

ERYTHEMATODES CHRONICUS PROFUNDUS AS DERMATOLOGY, SURGERY AND COSMETOLOGY PROBLEM

ERYTHEMATODES CHRONICUS PROFUNDUS JAKO DERMATOLOGICZNY, CHIRURGICZNY I KOSMETOLOGICZNY PROBLEM

Alendar Faruk, Soskic Samra, Helppikangas Hana,
Gavrankapetanovic Alma, Alendar Temeida

*Dermatovenerology Department Clinical Center University of Sarajevo,
Bosnia & Herzegovina* derm_alendar@hotmail.com

N Dermatol Online. 2011; 2(3): 141-143

Date of submission: 30.03.2011 / acceptance: 22.04.2011

Conflicts of interest: None

Abstract

Lupus profundus is a rare skin condition originally described by Kaposi in 1883. It is a clinical variant of LE in which the deep dermis and subcutaneous fat are predominantly affected. A 38-year-old female presented with history of development of well-circumscribed patches of hair loss since 1994 year. In 2004 on the skin of left cheek she noticed a single erythematous plaque on which place later became dint of subcutaneous tissue. Today she has massive defects across whole scalp, right cheek and the upper part of left arm with multiple ulcers (fig.3 and 4). case report, laboratory examinations, skin biopsy and influence of corticosteroids on further progression / regression of this disease.

Streszczenie

Lupus Profundus jest rzadkim schorzeniem skóry pierwotnie opisanym przez Kaposiego w 1883 roku. Jest to wariant kliniczny LE, w którym zmiany zlokalizowane są w przeważającym stopniu głęboko w skórze właściwej i tkance podskórnej. 38-letnia chora zgłosiła się z historią rozwiniętych, ograniczonych plackowatych ognisk utraty włosów od 1994 roku. W 2004 r. na skórze lewego policzka zauważyła pojedynczą rumieniową zmianę, w której miejscu później powstało wgłębienie do tkanki podskórnej. Obecnie pacjentka z rozległymi ubytkami na całej skórze głowy, policzkach i w górnej prawej części lewego ramienia, z wieloma owrzodzeniami. Opisano przypadek kliniczny, wykonane badania laboratoryjne, diagnostyczne oraz wpływ kortykosteroidów na dalszy rozwój / regresję choroby.

Key words: lupus profundus; chloroquin; corticosteroids

Słowa kluczowe: lupus profundus; chlorochina; kortykosteroidy

Introduction

Lupus profundus is a rare skin condition originally described by Kaposi in 1883 and later by Irgang in 1940. It is a clinical variant of LE in which the deep dermis and subcutaneous fat are predominantly affected. It may occur on its own or in association with discoid lupus erythematosus (DLE) or SLE. In cases associated with SLE, it may precede SLE by several years. In the majority of cases, it tends to have a mild chronic course marked by recurrent nodules or plaques.

Case report

A 38-year-old female presented with history of development of well-circumscribed patches of hair loss

since 1994. year. The hair loss was rapid and asymptomatic, with no hair re-growth.

There was no family history of any autoimmune rheumatic diseases.

In 2004. she was first time hospitalised on our clinic. Clinical symptoms were: on the skin of left cheek a single, erythematous plaque (Fig. 1). On left arm two small ulcers size 1 x 2 cm, oval in shape and firm in consistency (Fig. 2).

Since than she was treated with chloroquin in dose 250mg per day with gradually reduction, but with no results.

Today she have massive defects across whole scalp, right cheek and the upper part of left arm (Fig. 3,4).



Figure 1. On the skin of left cheek a single, erythematous plaque



Figure 2. On left arm two small ulcers size 1 x 2 cm, oval in shape and firm in consistency



Figure 3. Massive defects across whole the upper part of left arm



Figure 4. Massive defects across whole scalp, right cheek

The lesion, on the left arm in 2004, initially, was about 1 X 2 cm² in size, oval in shape and firm in consistency. It gradually increased in size to about 8 X 5 cm², in 2010, with multiple ulcers colonized with *Staphylococcus aureus*, and with extensive atrophy of cutaneous and subcutaneous tissue. There was no history of preceding trauma. No history of taking injection. She denied arthralgia, arthropathy, myalgia, fatigue, fever, Raynaud's phenomenon and gastrointestinal symptoms. She notifies pain in right lumbal area, and the ultrasound showed signs of bilateral nephropathy.

The following laboratory tests were done: complete blood cell count, differential cell count, ESR was 25, total serum proteins 86.0, with A:G ratio 1.0, y globulins were increased 0.27. Blood sugar, blood urea, serum creatinine, serum electrolytes, liver function tests and urinalysis were in normal rates. X-ray chest was normal. Rheumatoid factor was normal, but antistreptolysin was 221,0IU/ml. C3 1.090 g/l, C4 0.165 g/l, ANA positive, Anti ds DNA negative and CiC was increased and his amount was 58.08IU/ml.

Skin biopsy showed lymphocytic panniculitis, hyaline degeneration of the fat, hyaline papillary bodies, and lymphoid nodular structures in the lower dermis and subcutaneous tissue. Result is in high correlation with diagnosis of erytematodes cronicus profundus.

In 2010, we treated her with systemic corticosteroid therapy in dose of 60mg per day with gradually reduction of dose. After 2 month of therapy further progression has stoped (Fig. 5,6). Her maintenance dose today is 20 mg prednisolon/day.

Discussion

Lupus erythematosus profundus or panniculitis is an unusual but distinct clinical variety of lupus erythematosus. The inflammatory reaction in takes place primarily in the deep corium and the subcutaneous tissues leading to deep indurated nodules or sharply defined plaques. The overlying skin usually appears normal but there may be erythema, atrophy, ulceration or poikilodermatous or hyperkeratotic changes.



Figure 5. Lesions after 2 month of therapy



Figure 6. Lesions after 2 month of therapy

The lesions are most frequent on cheeks but other sites of predilection are face, upper arms, hands, chest, buttocks and thighs. It may develop in association with discoid lupus erythematosus or systemic lupus erythematosus or may occur as an isolated phenomenon. The differential diagnoses include panniculitis due to other connective tissue disorders like dermatomyositis or scleredema and Weber Christian panniculitis or Jessner's lymphocytic infiltration, lymphocytoma cutis and sarcoidosis.

Lupus panniculitis often responds to treatment with antimalarials, such as hydroxychloroquine (200 mg once

or twice a day). Some cases respond to a combination of antimalarials (for example, hydroxychloroquine 200 mg and quinacrine 100 mg daily) when monotherapy is ineffective. Systemic glucocorticoids should be reserved for widespread and resistant lesions. Intralesional glucocorticoids are usually ineffective and may exacerbate the atrophic healing process. Success with dapsone, azathioprine, and thalidomide has been described in isolated case reports. Surgical debridement or resection of individual lesions may be attempted when all other modalities have failed and there is appreciable debilitation.

Conclusion

Progress of Erythematodes profundus is unpredictable. As we can see on example of this patient therapy with cloroquine was insufficient to stop progression of disease.

With dose of 60 mg methylprednisolon there was no more further progression. Today, after 8 months of last hospitalisation we have not noticed any progression.

Defects on skin with which she needs to live daily, are horrifying, what caused her psychiatric problem and antidepressive therapy.

Today she stay as dermatology, surgery and cosmetology problem.

REFERENCES / PIŚMIENNICTWO:

1. Tuffanelli DL: Lupus erythematosus panniculitis (profundus). *Arch Dermatol* 1971; 103: 231-242.
2. Black MM, Cunliffe WJ: Inflammatory disorders of subcutaneous fat. In: Champion RH, Burton JL, Burns DA, et al., editors. *Textbook of Dermatology*. London: Blackwell Science LTD; 1998.
3. Braun-Falko O, Wolf HH: *Dermatology and venerology*, New York; 1997.
4. Sanchez NP, Peters MS, Winkelmann RK: The histopathology of lupus erythematosus panniculitis. *J Am Acad Dermatol*. 1981; 5: 673-680.
5. Ahmed I, Ahmed D. Lupus erythematosus panniculitis: a unique subset within the lupus erythematosus spectrum. *Am J Dermatopathol*. 2000; 22: 352.
6. Kündig TM, Trüeb RM, Krasovec M: Lupus profundus/panniculitis. *Dermatology*. 1997; 195: 99-101.
7. Diaz-Jouanen E, DeHoratius RJ, Alarcon-Segovia D, Messner RP: Systemic lupus erythematosus presenting as panniculitis (lupus profundus) *Ann Intern Med*. 1975; 82: 376-379.
8. Masood Q, Manzoor R: Lupus erythematosus profundus (panniculitis) *J K Practitioner*. 1995; 2: 135-136.