Acta Dermatovenerol APA

Acta Dermatovenerologica
Alpina, Pannonica et Adriatica

2012;21:79-80
doi: 10.2478/v10162-012-0023-2

Localized actinic nasal porokeratosis: a case report

Arzu Ataseven[1] ✉, Perihan Öztürk[2], Nursel Dilek[3], Ilknur Küçükosmanoğlu[1]

Abstract

Porokeratisis is a specific keratinization disorder. The presence of cornoid lamella is histologically characteristic of the disorder. This report describes a 23-year-old male patient with multiple porokeratotic lesions with bilateral symmetric localization on the ala of the nose, which may be a rare variant of porokeratosis.

Received: 17 October 2012 | **Returned for modification:** 17 November 2012 | **Accepted:** 11 February 2013

Introduction

Porokeratosis is an epidermal keratinization disorder with an unclear etiopathogenesis. Cornoid lamella, a histopathological marker, results from abnormal proliferation of keratinocytes. This abnormal proliferation clinically causes distinctive lesions with ridge-like borders. The lesions tend to expand to the peripheral areas and leave an area of central atrophy (1). We present the case of a 23-year-old male patient with multiple porokeratotic lesions with bilateral symmetric localization on the ala of the nose.

Case report

A 23-year-old male patient had had a nasal lesion for 3½ years. He complained about a burning sensation during sun exposure. The past medical and family histories were unremarkable. A dermatological examination revealed symmetrical, four bilateral plaques on the ala of the nose, 0.8 to 2 cm in diameter and almost 1 mm in height, with hyperpigmented sharp margins and relatively atrophic and hypopigmented central regions (Figure 1).

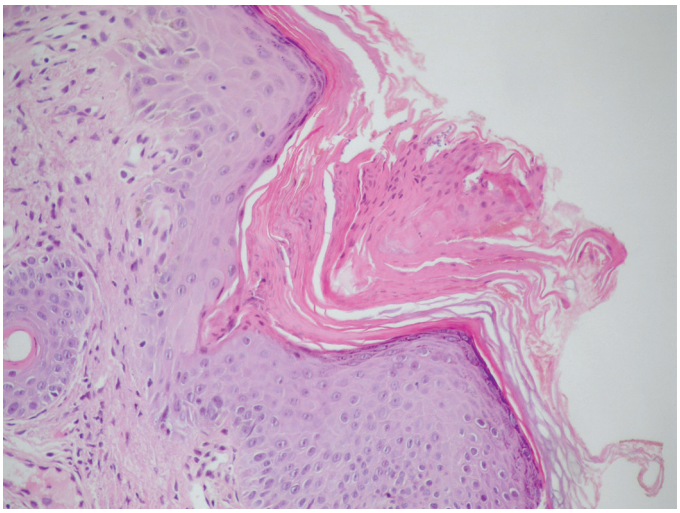

Figure 1 | The stratum corneum was seen as a parakeratotic column, without a visible granular layer in this area (cornoid lamella).

The epidermis appeared normal and the stratum corneum was orthokeratotic upon histopathological examination. The epidermis invaginated into the dermis in one area. The stratum corneum was seen as a parakeratotic column, without a visible granular layer in this area (cornoid lamella). Chronic inflammatory cell infiltration and a pilosebaceous unit were noted in the dermis (Figure 2). The laboratory results were as follows: hemoglobin: 6 g/dL, hematocrit: 23 g/dL, aspartate aminotransferase: 36 U/L. Other results such as complete blood count, biochemical parameters, erythrocyte sedimentation rate, C-reactive protein, and urine analysis were all within normal limits. Imiquimod 5% cream was prescribed for the patient; however, adequate relief was not obtained.

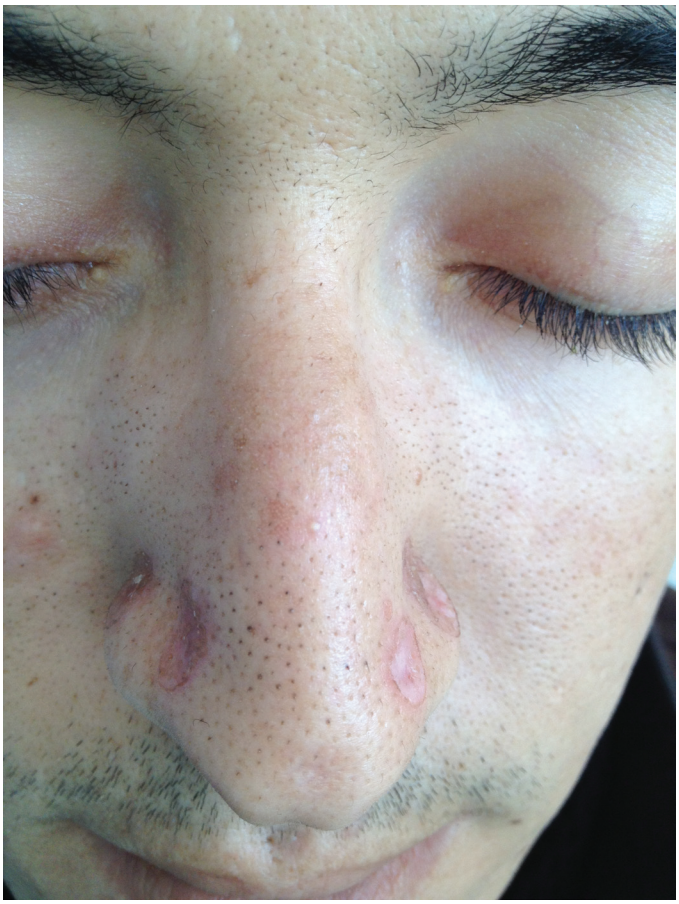

Figure 2 | Symmetric, four bilateral plaques on the ala of the nose, with hyperpigmented sharp margins and relatively atrophic and hypopigmented central regions.

Discussion

Mibelli reported porokeratosis for the first time in 1893 (2). There are six different types of porokeratosis: Porokeratosis of Mibelli (PM), giant porokeratosis, linear porokeratosis (LP), disseminated

[1]Department of Dermatology, Konya Education and Research Hospital, Konya, Turkey. [2]Department of Dermatology, Sütçü İmam University, Kahramanmaraş, Turkey. [3]Department of Dermatology, Rize University, Rize, Turkey. ✉ **Corresponding author: arzuataseven@hotmail.com**

superficial actinic porokeratosis (DSAP), palmoplantar porokeratosis (PPP), and punctate porokeratosis (PP) (3). DSAP is the most common type (4). Porokeratosis of Mibelli most often affects the limbs, particularly the hands and feet, the neck and shoulders, the face, and the genitals (5). Giant porokeratosis is a very rare form of porokeratosis and it is a morphological variant of PM (3). Linear porokeratosis can further be classified as localized, zosteriform, systematized, or generalized. The distribution of all of these variants follows Blaschko's lines, which may be explained by cutaneous mosaicism (6). The distribution of typical lesions is symmetrical and usually affects sun-exposed areas. The lesions generally spare the face, palms, soles, and mucosal surfaces (7). PPP initially occurs in the palmoplantar areas with subsequent involvement of the other areas of the body, including sites with sun exposure to ultraviolet radiation (8). Punctate porokeratosis is a rare variety of porokeratosis. It is characterized by seed-like punctate keratoses and/or pits on the palms and soles (9).

The disease tends to mostly affect the extremities. Less commonly, the body and the face can be affected. Isolated face involvement such as that seen in our patient is rarely reported. The type of porokeratosis of these cases were PM and DSAP. In one study, seven out of 197 cases of PM had primary facial lesions (10). Gutierrez et al. encountered only six cases of facial porokeratosis during 15 years of experience. These cases consisted of one male and five females, and the patients' age range was 16 to 30 years (11). Even though the age of our case, 23 years, was consistent with this report, the patient was male, whereas porokeratosis mostly affects females. Another patient with bilateral lesions on the ala of the nose similar to our case was reported by Ghorpade (4).

Porokeratosis can occur with a genetic predisposition and it can also be secondary to risk factors such as sun exposure, immune suppression, and ultraviolet exposure. The incidence of Bowen's disease and squamous cell and spinous cell carcinomas is 6.8 to 11% in this disease, and so follow-up is essential. The risk is greater if the patient has linear-type porokeratosis, or a large or long-standing lesion (11).

The etiology and pathogenesis of porokeratosis are obscure but certainly complex and multifactorial. It has been suggested that the lesions of porokeratosis result from the peripheral expansion of an abnormal, mutant clone of epidermal keratinocytes (which would be inherited) located at the base of the parakeratotic column (12).

The following disorders should be considered in the differential diagnosis of porokeratosis: elastosis perforans for PM, actinic keratosis and stucco keratosis for DSAP, lichen sclerosus et atrophicus, lichen planus, acrokeratosis verruciformis, Bowen's disease, and squamous cell carcinoma for single lesions (13). Discoid lupus erythematosus-like case can also be expressed with similar clinical presentations (14).

The histopathological presence of a cornoid lamella is often associated with a diagnosis of porokeratosis. However, this feature is not pathognomonic for porokeratosis and can be found in a number of other dermatological conditions, which include seborrheic keratosis, verruca vulgaris, actinic keratosis, squamous cell carcinoma in situ, basal-cell carcinoma, milia, and scars. Notably, none of these entities has an inflammatory etiology (15).

Keratolytic agents, topical 5-flourorasil, topical and oral retinoid agents, topical imiquimod, cryotherapy, photodynamic therapy, carbon dioxide laser application, dermabrasion, and excision are used for treatment (16). We preferred to prescribe topical imiquimod our patient for three months.

In conclusion, our case was a rare case of PM, with only symmetrical bilateral involvement of the ala of the nose. The case may be a rare variant of porokeratosis and the differential diagnosis of annular lesions should be borne in mind.

References

1. Lembo S, Panariello L, Nugnes L, Lembo C, Alaya F. Porokeratosis: two faces, one family. Case Rep Dermatol. 2009;1:52-5.
2. Mibelli V. Contributo allo studio della ipercheratosi dei canali sudoriferi (porokeratosi). G It Mal Vener Pele. 1893;28:313-55.
3. Bozdağ KE, Bicakci H, Ermete M. Giant porokeratosis. Int J Dermatol. 2004;43:518-20.
4. Ghorpade A. Localized actinic nasal porokeratosis. Clin Exp Dermatol. 2010;35:211-2.
5. Odeyinde S, Belcher H. Isolated single digit Porokeratosis of Mibelli: an unusual case. Dermatol Online J. 2012;18:13.
6. Kudligi C, Bhagwat PV, Giriyan SS, Eshwarrao MS. Unilateral systematized linear porokeratosis: a report of a rare case. Indian J Dermatol. 2011;56:452-3.
7. Lee HR, Han TY, Son SJ, Lee JH. Squamous cell carcinoma developing within lesions of disseminated superficial actinic porokeratosis. Indian J Dermatol. 2011;56:452-3.
8. Irisawa R, Yamazaki M, Yamamoto T, Tsuboi R. A case of porokeratosis plantaris palmaris et disseminata and literature review. Dermatol Online J. 2012;18:5.
9. Ghorpade A, Ramanan C. Punctuate porokeratosis. Indian J Dermatol Venereol Leprol. 1995;61:123-4.
10. Wang NS, Gruson LM, Kamino H. Fasial follicular porokeratosis: a case report. Am J Dermatopathol. 2010;32:720-2.
11. Gutierrez EL, Galarza C, Ramos W, Tello M, De Paz PC, Bobbio L, et al. Facial porokeratosis: a series of six patients. Australas J Dermatol. 2010;51:191-4.
12. Kanitakis J, Euvrard S, Faure M, Claudy A. Porokeratosis and immunosuppression. Eur J Dermatol. 1998;8:459-65.
13. Ikizoglu G. Genodermatozlar. In: Tüzün Y, Gürer MA, Serdaroglu S, Oguz O, Aksungur VL (eds). Dermatoloji, 3rd ed. Istanbul: Nobel Tıp Kitabevi; 2008;1609-736.
14. Nabai H, Mehregan AH. Porokeratosis of Mibelli. A report of two unusual cases. Dermatologica. 1979;159:325-31.
15. Tran H, Bossenbroek NM, Rosenman K, Meehan SA, Sanchez M, Prystowsky S. Steroid-responsive facial eruption with cornoid lamella-a possible new entity. Dermatol Online J. 2008;14:9.
16. Lee Y, Choi EH. Exclusive facial porokeratosis: histopathologically showing follicular cornoid lamella. J Dermatol. 2011;38:1072-5.