Our Dermatology Online

Letter to the Editor

Facial porokeratosis - Rare presentation of an uncommon disease

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Sir,

Porokeratoses are a group of hereditary or acquired disorders of keratinization. Several types have been described. Disseminated superficial actinic porokeratosis (DSAP) is the most common presentation of the disease. Facial lesions have been seen in 15% of the DSAP patients while majority of the reported cases involve extrafacial sites such as the extensor surfaces of the extremities. However, Exclusive facial presentation of DSAP has rarely been reported. In this article, the authors report a 20 year female with exclusive facial porokeratosis.

A 20 year old female visited our dermatology outpatient department (OPD) for multiple brownish lesions on the face for the last 3 months. There is no history of photosensitivity. There is no history of such lesion in her family members. There is no excessive sun exposure during these 3 months with respect to her routine work. There was no history of recurrent infections. Dermatological examination revealed multiple brownish hyperkeratotic macules of size 2-8 mm present over cheeks and infraorbital area on both sides with majority of lesions on the right side of face [Figure 1]. Most of the lesions had dense hyper pigmented (brownish) and hyperkeratotic borders with less pigmented atrophic centres. The examination of hair, mucosae and nails was normal. Systemic examination was unremarkable. Her laboratory investigations were within normal limits. Histopathology of skin biopsy revealed hyperkeratosis, columns of parakeratotic cells (coronoid lamellae) overlying an interrupted stratum granulosum and dyskeratotic keratinocytes which confirmed the diagnosis of porokeratosis. She was prescribed topical tazarotene and sunscreens. Though initially there was some benefit with respect to hyperpigmentation and scaling, but at 3 months follow up her lesions were persistent with minimal improvement.

Porokeratoses, a disorder of epidermal keratinization, includes a heterogeneous group of disorders that are mostly inherited in an autosomal dominant fashion [1]. Several clinical variants have been described which include classic porokeratosis of Mibelli, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, porokeratosis palmaris et plantaris disseminata and porokeratosis punctata [2]. Other uncommon variants are giant porokeratosis, porokeratosis ptychotropica and porokeratoma, Porokeratotic Eccrine Ostial and Dermal Duct Nevus (PEODDN), punched-out, hypertrophic verrucous and reticulate porokeratosis [3,4]. Most of these variants are clinically characterized by a thin hyperkeratotic rim bordering a slightly atrophic macule or plaque with a tendency to extend in a centrifugal fashion [5]. DSAP is the most common clinical variant of the disease with multiple lesions predominantly in sun-exposed sites in middle-aged individuals [6]. Majority of the lesions occur on extrafacial sites including extremities with only 15% cases having lesions on face. Lesions of DSAP exclusive to face are rare and hence reported [7].

Various conditions have been associated with porokeratosis like Psoriasis and phototherapy (UVA, NB-UVB and BB-UVB). In addition, DSAP has also been linked with HIV infection, following administration of

How to cite this article: Arif T, Amin SS, Adil M. Facial porokeratosis - Rare presentation of an uncommon disease. Our Dermatol Online. 2016;7(3):353-354.
Submission: 19.01.2016; **Acceptance:** 24.04.2016
DOI: 10.7241/ourd.20163.97

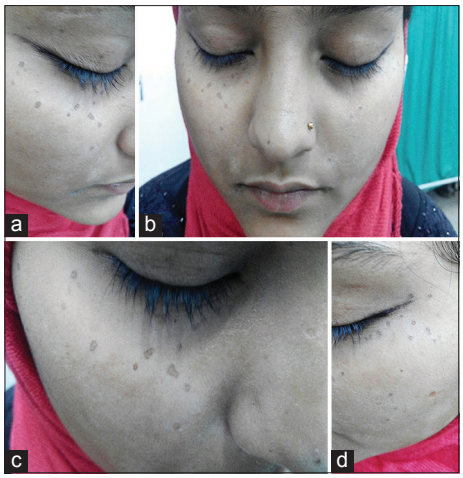

Figure 1: (a,b,c,d) Multiple 2-8 mm brownish macules with well marginated hyperkeratotic and hyperpigmented borders with less pigmentation at the centre.

immunomodulating drugs used to treat autoimmune diseases and after organ transplantation, diabetes mellitus, liver cirrhosis, acute pancreatitis, solid malignancy, Crohn's disease, etc. However our case had no systemic associations [8,9].

The histopathology is diagnostic in porokeratosis. The edge of the lesion shows characteristic microscopic findings. A hyperkeratotic stratum corneum, column of poorly staining parakeratotic keratinocytes termed as coronoid lamella with underlying interrupted granular layer and dyskeratotic cells are highly suggestive of porokeratosis. The central area of a lesion may be atrophic or normal or grossly hyperkeratotic [5,6].our case had similar findings on histopathology.

The treatment of porokeratosis is difficult.Various treatments have been tried which include cryotherapy, photodynamic therapy, erbium YAG and CO2 lasers, 3% diclofenac gel, 5- fluorouracil cream, Keratolytics, topical tacalcitol, imiquimod cream and oral etretinate [6,7]. However our case was prescribed topical tazarotene 0.05% gel to be applied at bed time on the lesions and topical sunscreens during the day. Initially the hyperkeratosis and the pigmentation improved. After a follow up of 3 months the lesions were persistent with minimal improvement in the lesions.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article and any accompanying images.

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Source of Support: Nil, **Conflict of Interest:** None declared.