Porokeratozis Mibelli

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Özet

Porokeratozis, nadir görülen, lokalize bir keratinizasyon bozukluðudur. Porokeratozisin patogenezisi tam olarak açýk olmamakla birlikte anormal epidermal hücre klonlarýndan geliþtiði bilinmektedir. Knuckle pad ise parmak eklemlerinin dorsal yüzlerine yerleþen fibromatöz kalýnlaþmadýr. Burada knuckle padleri olan porokeratozis Mibellili bir hasta sunulmuþtur.

Anahtar Kelimeler *: Porokeratozis, Knuckle pad.*

Porokeratosis Of Mibelli With Knuckle Pads

Abstract

Porokeratosis is an uncommon localized disorder of keratinization. Porokeratosis appears to develop from abnormal clones of epidermal cells, but pathogenesis of porokeratosis is unclear. Knuckle pads are well-defined thickening of the skin over the articular areas of the dorsal surface of the fingers or toes. We describe here porokeratosis of Mibelli in a patient with knuckle pads..

Key words*: Porokeratosis, Knuckle pads.*

Porokeratozis Mibelli is an uncommon hereditary disorder. PM is inheridet as an autosomal dominant trait but non familial forms probably exist. PM affects males two or three times than females (1).

The onset of porokeratosis is visible as a brownish keratotic papule, about 1 mm in diameter. The top is occupied by a horny plug, the removal of which leaves a depression or crater. Peripheral extension of the border, over a long period of time, is accompanied by enlargement of the crater, producing an annuler or oval figure. The border is wall - like, keratotic, delicate or coarse and continuous or intermittent. Centrally the color of the lesion is gray, tan or yellowish, whereas the border is usually a darker gray, dirty brown or even black. The center is dry, smooth and slightly sclerotic, the follicular and poral orifices usually are obliterated and the normal skin markings are decreased (1.2).

The eruption may consist of one, a few or many small lesions or exclusively of large lesions.

There are no subjective symptoms. Sites of predilection are the extremites, especially their distal dorsal portions, the face neck and the genitalia, although any area of the body may be involved. Porokeratosis may appear on the buccal and mucous membrane as opalescent, macerated round patches with slightly raised whitish borders and a hyperemia periphery (1.3).

A variety of clinical presentations has been recognised : porokeratosis of Mibelli, disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminate (PPPD), linear porokeratosis, punctate porokeratosis, disseminated superficial porokeratosis (DSP). In addition the following types of porokeratosis is reported in the literature, giant porokeratosis (4), zosteriform porokeratosis of Mibelli (5), hyperkeratotic form porokeratosis of Mibelli (6). Sometimes different porokeratotic variants may coexits in one patient.

Case Report

The patient was a 16 year-old, house-wife. She was admitted to the Dermatology Department of Süleyman Demirel University for her facial lesions. Her complaints started at early childhood at the age of 3 years. Her suffering was increasing during summer months. The patient was more warried about her appearence than the discomfort of the

lesions. Dermatological examination revealed numerous lesions with the largest one at her nose. There were also lesions located at the cheek, at the edge of her right eye, at the upper eyelid and upper lip. Lesions had different shapes such as round, oval or semilunar. The color of the center of the lesions were tan or reddish-brown. The central part of the lesion were dry and had a shiny appearence with atrophia. The border was wall-like and keratotic (Figure 1).

Figure 1

The nail of right index finger was deformed due to a trauma occurned in childhood. The skin overlying proximal interfalangial joints was thickened in a roundend shape on both hands (Figure 2).

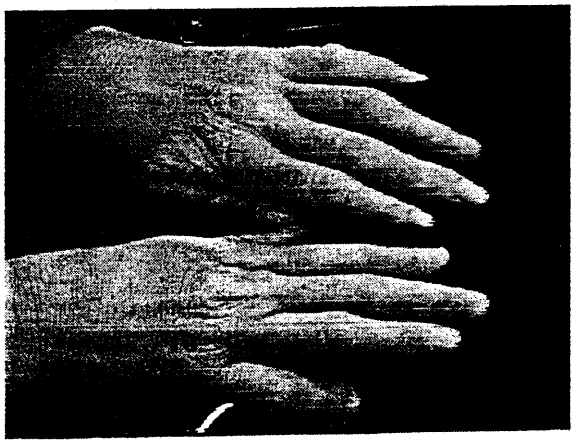

Figure 2

Routine laboratory examination and general medical check-up was normal. A biopsy was taken from left cheek. The examination of the biopsy specimen showed pronounced acanthosis and hyperkeratosis. Also a deep invagination was seen together with a parakerotic column in the acanthotic region. An inflammatory cell infiltration was also seen under this invagination in the dermis (Figure 3).

Figure 3

These findings enabled us to make a diagnosis of porokeratosis of Mibelli (PM). Based on the clinical findings, the lesions at her hands were diagnosed as knuckle pads. Topical 5% 5 fluorouracyl-topical corticosteroid combination was used for treatment of the lesions at her face. She was also adviced to avoid sunlight and use effective sunscreens. The lesions of the patient responded well to the treatment within one month. The small lesions disappeared without any scar formation whereas big lesions left atrophic scars behind. During the treatment period no new lesions were observed. There were no side effects of the treatment. The patient is called at regular intervals. Knuckle pads were not treated since they were asymptomatic.

Discussion

PM is an uncommon hereditary disorder, characterized by a keratotic lesion whose peripheral spread procudes a wall-like border and an atrophic center. PM is inherited as an autosomal dominant trait but nonfamilial forms probably exist. PM affects males two or three times than females (1).

The histogenesis of PM is not completely clear. Reed and Leone proposed that PM could be related to mutant cellular "clones" of epithelial cells in the epidermis, and the cornoid lamella was a marker for the boundary between the abnormal clonal population and the normal epithelium. Changes in the dermis beneath porokeratotic lesions were considered secondary to those in the epidermis and not part of the causal mechanism (2, 7). In contradiction to Reed and Leone's hypothesis there are many reports suggesting that the dermal injury could be an initiating factor in the pathogenesis of PM (8). Although the initiating event is often not apparent, clinical disease may be triggered by a number of stimuli including irridiation with

ultraviolet light or x-ray, viral activation, repeated trauma, immunosuppression, therapy for chronic active hepatitis (9.10.11).

Knuckle pads are circumscribed fibromatous thickening overlying the finger joints, often occuring as a sporadic and apparently isolated defect, but sometimes familial. Knuckle pads may be idiopatic or an occupational disease. They are found as a result of occupational trauma in bakers, carpet installers and sheep shearers (1). The knuckle pads in our patient were not of traumatic origin. Her family members did not have similar lesions on their faces and hands. So this case was regarded to be sporadic.

The role of immunosuppression is well known in the etiopathogenesis of PM. Our patient did not use any immunosuppressive drugs nor had a previous immunosupressive disease.

It is known that ultraviolet light is an important factor especially in the occurrance of disseminated superficial actinic porokeratosis, but our patient's lesions were classified as PM type both by clinically and dermatopathologically. On the other hand because the lesions were at the face and increased in summer month, ultraviolet light seems to play an important role in the formation of PM as well.

Porokeratosis is believed to be a premalignant condition of the skin. Recent flow cytometric studies showing DNA aneuploidy in the epidermis of some type of porokeratosis support this conclusion. The premalignant potential of porokeratosis is well illustrated by the numerous reports of squamous cell carcinoma and Bowen's disease developing in preexisting lesions of porokeratosis (4.7). A possible association between porokeratosis especially punctate porokeratosis and internal malignancy has been reported (10). This association led us to speculate that porokeratosis could be a sign of internal neoplasia. For this reason, the cases of porokeratosis should be controlled at regular intervals for the risk of malignant development on lesions and for the signs of internal malignancy. The patient was also warned about this possibility and she was followed up regularly.

Kaynaklar

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