aunque otras áreas del cuerpo pueden afectarse. Las lesiones por lo general ocurren en la pubertad, lo que sugiere cierto control androgénico [5,6].
Epitelio: escamoso (sin granulosa) corrugado y contiene una cutícula eosinofílica en su superficie.
Pared: glándulas sebáceas adyacentes a la pared del quiste.
Contenido: desprovista de elementos.

Con revestimiento epitelial:

1. Tapizados por epitelio no escamoso [7]:
   - Cilíndrico pseudoestratificado: Quiste del rafe medio.
   - Cúbico: Hidrocistomas ecrino y apocrino.

Hidrocistomas ecrino y apocrino:
Proliferaciones quísticas de las glándulas sudoríparas con diferenciación apocrina (la mayoría) o ecrina. Apocrinos: derivan de la porción secretora. Ecrinos: derivan del conducto excretor. Son raros, 1/1.000 biopsias cutáneas remitidas. Solitarios. Se han descrito casos múltiples. Se presentan en la edad media o superior y se describen casos en niños y adolescentes sin predominio de sexo. Predilección por el área de la cabeza y cuello (región periorbitaria y cuero cabelludo) y periné (Fig. 3). Su etiología es desconocida. Se exacerban con las altas temperaturas y desaparecen con el frío y el tto. con atropina [9-11].
Se presentan como una pápula o nódulo quístico firme, superficie lisa, color azulado. En algún caso el contenido del quiste es marrón a negro.
Macroscopía: 0.5-1.0 cm. (hasta 7cm.). Dermis. Se han descrito en el tejido celular subcutáneo. Corte cavidad quística uni o multilocular.
Histopatología: Doble capa de células epiteliales: Interna: células columnares, citoplasma eosinófilo, secreción por decapitación.

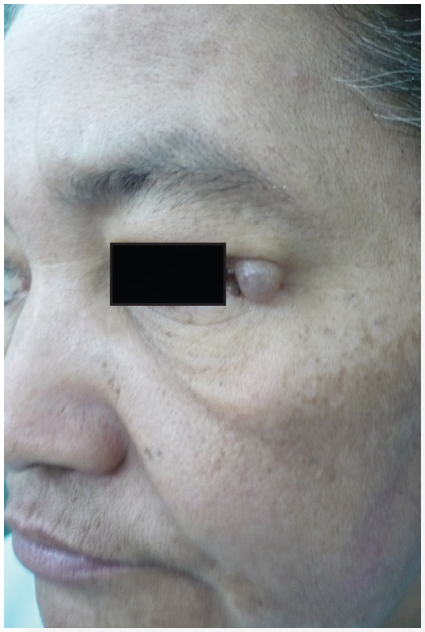

Figura 3. Clínica. Hidrocistoma. La lesión aparece como una pápula o nódulo palpebral traslucente.
Figure 3. Clinic. Hidrocystoma. The lesion appears as a translucent lid papule or nodule.

Externa: células planas y células mioepiteliales. Pueden haber proyecciones papilares hacia la luz. Epitelio aplanado por la secreción. Los quistes carecen de conexión con la epidermis.
La resección es curativa. Se pueden tratar con atropina y escopolamina y evitar ambientes cálidos.

Síndromes asociados [12-14]:
· **Síndrome de Goltz-Gorlin:**
- Esporádicos. Pocos casos familiares con herencia ligada a X.
- Mujeres.
- Microcefalia, hipoplasia mediofacial, orejas mal formadas, microftalmia, hidrocistomas múltiples perioculares, papilomas de lengua-labio-ano-axila, anomalías esqueléticas y retraso mental.
· **Síndrome de Schopf-Schulz-Passage:**
- AR.
- Hidrocistomas apocrinos múltiples del párpado, hiperqueratosis palmo-plantar, hipodoncia e hipotricosis.
· **Enfermedad de Graves:**
- Múltiples hidrocistomas ecrinos (relacionado con la hiperhidrosis).
· **Sin revestimiento epitelial (pseudoquistes):**
- Quiste mucoso labial (mucocele) y digital (ganglion)
- Seno pilonidal
- Pseudoquiste auricular [15].

Quiste mucoso labial y digital:
Digital: Asientan en dorso de los dedos cerca de la articulación interfalángica distal y comunican con el espacio sinovial al cual están adheridos por un pedículo [16].
Mucocele: producido por la ruptura del conducto excretor de una glándula salivar menor, asentando en labio, mucosa oral o lengua.
La histología es similar en ambos casos:
· Pared: fibrosa de colágeno comprimida.
· Contenido: mucopolisacáridos.

Seno pilonidal:
Asientan en la región sacrococcígea de hombres hirsutos. Muy importante biopsiar por el desarrollo de un carcinoma epidermoide en lesiones de largo tiempo de evolución [17,18].
Se trata de un trayecto sinusal tapizado por tejido de granulación. Se observan pelos en la cavidad o en la pared. Fibrosis de la pared en la porción más profunda de la cavidad y denso infiltrado inflamatorio.

CASOS CLÍNICOS:

Caso Nº 1:
· Varón, 42 años, procedente de Asunción (Paraguay).
· Motivo de consulta (MC): lunar en la cabeza.
· Antecedentes de la enfermedad actual (AEA): lunar negruzco, redondo, desde el nacimiento, que va creciendo, no duele ni sangra.
· Antecedentes patológicos personales (APP): sin particularidades (s/p).
· Examen físico (EF): tumoración hiperpigmentada, negro-azulada, con superficie lobulada, de 1.5 cm. de eje mayor, en sien derecha.
· Diagnósticos clínicos presuntivos: Tumor de etiología a determinar (hemangioma, fibroma o nevus).
· Macroscopía: Fragmento cutáneo de 1.5x1x0.5 cm. de ejes mayores, en el que al corte se individualiza una formación quística multilocular de paredes milimétricas y contenido seroso amarillento, de 0.6cm de eje mayor (Fig. 4).
· Microscopía: Varias capas de células epiteliales columnares. Se observa secreción por decapitación (Fig. 5).
· Diagnóstico anatomo-patológico final:
HIDROCISTOMA APOCRINO.

Caso Nº2:
· Varón, edad desconocida, procedente de Asunción (Paraguay).
· MC: lesiones en piel del pecho.
· APP y APP: s/p.
· EF: múltiples lesiones tipo pápulas color piel en tórax anterior, de dimensiones milimétricas.
· Diagnóstico clínico presuntivo: quistes epidérmicos.
· Microscopía: cavidad quística que asienta en dermis tapizada por un epitelio escamoso estratificado sin capa granulosa, corrugado, y con una cutícula eosinofílica en su superficie. Se observan glándulas sebáceas adyacentes a la pared del quiste y la cavidad está desprovista de elementos (Fig. 6).
· Diagnóstico anatomo-patológico final:
ESTEATOCISTOMA.

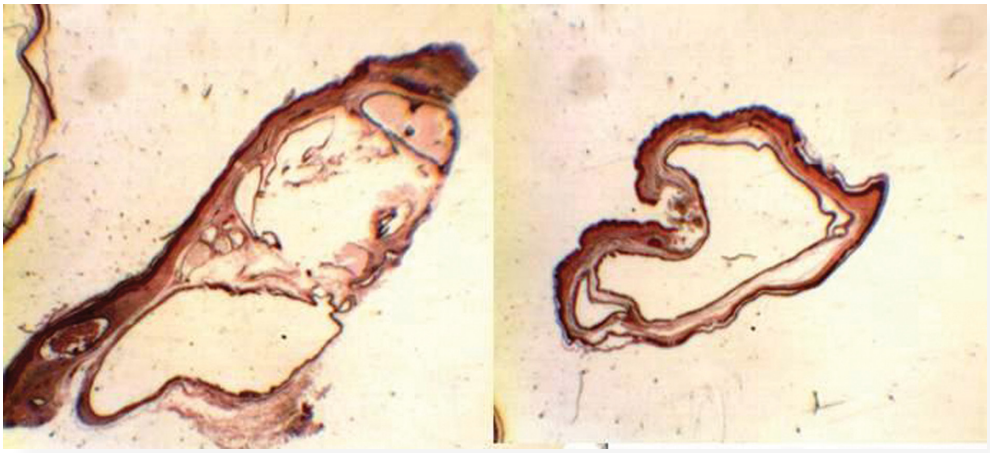

Figura 4. Caso 1. Macro-micoscopía. Formación quística multilocular asentando en dermis, de paredes milimétricas y contenido seroso amarillento, de 0.6 cm. de eje mayor.
Figure 4. Case 1. Macro-microscopy. Multilocular cystic settling in dermis, millimeter walls and yellowish serous content, of 0.6 cm. of major axis.

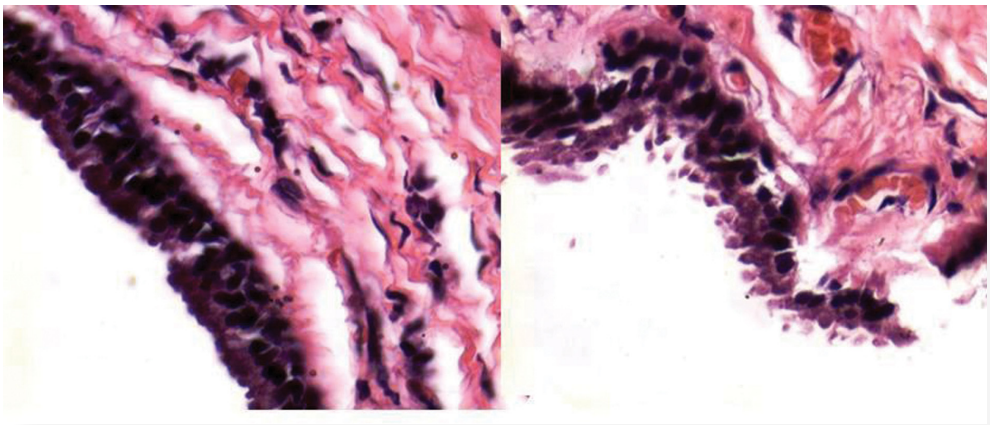

Figura 5. Caso 1. Histopatología. Epitelio de revestimiento del quiste constituido por varias capas de células epiteliales columnares. Se observa secreción por decapitación.
Figura 5. Case 1. Histopathology. Cyst lining composed of several layers of columnar epithelial cells. Secretion by decapitation is observed.

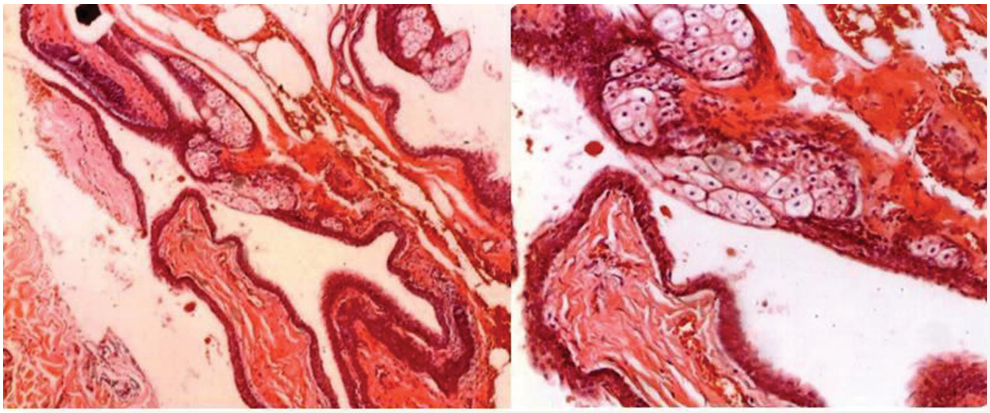

Figura 6. Caso 2. Histopatología. Cavidad quística que asienta en dermis, tapizada por epitelio escamoso poliestratificado corrugado con cutícula eosinofílica superficial y glándulas sebáceas adyacentes a la pared. La cavidad está desprovista de elementos.
Figure 6. Case 2. Histopathology. The cystic cavity sits in dermis and it's lined by stratified corrugated squamous epithelium without granular layer and a eosinophilic surfacecuticle. Sebaceous glands are observed adjacent to the wall of the cyst. The cavity is devoid of elements.

Caso Nº 3:

· Niña, edad desconocida. Procedente de Asunción (Paraguay).

· MC: lesiones en piel del pecho y miembros superiores.

· APP y APP: desconocidos.

· EF: múltiples lesiones pequeñas, de dimensiones milimétricas, papulosas, pigmentadas en tórax anterior y ambas extremidades superiores.

· Diagnóstico clínico presuntivo: quistes epidérmicos, esteatocistomas o quistes vellosos eruptivos.

· Microscopía: cavidad quística situada en dermis media e inferior tapizada por un epitelio escamoso estratificado muy bajo (quizás aplanado y comprimido por el contenido del quiste) con capa granulosa y pared desprovista de elementos. El contenido de la cavidad muestra múltiples pelos cortados transversalmente, mezclados con queratina (Fig. 7, 8).

· Diagnóstico anatomo-patológico final:

QUISTE DE PELO VELLOSO.

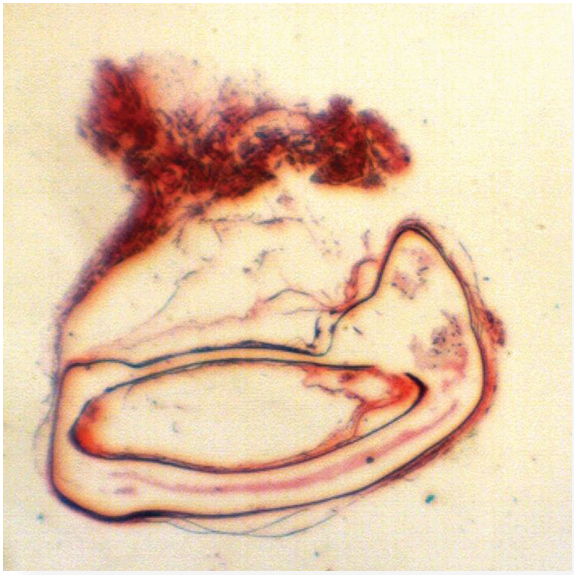

Figura 7. Caso 3. Histopatología. Cavidad quística situada en dermis media e inferior.

Figure 7. Case 3. Histopathology. Cystic cavity is located in the middle and lower dermis.

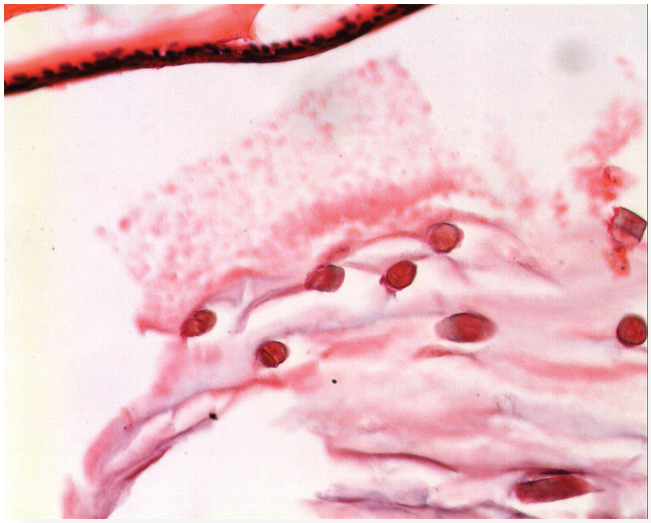

Figura 8. Caso 3. Histopatología. La cavidad está tapizada por un epitelio escamoso aplanado y comprimido por el contenido del quiste, con capa granulosa y pared desprovista de elementos. En el interior se observa múltiples pelos cortados transversalmente, mezclados con queratina.

Figure 8. Case 3. Histopathology. The cavity is lined by flattened squamous epithelium that is compressed by the contents of the cyst. The wall has granular layer and it's devoid of elements. Inside there are multiple hairs cut transversely, mixed with keratin.

Conclusiones:

1. Los quistes cutáneos son patologías frecuentes en la práctica dermatológica.
2. Ya que su apariencia clínica no es específica su diagnóstico de certeza es estrictamente anatomo-patológico [19].
3. Los pacientes con quistes pueden consultar debido a preocupaciones médicas o cosméticas, o debido a las molestias de la irritación mecánica o inflamación del quiste.
4. Siempre deben ser biopsiados, ya que su etiología no es inflamatoria en todos los casos y algunos surgen a causa de alteraciones del desarrollo o son complicaciones de procedimientos médico-quirúrgicos (dermoabrasión, toma de biopsia, etc.).
5. Pueden acompañar algunos síndromes esporádicos o hereditarios.
6. Las lesiones pueden ser proliferantes e incluso pueden desarrollarse carcinomas epidermoides sobre lesiones de larga data [17,18,20].

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FACIAL SPOROTRICHOSIS IN CHILDREN FROM ENDEMIC AREA IN PERU

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Sporotrichosis is a subcutaneous mycosis subacute or chronic evolution, caused by the dimorphic Sporothrix complex, which includes five species: *Sporothrix albicans*, *Sporothrix brasiliensis*, *Sporothrix globosa*, *Sporothrix mexicana* and *Sporothrix schenckii (sensu stricto)*. The infection occurs after trauma with contaminated material, which inoculated the fungus on the skin. The clinical types of sporotrichosis are lymphocutaneous sporotrichosis, fixed cutaneous (nodulopapular, ulcerative, verrucouse and furunculoide) and extracutaneous [1,2]. It is a mycosis of a universal distribution, but more commonly seen in tropical or subtropical countries. The majority of cases are found in Mexico, Central America and Peru [1]. However, the region of Abancay in Peru is considered hyperendemic and has an annual incidence of 48-60 cases per 100 000 population [3]. Also, the incidence is three times higher in children aged 0-14 in ≥ 15 years of age, approximately 1 case/1000 children 0-14 years and a frequency of 60% [4,5]. In pediatric patients from endemic areas the most anatomical site affected is the facial region (40-42%) [5,6], with lesions on the nose, face, chin, malar, genian and palpebral that is clinically rarely mentioned in the literature [7].

Diagnosis:
The gold standard for diagnosis is the mycological culture of secretions from injuries. In the crop in Sabouraud dextrose agar with chloramphenicol at 25 ° C colonies are observed finely radiated creamy white and brown. Lactophenol blue stained observed microscopically thin and branched hifas. In BHI agar at 37°C are observed yeast creamy, moist, and whitish. Microscopically with oval and globular structures [1].

Treatment:
In our experience treatment with potassium iodide in the form of saturated (SSKI) at doses of 2-20 drops / 3 times / day is preferred for pediatric cases and lymphocutaneous fixed cutaneous form [5]. Other treatment options are oral imidazole, ketoconazole, itraconazole or fluconazole and terbinafine at standard doses with favorable response in terms of three to five months of continuous administration [1].
We present photographs of pediatric patients with cutaneous sporotrichosis fixed facial skin and lymphatic topographical characteristics can be useful to be considered in the differential diagnosis against other infections in children from endemic areas also are images of the etiologic agent *Sporothrix schenckii*. The photographs were obtained from patients treated at Santa Teresa Clinic of Abancay, Peru.
Ethical considerations: For taking photographs and publishing these. Informed consent was obtained in writing and signed by the parents of the patients. Figures 3, 4 and 5 represent the palpebral forms sporotrichosis. Magazines were published previously in Peru [5,6,8].

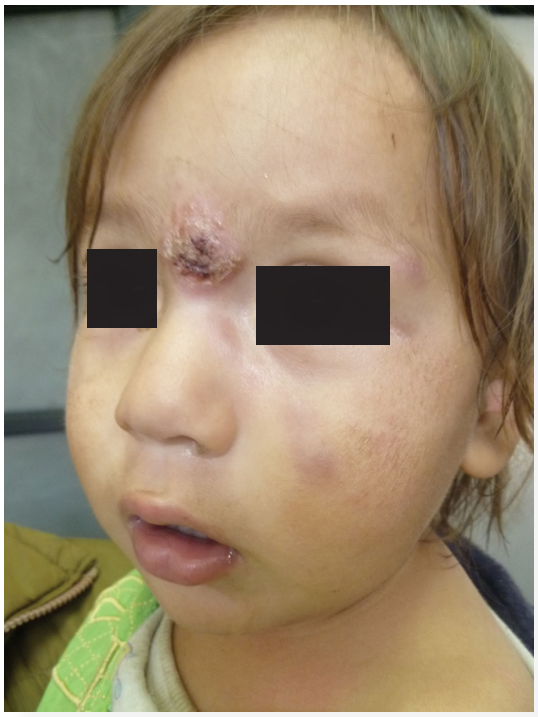

Figure 1. Cutaneous lymphatic sporotrichosis crusted-ulcer on the forehead and nose with nodules on the cheeks

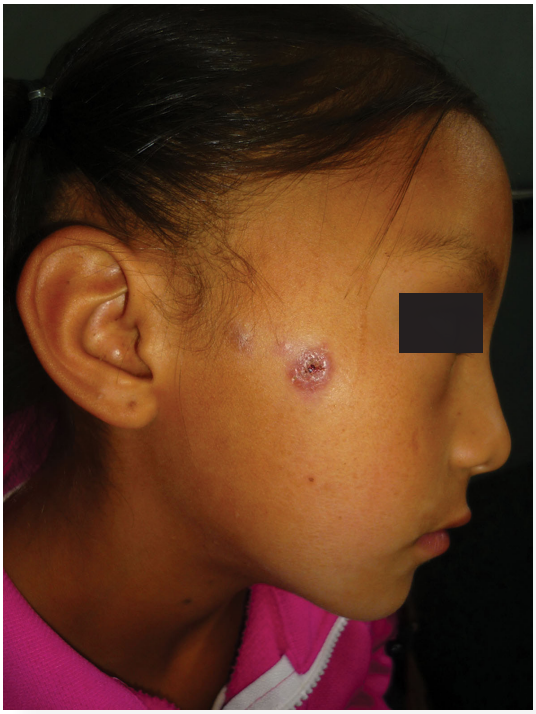

Figure 2. Sporotrichosis lymphocutaneous on the malar region

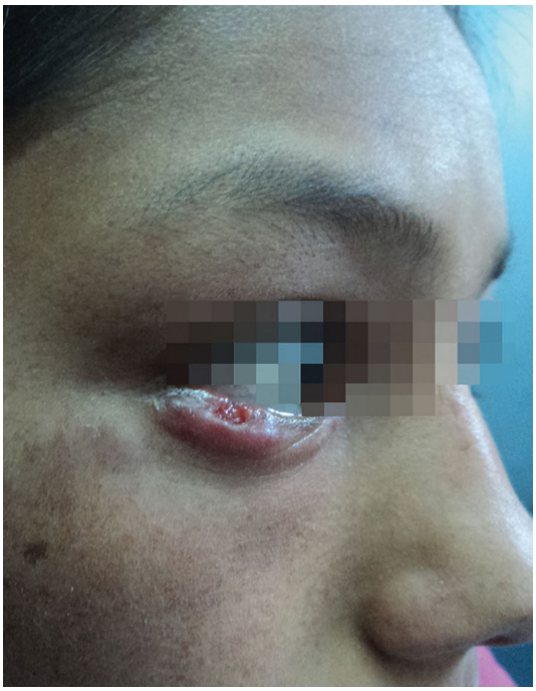

Figure 4. Ulcerated cutaneous lymphatic sporotrichosis in the right lower eyelid with nodules in the malar region

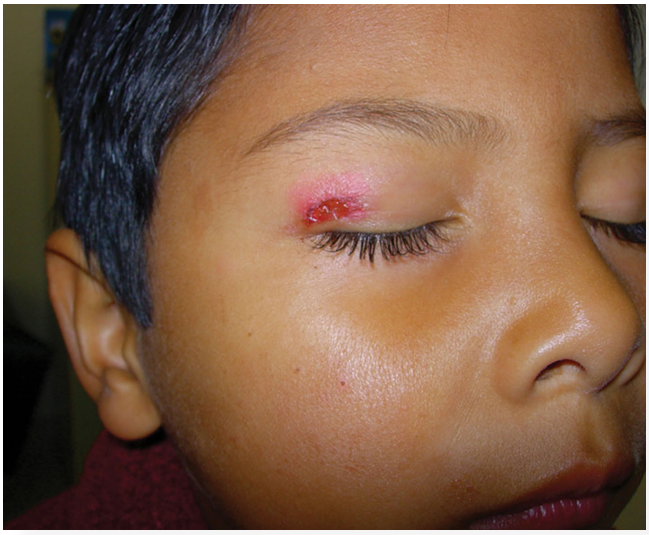

Figure 3. Fixed cutaneous sporotrichosis ulcerated right upper eyelid

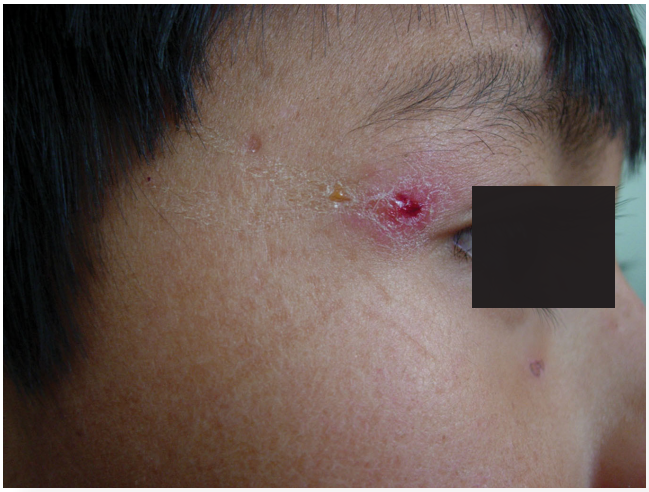

Figure 5. Fixed cutaneous sporotrichosis right upper eyelid

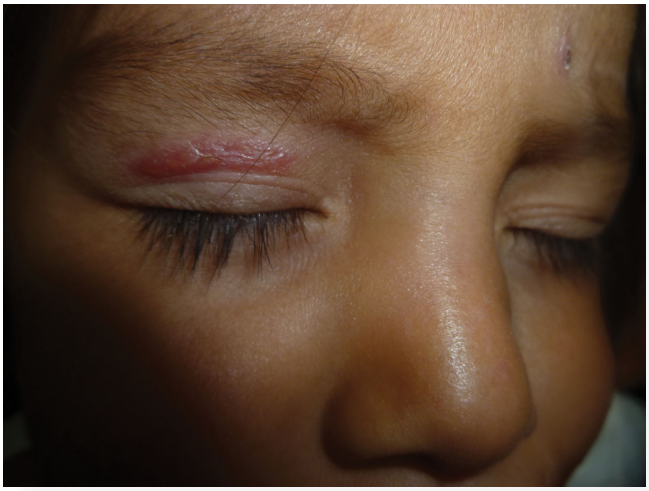

Figure 6. Palpebral fixed cutaneous sporotrichosis in child of two years old

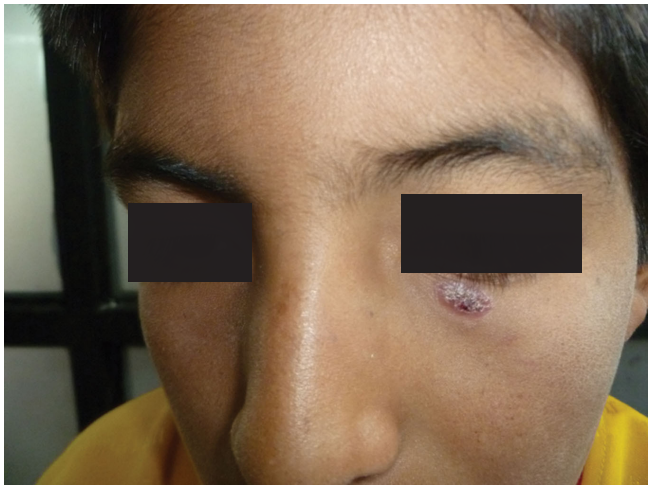

Figure 7. Fixed cutaneous sporotrichosis left lower eyelid

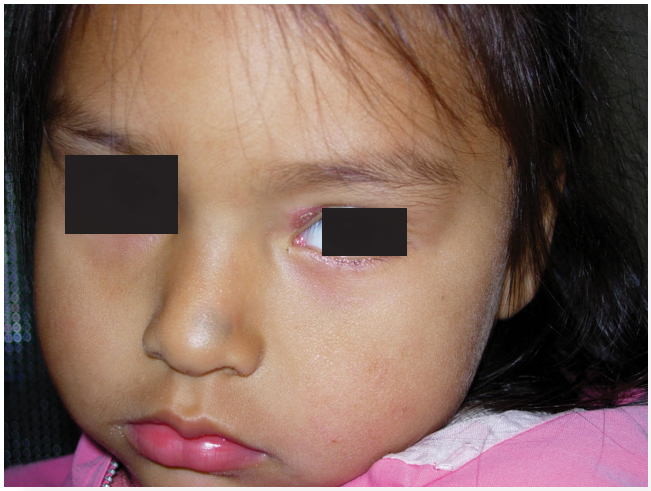

Figure 8. Fixed cutaneous sporotrichosis left upper eyelid

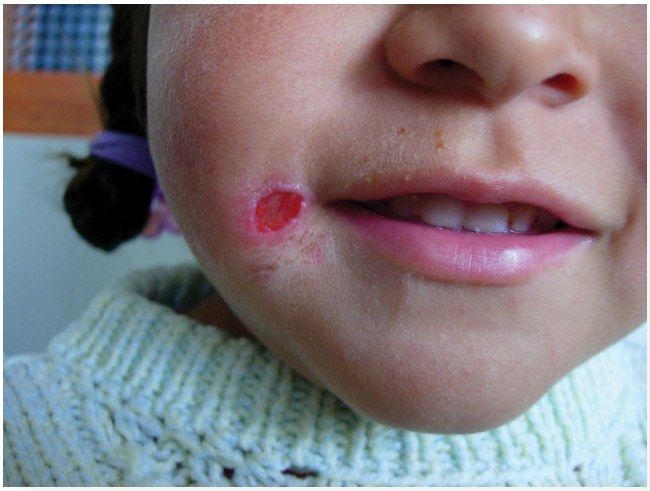

Figure 9. Fixed cutaneous sporotrichosis ulcerative the genian region

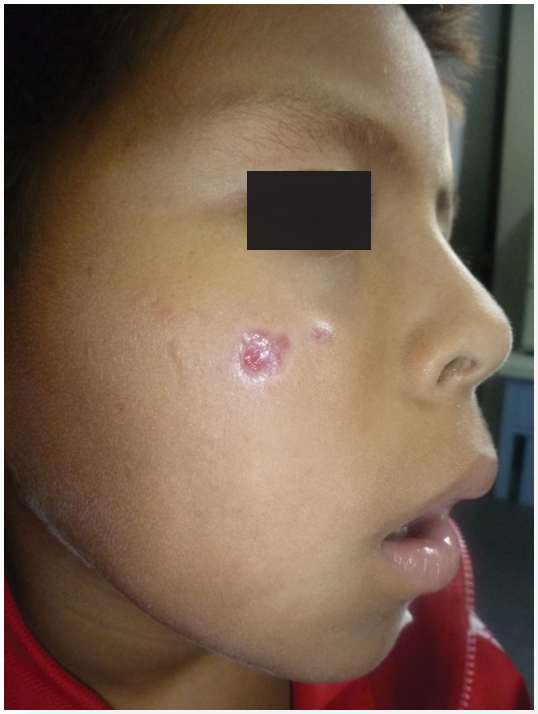

Figure 10. Fixed cutaneous sporotrichosis satellitosis cheek

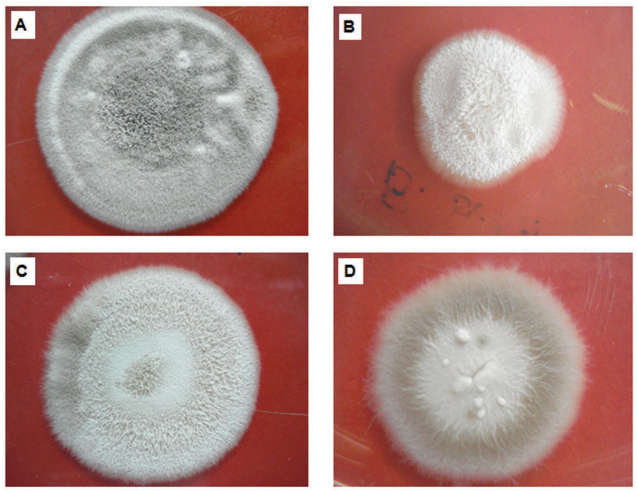

Figure 11. Sporothrix schenckii cultivation on Sabouraud dextrose agar with chloramphenicol at 25 ° C, colonies are observed with different shapes, white and brown finely grooved radiated

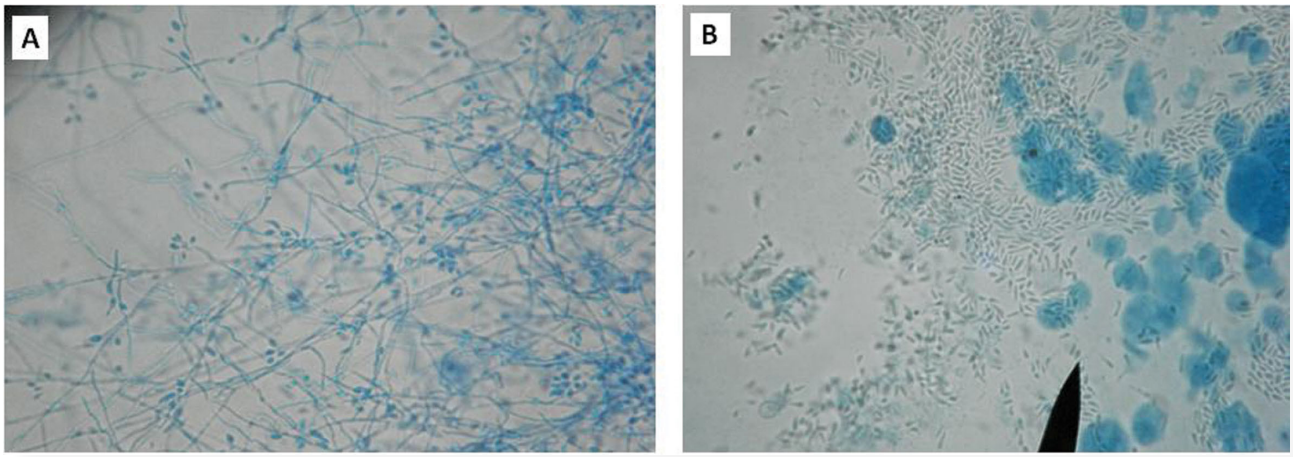

Figure 12. Photomicrograph of Sporothrix schenckii: A. Mycelial form: displayed twisted hifas with microconidia; lactophenol blue staining. B. Parasitic form: Yeast is observed in the form of cigar

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PETECHIAE - ADVERSE REACTIONS TO CIPROFLOXACIN

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Sir

There are many dermatological adverse reactions to Ciprofloxacin reported in the literature: allergic reaction, pruritus, urticaria, photosensitivity/phototoxicity reaction, flushing, fever, chills, angioedema, edema of the face, neck, lips, conjunctivae or hands, cutaneous candidiasis, hyperpigmentation, erythema nodosum, sweating [1-5].

We report a case of petechiae appeared 24 hours after the initiation of Ciprofloxacin treatment for urinary infection,with long lasting evolution, despite the immediate withdrawal of the medication.The petechiae were present on the lower limbs, with intense pruritus, no systemic reactions.

Case presentation

A 65-year-old woman, with a history of chronic urinary infections, for the first time treated with Ciprofloxacine 500mg twice daily, presented to our Department for the sudden onset (24 hours before the admission to the hospital) of a petechial rash on the lower limbs (Fig. 1a, 1b). No fever, no gastro-intestinal symptoms, just a slight pruritus on the site of the lesions. She was in good health state and she reported the first cutaneous manifestations after the second intake of the drug.

All the laboratory parameters were within normal range (no thrombocytopenia).

The medication was stopped and the patient was under observation and a short course of antihistamines, with wonderful results, but only after 21 days after the withdrawal of Ciprofloxacine therapy.

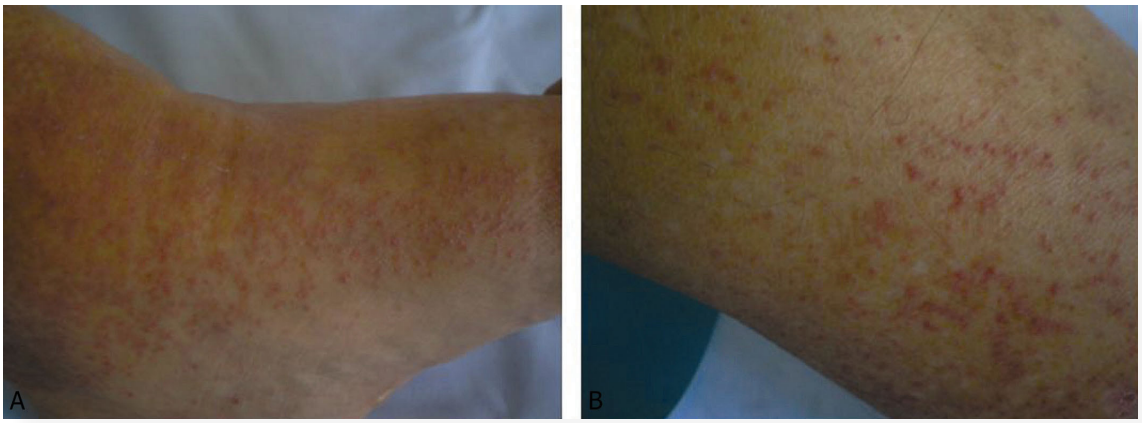

Figure 1 a,b. Petechiae on the lower limbs

Discussions

Ciprofloxacin is one of the most commonly used antibacterial agents with relatively few side effects. Serious adverse reactions reported with ciprofloxacin are rare with an incidence of 0.6% (Tabl. I).

It is well known that Fluoroquinolones can induce drug-dependent, platelet-reactive antibodies causing complement-mediated destruction of platelets and thrombocytopenia [1], but it was the case of our patient who had a normal number of platelets.

Also there have been reported a few cases of photo exposed purpuric eruptions during treatment with Ciprofloxacine [2], but our patient denied any exposure to sun or UV light and it was winter when the diagnosis of petechias induced by Ciprofloxacine was made [5].

There are few reports on petechial adverse reaction to Ciprofloxacine therapy and we want to aware clinicians about this possible side effect of this widely use medication.

Adverse reaction (L Mandell 2002)	Range of incidence (%)
Gastrointestinal (diarrhea, vomiting)	0.8 - 6.8
Central nervous system (dizziness, headache)	0.9 - 11
Skin (rashes)	0.4 - 2.1
Blood disorders	0.5 - 5.3
Cardiovascular (palpitations)	0.5 - 2.0
Musculoskeletal	0.5 - 2.0
Phototoxicity or photoallergy	0.5 - 2.1
Serious reactions, eg, hemolytic uremic syndrome, Stevens Johnson syndrome	<0.5

Table I. Adverse reaction (L Mandell 2002) to Ciprofloxacin [5]

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EPONYMS IN DERMATOLOGY LITERATURE LINKED TO GENITAL SKIN DISORDERS

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There are numerous eponymous systemic diseases which may affect genital skin or sexual organs in both genders. For example Behçet disease which is characterized by relapsing oral aphthae, genital ulcers and iritis.

This disease is named after Hulusi Behçet [1] (1889–1948), (Fig. 1), the Turkish dermatologist and scientist who first recognized the syndrome. This disease also called „Adamantiades' syndrome" or „Adamandiades-Behçet syndrome", for the work done by Benediktos Adamantiades. Benediktos Adamantiades (1875-1962), (Fig. 2) was a Greek ophthalmologist [2].

Another example of systemic disease with genital involvement is Reiter syndrome which is recently called reactive arthritis. This syndrome is characterized by arthritis, urethritis, and conjunctivitis.

It is named after German physician Hans Reiter [3] (1881-1969), (Fig. 3). However, the name „Reiter's syndrome", has become unpopular in the past decade as Reiter's history of Nazi party membership, forced human experimentation in the Buchenwald concentration camp, and subsequent prosecution in Nuremberg as a war criminal, have come to light.

One more example is syphilis. In 1530, the name „syphilis" was first used by the Italian physician and poet Girolamo Fracastoro [4] (1478-1553), as the title of his Latin poem in dactylic hexameter describing the ravages of the disease in Italy. In his well-known poem „Syphilidis sive de morbo gallico libri tres" (Three books on syphilis or the French disease), he coined the name by which we now know the disease from the legend of a shepherd called Syphilus who had purportedly gotten the illness as a punishment for defying the gods.

In Table I [5-20], however, we highlighted in particular to eponymous conditions which are primarily a disorders of genital area.

Figure 1. Hulusi Behçet (1889–1948)

Figure 2. Benediktos Adamantiades (1875-1962)

Figure 3. Hans Reiter (1881-1969). A courtesy of National library of Medicine

Eponyms in dermatology literature linked to the disorder of genital area	Remarks
Bartholin cyst [5]	It occurs as a result of blockage to the Bartholin gland which was first described in 1677 by Danish anatomist Caspar Secundus Bartholin (1655–1738).
Buschke-Loewenstein Tumor [6]	This is another name for giant condyloma accuminata of genital skin. It is named after dermatologists from Germany, Abraham Buschke (1868-1943), (Fig. 4) and Ludwig Loewenstein (1895-1959).  **Figure 4. Abraham Buschke (1868-1943). Reproduced from reference number 6 . The original caption reads: "Photo of Prof. Buschke taken in 1907." From Curth W, Ollendorff-Curth H: Remembering Abraham Buschke (1860-1943). Am J Dermatopathol. 1983:5; Figure 1, page 28.**
Fournier's gangrene [7]	Fulminating infection of the scrotum leading to gangrene and commonly associated with diabetes. It is a type of necrotizing infection or gangrene usually affecting the perineum. It was first described by Baurienne in 1764 and is named after, Jean Alfred Fournier (1832-1914), French dermatologist (Fig. 5).  **Figure 5. Jean Alfred Fournier (1832-1914). A courtesy of National library of Medicine.**
Klingsor Syndrome [8]	This syndrome denotes to self-mutilation of the external genitals in psychiatric patients. It was applied only to acts of genital self-mutation, involving religious delusions. However, it was suggested that the syndromeshould also include cases which involve genital self. Mutilation associated with all delusional syndromes. The name "Klingsor" was based on a fictitious characterin Wagner's opera where Klingsor was a magician who Castrated himself in an unsuccessful attempt to gain acceptance from the Knights of the Grail.
Lipschütz' ulcer [9,10]	Lipschütz acute genital ulcer is a rare distinctive cause of nonvenereal acute genital ulcers that occurs particularly in adolescents described in 1913. The etiology is unknown, although recent reports have associated it with the Epstein-Barr virus. The diagnosis is made by exclusion after ruling out sexually transmitted diseases, autoimmune causes, trauma, and other etiologies of genital ulcerations. It is named after Benjamin Lipschütz (1878-1931) Austrian dermatologist and microbiologist.
Mondor's disease [11,12]	Superficial thoracic wall and dorsal vein of the penis phlebitis are uncommon diseases. Both are known as Mondor's disease. It is named after Henri Mondor (1885-1962), (Fig. 6), a surgeon in Paris, France who first described the disease in 1939.

Table I. Selected Eponyms in dermatology literature linked to the disorder of genital area

Figure 6. Henri Mondor (1885-1962).

Eponyms in dermatology literature linked to the disorder of genital area	Remarks
Paget disease (extramammary) [13,14]	Vulvar Paget, disease is the most common site of extramammary Paget's disease (EMPD). The disease frequently associated with the underlying invasive skin adnexal carcinoma or representing the migration of underlying internal malignancy, especially anorectal and genitourinary cancer. It is named after an English surgeon and pathologist, Sir James Paget, 1st Baronet (1814-1899), (Fig. 7).  **Figure 7. Sir James Paget, 1st Baronet (1814 – 1899). A courtesy of National library of Medicine.**
Priapism [15]	It is a potentially painful medical condition, in which the erect penis or clitoris does not return to its flaccid state, despite the absence of both physical and psychological stimulation, within four hours. The name comes from the Greek god Priapus, a fertility god often represented with a disproportionately large and permanent erection. The acronym, ASPEN syndrome, was proposed for the association of sickle cell disease, priapism, exchange transfusion and neurological events.
Prince Albert's piercing [16,17]	Prince Albert penile piercing is a metallic bead, which is anchored to the urethral opening. Most of the body piercing is individually named, but the renowned piercer Jim Ward, who developed the magazine Piercing Fans International Quarterly in the late 1970s, accepted that most of the names were contrived. One of the most renowned piercings of the male genitals is the Prince Albert (1819–1861), (Fig. 8). However, the idea that Prince Albert wore a penis ring to tie his member down and prevent an offensive bulge in the breeches is a modern myth. Concerns over the possibility of hepatitis B and C and human immunodeficiency virus (HIV) transmission from body piercing are probably well founded. As with any surgical procedure that involves piercing the skin, the possibility of bleeding and infection must be considered. Other documented complications include urethral stricture, priapism, paraphimosis, and recurrent condyloma acuminatum. Due consideration should also be given to possible complications to the partner of the individual who has been genitally pierced. A review of some of the piercing Web sites reveals anecdotal complications in the form of trauma to the vagina or anus, teeth chipping, and choking.

Table I. Selected Eponyms in dermatology literature linked to the disorder of genital area (continued)

Figure 8. Prince Albert (1819–1861).

Figure 9. François Gigot de La Peyronie (1678-1747). A courtesy of National library of Medicine.

Figure 10. J.J.Zoon (1902-1952). Reproduced from reference number 20.

Eponyms in dermatology literature linked to the disorder of genital area	Remarks
Peyronie's disease [18]	Also known as „Induratio penis plastica", or more recently Chronic Inflammation of the Tunica Albuginea (CITA), is a connective tissue disorder involving the growth of fibrous plaques in the soft tissue of the penis. It causes erectile dysfunction. It is named after François Gigot de La Peyronie (1678-1747), (Fig. 9) the first surgeon to Louis XV.
Zoon balanitis [19,20]	This is another name for Plasma cell balanitis, which is a benign asymptomatic but chronic and erosive inflammatory condition of the glans penis and prepuce that generally affects uncircumcised men in later years. Clinical presentation involves a single, shiny, well defined reddish patch. It is named after J.J.Zoon (1902-1952), (Fig. 10).

Table I. Selected Eponyms in dermatology literature linked to the disorder of genital area (continued)

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EPONYMS IN THE DERMATOLOGY LITERATURE LINKED TO THE ORAL DISORDERS

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Diseases which involve the oral cavity usually derive their names from either Greek or Latin. These terms are customarily based on etiology or description of the lesion [1]. However, there are many eponyms as well.

Some of these names are misnomers. The misnomers encountered in oral pathology may arise from lack of understanding of underlying etiology, pathogenesis, histopathology, and/or concepts. Some misnomers are due to imprecise translations from word origins, etymological bungles, and/or factual errors [1].

In this manuscript, we are reviewing only, some selected examples of eponyms linked to the oral disorders (Tabl. I) [2-13].

Eponyms in the dermatology literature linked to oral disorders	Remarks
Behcet disease [2]	It is characterized by relapsing oral aphthae, genital ulcers and iritis. This disease is named after Hulusi Behçet (1889–1948), (Fig. 1), the Turkish dermatologist and scientist who first recognized the syndrome.

Figure 1. Hulusi Behçet (1889–1948)

Table I. Selected Eponyms in the dermatology literature linked to oral disorders

Figure 2. John Addison Fordyce (1858 - 1925)

Figure 3. Sir Jonathan Hutchinson (1828 – 1913)

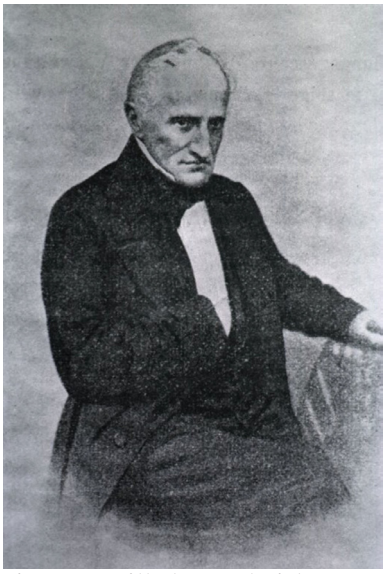

Figure 4. Wilhelm Frederick von Ludwig (1790 – 1865)

Eponyms in the dermatology literature linked to oral disorders	Remarks
Fordyce's spots [3]	The spots are a form of ectopic sebaceous gland which may occur on the lips and other body sites. Named after an American dermatologist, John Addison Fordyce (1858-1925), (Fig. 2).
Hutchinson teeth [4]	Hutchinson's teeth are a sign of congenital syphilis. It is named after Sir Jonathan Hutchinson (1828-1913), (Fig. 3), an English surgeon and pathologist, who first described them.
Ludwig angina [5,6]	Wilhelm Frederick von Ludwig (1790-1865), (Fig. 4), a German physician first described in 1836 a potentially fatal, rapidly spreading soft tissue infection of the neck and floor of the mouth. The condition was later named „Ludwig's angina", a term which persists in medicine to this day.
Miescher's cheilitis [7]	Miescher's cheilitis is another less commonly used name for Granulomatous cheilitis. Miescher's cheilitis is named for Alfred Guido Miescher (1887-1961), (Fig. 5), who was an Italian-born Swiss dermatologist. Granulomatous cheilitis or cheilitis granulomatosa is a monosymptomatic form of the Melkersson–Rosenthal syndrome (MRS). MRS is characterized by a triad of symptoms, typically with an onset in childhood or youth. It comprises recurrent facial paralysis (in 30% of cases), chronic edema of face and lips and fissured tongue (lingua plicata). MRS was described by Melkersson in 1928 and, Rosenthal in 1931 emphasized that lingua plicata (fissured tongue) is commonly related. However, there are several earlier descriptions of the condition-by Paul Hübschmann (1894), Lothar von Frankl-Hochwart (1891) and Grigorii Ivanovich Rossolimo (1901). Ernst Gustaf Melkersson (1898-1932) was born and educated in Sweden. Later, he worked at the medical department of the Gothenburg Sahlgrenska sjukhuset. Curt Rosenthal (1892-1937), was born in Germany and worked at the University of Breslau psychiatry and neurology clinic. The designation Melkersson's syndrome was suggested to honor Melkersson, who had died so young, but the term Melkersson–Rosenthal syndrome has now been generally accepted.

Table I. Selected Eponyms in the dermatology literature linked to oral disorders (continued)

Figure 5. Alfred Guido Miescher (1887-1961)

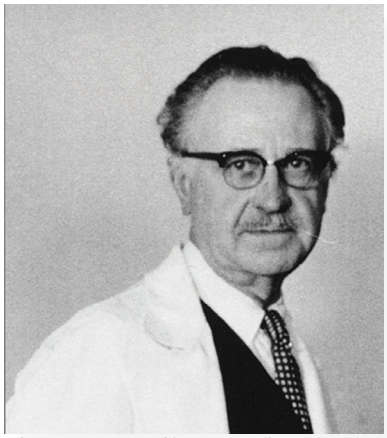

Figure 6. Henrik Samuel Conrad Sjögren (1899-1986). A courtesy of the South Swedish Society for the History of Medicine

Eponyms in the dermatology literature linked to oral disorders	Remarks
Sjögren's syndrome [8]	Sjögren syndrome (SS) is a chronic autoimmune disease - an inflammatory exocrinopathy - affecting mainly postmenopausal women (80-90%) or younger women after artificial menopause. It is named for, Henrik Samuel Conrad Sjögren (1899-1986) (Fig. 6), a Swedish ophthalmologist. SS is also known as, Gougerot-Houwer-Sjögren syndrome, Gougerot--Sjögren syndrome, Sjögren disease and von Mikulicz-Gougerot-Sjögren syndrome. In 1925, Henri Gougerot (1881-1955), a French dermatologist, described three cases of salivary gland atrophy associated with dry eyes, mouth and vagina. Houwer (1927) and Wisssmann (1932) noted the joint occurrence of keratoconjunctivitis sicca and arthritis. Sjögren in 1933 published the complete disease picture.Sjögren described his syndrome in 1933 in his doctoral thesis „Zur Kenntnis der keratoconjunctivitis sicca". Jan Mikulicz-Radecki (German: Johann von Mikulicz-Radecki) (1850-1905), was a Polish-Austrian surgeon. His name is also associated with one of the eponyms of this syndrome.
Stevens–Johnson syndrome [9]	Also known as erythema multiform major. It is characterized by mucous membranes erosions. The main known cause is certain medications, followed by infections and, rarely, cancers. Stevens–Johnson Syndrome is named for Albert Mason Stevens and Frank Chambliss Johnson, American pediatricians who jointly published a description of the disorder.
Takahara disease [10]	This is another name for Acatalasemia, a rare disease in which the enzyme catalase is deficient in the liver, muscles, bone marrow, erythrocytes, and skin. The absence of catalase leads to progressive gangrene of the mouth, with recurrent ulcerations resulting from increased susceptibility to infection by anaerobic organisms. In 1948, Dr. Shigeo Takahara (1908-1994), a Japanese otolaryngologist first reported this disease.
Van Der Woude syndrome (VDWS) [11]	It is a genetic disorder characterized by the combination of lower lip pits, cleft lip with or without cleft palate, and cleft palate alone (CP). The association between lower lip pits and cleft lip and/or palate was first described by Anne Van der Woude in 1954.
Vincent disease [12]	This is a synonym for Acute necrotizing ulcerative gingivitis , a disease characterized by a rapid onset of characteristic punched-out ulcerations appearing on the interdental papillae and marginal gingivae. The lesions may spread rapidly and involve entire respiratory tract. There is a characteristic foul, fetid odor that is always present. It is named after French physician Jean Hyacinthe Vincent (1862–1950), (Fig. 7).
Wickham striae [13]	Wickham striae are whitish lines visible in the lesions of lichen planus. Named after, Louis Frédéric Wickham (1861-1913), (Fig. 8), a French physician and pathologist.

Table I. Selected Eponyms in the dermatology literature linked to oral disorders (continued)

Figure 7. Jean Hyacinthe Vincent (1862 – 1950)

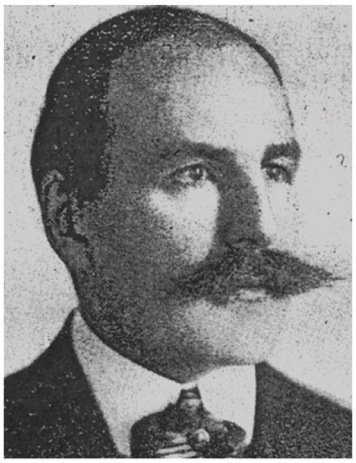

Figure 8. Louis Frédéric Wickham (1861 – 1913)

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NAMES OF „LINES" IN DERMATOLOGY LITERATURE

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The aim of this communication is to shed some lights on the names of some of the „lines" encountered in dermatology, which are summarized in Table I.

Names of "Lines", in dermatology literature	Remarks
AA lines [1]	Linear, slightly depressed transverse line of the upper part of the forearm. Named by the team who reported it as AA (antecubital Ahn's) lines. Ahn is the first author of the report.
Beau's lines [2]	Beau's lines are deep grooved lines in the nails. They are named after, a French physician, Joseph Honoré Simon Beau (1806–1865), who first described it in 1846.
Bunny lines [3]	Bunny lines are the subtle crinkly wrinkles on both sides of the nose. They get their name from the cute way bunnies scrunch up their noses.
Futcher's lines or Voigt's lines or Futcher-Voigt's lines or Ito lines [4,5]	These are less common names for what is known as Pigmentary demarcation lines (PDL). PDL are physiological abrupt transition lines from areas of deeper pigmentation to the area with less pigmentation. Five types (A-E) have been described. More recently; facial lines F, G and H were added. Type A (Futcher's / Voigt's lines), the most common lines, seen over the dorso-ventral aspect of the arms. These lines are named after Howard Palmer Futcher (1910-2004), (Fig. 1) an American-Canadian physician. He was the former member of the faculty of the Johns Hopkins University School of Medicine, executive director of the American Board of Internal Medicine.  **Figure 1. Howard Palmer Futcher (1910-2004). Reproduced from reference number 4**

Table I. Selected Names of „Lines", in dermatology literature

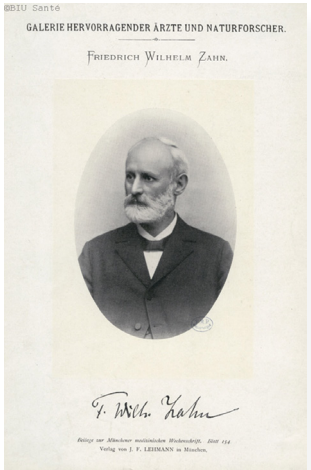
Names of "Lines", in dermatology literature	Remarks
Hart's line [6]	Natural demarcation between the labia minora and the limits of the vestibule. It is named after David Berry Hart (1851-1920), who was a Scottish surgeon.
Langer's lines [7,8]	Langer's lines, sometimes called cleavage lines, are topological lines drawn on a map of the human body. The lines were first described in 1861 by Austrian anatomist Karl Langer (1819-1887). However, for skin incision, Kraissl preferred lines oriented perpendicular to the action of the underlying muscles. Later, Borges described relaxed skin tension lines, which follow furrows formed when the skin is relaxed and are produced by pinching the skin. Some authors think that Borges's and Kraissl's lines (not Langer's) may be the best guides for elective incisions of the face and body, respectively.
Lines of Blaschko [1,9]	Imaging lines represent a pattern followed by many skin disorders. It is named after Alfred Blaschko (1858-1922), (Fig. 2), a German dermatologist.  **Figure 2. Alfred Blaschko (1858-1922). A courtesy National Library of Medicine**
Lines of Zahn [10]	Lines of Zahn are microscopic thing which can be seen in dermatopathology. They are a characteristic of thrombi. They have visible and microscopic alternating layers (laminations) of platelets mixed with fibrin, which appear lighter and darker layers of red blood cells.They are been named after German pathologist Friedrich Wilhelm Zahn (1845-1904), (Fig. 3).  **Figure 3. Friedrich Wilhelm Zahn (1845-1904). Courtesy of BIU Sante (Paris) Available online from; http://www2.biusante.parisdescartes.fr/**
Marionette lines [11]	Melomental folds, or marionette lines, are one of the consequences of facial aging. The curvilinear wrinkles formed because of facial movements and the aging process extends downward from the oral commissures. These lines are referred to as „Marionette Lines" named after the classic marionette puppets as they look like the mouth pieces of a ventriloquist's doll.
Mees' lines [2]	Also called Aldrich-Mees' lines or leukonychia striata. They are lines of discoloration across the nails of the fingers and toes. They are named after Dutch physician R.A. Mees, who described the abnormality in 1919. However earlier descriptions of the same abnormality were made by Englishman E.S. Reynolds in 1901and by American C.J. Aldrich in 1904.

Table I. Selected Names of „Lines", in dermatology literature (continued)

Names of "Lines", in dermatology literature	Remarks
Muehrcke lines [2]	Muehrcke's lines are white lines (leukonychia) that extend all the way across the nail and lie parallel to the lunula. Muehrcke's lines were described by Robert C. Muehrcke (1921 -2003), an American Physician in 1956.
Pastia's line [12]	Transverse red streaks in the skin folds due to capillary fragility in patients with scarlet fever it is named after a Romanian physician, Constantin Chessec Pastia (1883-1926).

Table I. Selected Names of „Lines", in dermatology literature (continued)

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EPONYMS IN DERMATOLOGY LITERATURE LINKED TO FINLAND

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Finland is a Nordic country situated in the Fennoscandian region of Northern Europe. The population of Finland is currently about 5.4 million [1]. It is developed in many fields, and particularly in education.

This year in the 2013 Reporters Without Borders World Press Freedom Index, and for the third year running, Finland has distinguished itself as the country that most respects media freedom.

There are names in medicine linked to Finland. These names might be after a place or after scientists from Finland [1].

For instance, Aland Island eye disease (AIED), also known as Forsius-Eriksson syndrome, is an X-linked recessive retinal disease characterized by a combination of fundus hypopigmentation, decreased visual acuity, nystagmus, astigmatism, protan color vision defect, progressive myopia, and defective dark adaptation. Electroretinography reveals abnormalities in both photopic and scotopic functions. The gene locus for AIED has been mapped to the pericentromeric region of the X-chromosome [2].

It is named after Henrik Forsius, Finnish ophthalmologist and Aldur Wictor Eriksson, Finnish human geneticist.

It is named Åland Island eye disease because it is reported first and common in Åland which is a group of islands in the Bay of Finland, between Finland and Sweden.

However, one of the commonest eponym linked to Finland mentioned in dermatology literature and the literature of medicine in general is Von Willebrand's disease (vWD). vWD is the most common inherited bleeding disorder. It is characterized by a deficiency in the clotting protein called von Willebrand's Factor; the most common symptom is prolonged bleeding time. The clotting protein Factor VIII may also be involved.

vWD may present with cutaneous bruising and/or bleeding. However the latter may be a manifestation of a hereditary or acquired qualitative or quantitative platelet disorder, disturbance of the vascular or supporting structure, or it may be due to one of several acquired systemic disorders3.

vWD is named after Erik Adolf von Willebrand (1870-1949) [4-7].

Erik Adolf von Willebrand (Fig. 1) is a Finnish internist, born in Vasa; a seaport city located in western Finland and died in, Pernå. He discovered the most common inherited bleeding disorder while studying the genetic traits of a family in the Åland Islands in Finland [4-7].

Von Willebrand published two papers on Physiology and Clinical Management in Treatment with Hot Air. Throughout his lifetime he maintained his interest in the latter form of treatment as well as in metabolic disorders and haematological problems. He focused on blood changes during muscular exercise, metabolism and obesity, as well as carbon dioxide and water exchange through the human skin. Von Willebrand wrote many articles about obesity, gout and diabetes mellitus. He detailed a technique for evaluating ketone bodies in urine in 1912. He wrote about managing diabetes with diet, and he was a pioneer in insulin use. In 1922, von Willebrand wrote about using insulin to treat diabetic coma. He was the author of several hematology articles as well [5].

Von Willebrand remains most famous, however, for his description of vWD. A disease he encountered among the inhabitants of the Åland Islands.

In 1925, he examined a 5-year-old girl with a history of bleeding who had been brought to Helsinki for treatment [5]. The little girl was the ninth of 12 children. Four of her siblings bled to death at an early age. Both of her parents came from families with bleeding disorders.

Von Willebrand was curious to know more, so he traveled to the Åland Islands to study the disease in depth. He mapped the family pedigree and found that 23 of the 66 family members had bleeding problems. Von Willebrand concluded that this was a previously unknown type of hemophilia. Initially, he called the disease „hereditary pseudo-hemophilia" because of the prolonged bleeding time. As he studied the disease more, he came to believe that platelets were involved, so he renamed it „constitution-al thrombopathy". He noted his findings about the family in a 1926 report [5].

In 1994, the Åland Islands issued a postal stamp to honor von Willebrand's work [5].

Figure 1. Erik Adolf von Willebrand (1870-1949). A courtesy of Helsinki University Museum

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NASZA DERMATOLOGIA Online
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DERMATOLOGY EPONYMS – SIGN – LEXICON – (I)

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Abstract
Eponyms are used almost daily in the clinical practice of dermatology. And yet, information about the person behind the eponyms is difficult to find. Indeed, who is? What is this person's nationality? Is this person alive or dead? How can one find the paper in which this person first described the disease? Eponyms are used to describe not only disease, but also clinical signs, surgical procedures, staining techniques, pharmacological formulations, and even pieces of equipment. In this article we present the symptoms starting with (I) and other. The symptoms and their synonyms, and those who have described this symptom or phenomenon.

Key words: eponyms; skin diseases; sign; phenomenon

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„I LOVE YOU" SIGN

The appearance of this hand is very typical of infants with trisomy 18, occurring in about 50% of affected infants. The clenched hand with a tendency for the index finger to overlap the third and for the fifth finger to overlap the fourth. At times these fingers are extended, giving the appearance of the sign for "I love you" in American sign language (Fig. 1). Infants with trisomy 18 also commonly have hypoplasia of the nails on both the fingers (especially the fifth finger) and the toes [1].

IDIOPATHIC DISEASE SIGN

One not consequent upon other disease, nor upon any known lesion or injury.

INMAN'S SIGN

=myalgia [2]

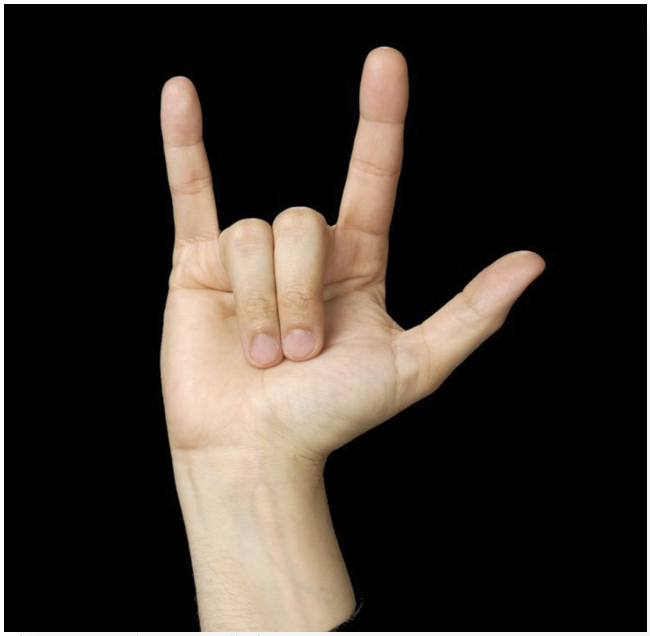

Figure 1. „I love you" sign

THOMAS INMAN

English physician, 1820-1876. House-surgeon to the Liverpool Royal Infirmary. In his lifetime he had numerous medical papers published. He was also an amateur mythologist, and wrote Ancient Pagan and Modern Christian Symbolism, first published in 1869 and then again in 1875. He entered King's College, London, where he graduated M.B. in 1842 and M.D. in 1844 at the University of London. Declining a commission as an army surgeon, Inman settled in Liverpool as house-surgeon to the Royal Infirmary. He obtained a good practice as a physician, and was for many years physician to the Royal Infirmary. Inman's publications on personal hygiene are practical advice.

INNOCENCE'S SIGN

In the estern region of Nigeria natives used the extract of the Calabar bean (*Physostigma vevevosum*) (Fig. 2) which is the seed of a leguminios plant for judical execution. However, if after ingestion the man vomited it back, then he was considered innocent [3].

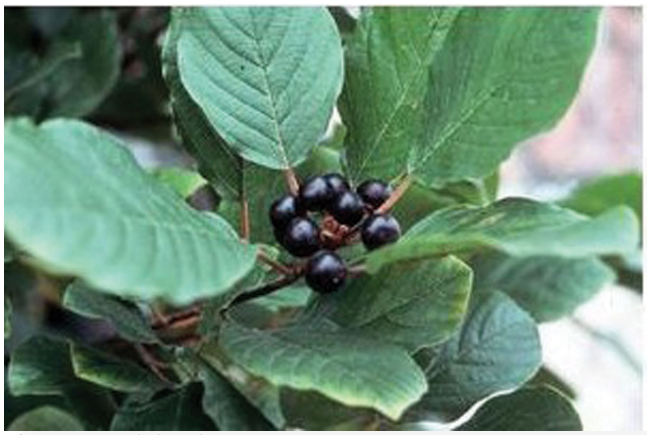

Figure 2. Calabar bean

DOUGLAS MORAY COOPER LAMB AGRYLL ROBERTSON

Figure 3. Douglas Moray Cooper Lamb Agryll Robertson

Scottish ophthalmologist and surgeon, 1837-1909 (Fig. 3). After earning his degree in 1857 from the University of St Andrews, he went to Berlin to study under Albrecht von Graefe. Robertson spent most of his medical career in Edinburgh as an eye surgeon at the Edinburgh Royal Infirmary and teacher of ophthalmology at the University of Edinburgh. For a while he was honorary eye physician to Queen Victoria and King Edward VII. Robertson made several contributions in the field of ophthalmology; in 1863 he researched the effects on the eye made by physostigmine, an extract from the Calabar bean (Physostigma venenosum), which is found in tropical Africa. He correctly predicted that physostigmine would become very important in the treatment of eye disorders. He also described a symptom of neurosyphilis that affects the pupils of the eye, which is known today as Argyll Robertson pupils [4].

INTERCURRENT DISEASE SIGN

A disease occuring during the course of another disease with which it has no connection.

INTERFERENCE SIGN

1. The interference of one drug with the therapeutic activity of another drug; especially a sort of drug-fastness toward full therapeutic doses of one drug conferred on a parasite by subtherapeutic doses of another drug.
2. The interference with the replication or virulence of a virus by the simultaneous infection with another that mayor may not be related. Called also preemptive immunity or interference phenomenon.

ITALIAN SIGN

=syphilis. Also called mal d'Italie.

HIERONYMUS FRACASTORIUS

Figure 4. Hieronymus Fracastorius

Physician, astronomer, a naturalist, a poet and a philosopher, 1483-1553 (Fig. 4). The poem Syphilis sive morbus gallicus was published by Fracastor in 1530.

In De morbis contagiosis, printed in Venice in 1546, Fracastor describes the cause of syphilis and appears as a precursor of bacteriology. Fracastor was born in 1483 within a well-known medical family. He studied the Fine Arts, Mathematics and Medicine in Padova [5-9].

ISOMORPHIC SIGN
Development of lesions after injury [10-12].

BIEDERMAN SIGN
A dusky redness of the lower portion of the anterior pillars of the fauces in certain cases of syphilis [13].

JOSEPH B. BIEDERMAN
Australian physician, (1907-…).

BITOT SIGN
Bitot's spots are the buildup of keratin debris located superficially in the conjunctiva, which are oval, triangular or irregular in shape. These spots are a sign of vitamin A deficiency and are associated with conjunctival xerosis. In 1863, Pierre Bitot, first described these spots [14].

PIERRE ALAIN BITÔT
French physician, anatomist and surgeon, (1822-1888). He attended medical school in Bordeaux, qualifying in 1846. He gained his M.D. in 1848 from the faculty of Paris, and joined the anatomy department in Bordeaux. He became Professor of anatomy in 1854, and gained his Chirurgien des Hôpitaux in 1878. Bitôt published on a wide variety of topics, ranging from hare lip to studies of the best form of ligatures to use in limb amputations, the use of quinine sulphate to prevent fever following blood transfusions, as well as some aspects of cerebral anatomy and function [15].

BORSIERI SIGN
Positive if white line that results when fingernail drawn across skin subsequently turns red; seen in early scarlet fever [16,17].

GIOVANNI BATTISTA BORSIERI DE KANIFELD
Italian parasitologist, (1725-1785). Physician and medical writer. Pupil of Morgagni and Vallisnieri, taught medical clinics at the University of Pavia. He wrote, in Latin, a very popular book, Institutiones medicinae practicae (1781–1789), which was later translated into Italian and English. Borsieri recommended the use of emetic tartar against tapeworms; also, in De anthelminthica argenti vivi facultate (1753) he reported on the successful use of mercury for the expulsion of roundworms.

DEMARQUAY SIGN
Absence of elevation of the larynx during deglutition, said to indicate syphilitic induration of the trachea.

JEAN NICHOLAS DEMARQUAY
French surgeon, 1814–1875 (Fig. 5). In 1863, became the first to record the observation of microfilariae in fluid extracted from a hydrocoele (another common symptom of lymphatic filariasis) [19].

Figure 5. Jean Nicholas Demarquay

FROSTED GLASS SIGN
syn. Hair in the eye sign
Inflamed and thickened eyelids which curl in upon themselves, inverting the eyelashes, which begin to scratch the cornea causing a frosted glass appearance and blindness. An indication of infection by zoonotic Chlamydia trachomatis transmited by the fly known as Musca sorbens. Also known as Frosted Glass sign [9].

HONEYCOMB SIGN CELSI

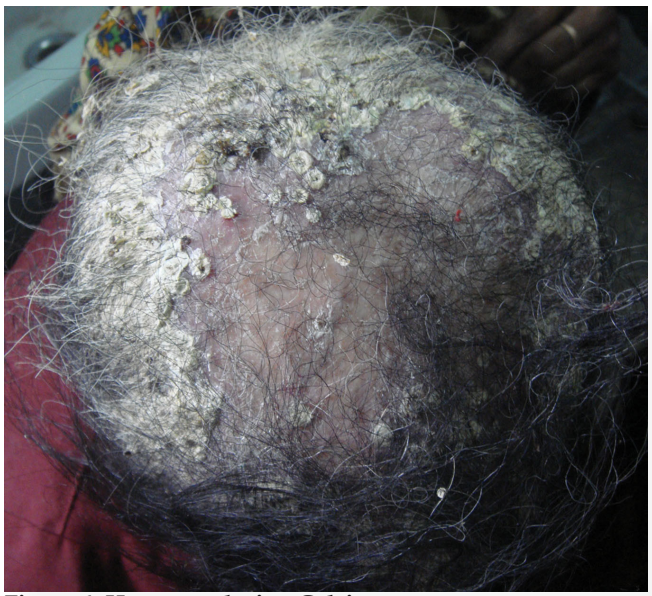

Figure 6. Honeycomb sign Celsi

There are three type of tinea capitis, microsporosis, trichophytosis, and favus (Fig. 6). Favus is caused by *T. schoenlenii*, and is endemic in South Africa and the Middle East. It is characterized by a number of yellowish, circular, cup-shaped crusts (scutula) grouped in patches like a piece of honeycomb, each about the size of a split pea, with a hair projecting in the center. These increase in size and become crusted over, so that the characteristic lesion can only be seen around the edge of the scab [20-22].

AULUS AURELIUS CORNELIUS CELSUS

(25 BC-AD 50) was a Roman writer on medicine and surgery (Fig. 7). He wrote several works, of which only one remains entire, his treatise De Medicina in eight books. Probably lived in Gallia Narbonensis.
(ur. ok. 25 p.n.e., zm. ok. 50), pisarz rzymski w medycynie i chirurgii. Napisał wiele dzieł, z których tylko jedno pozostaje kompletne jego traktat De Medicina w ośmiu ksią?kach. Mieszkał prawdopodobnie w Gallia Narbonensis [6].

Figure 7. Aulus Aurelius Cornelius Celsus

GERDY SIGN

In typical plaques of alopecia areata, the diagnosis is easy to do, because these features are clear: precise edges, glowing skin and hair with orthostatic around as an exclamation point [23].

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We are pleased to extend a warm welcome to the 23RD World Congress of Dermatology (23RD WCD), to be held in Vancouver, Canada from June 8-13, 2015. Held under the auspices of the International League of Dermatological Societies, the 23RD WCD will be the largest international gathering of dermatologists and people dedicated to skin health from all sectors. Our vision for the world's premier dermatology conference includes celebration, innovation, and inclusiveness. Our award-winning world class Vancouver Convention Centre will serve as one of the most beautiful venues to ever host the WCD. Strategically situated on the waterfront in the heart of downtown Vancouver, participants will enjoy spectacular views of the harbour and mountains as they move between their sessions. This unique convention centre is within walking distance of a spectacular variety of accommodation, dining, shopping, tourist attractions, and transportation.

We look forward to celebrating with you in Vancouver, where the world of dermatology will gather in 2015.

Dr. Jerry Shapiro and Dr. Harvey Lui
President and Secretary-General

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