Asymptomatic solitary plaque in the groin

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A 28-year-old male patient complained of asymptomatic skin lesion in the genital area for 3 years. The lesion was gradual in onset and had slowly progressive course. Topical steroids were used as an initial treatment for about 7 months but without any significant improvement. 3 sessions of cryotherapy failed to show any improvement, and the patient stopped treatment for about one year. The patient was treated again with potent topical steroid for 3 months but no response was achieved. There was no previous history of similar condition and family history for similar disease was negative.

Skin examination showed solitary, well defined plaque on the left groin, measured about 1.6 x 1.2 cm (Fig. 1). The lesion was firm in consistency and the surface was slightly pigmented (Fig. 2). The border of the lesion was slightly scaly and stria rubra were obvious on the inner aspect of left thigh due to prolonged use of topical steroids.

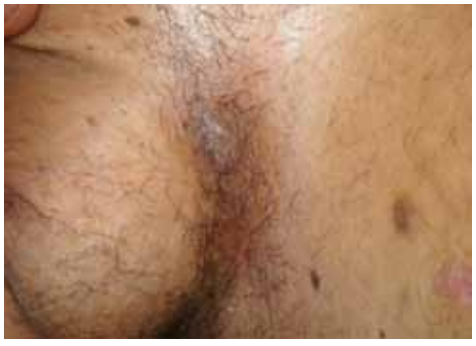

Fig. 1 A solitary, well defined plaque in the left groin, measuring about 1.6x1.2cm.

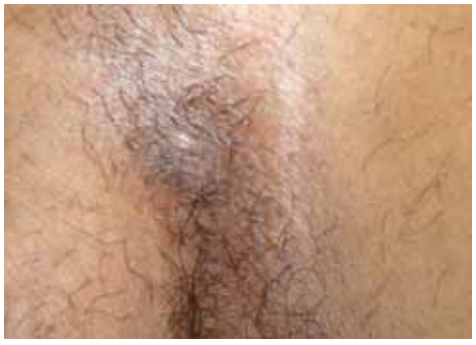

Fig. 2 The border of the lesion was slightly scaly while the surface was slightly pigmented.

Examination of inguinal lymph nodes showed no significant findings. General examination was irrelevant, while routine investigations showed no significant abnormalities.

What is the clinical diagnosis?

1. Bilharzial granuloma
2. Granular cell tumor
3. Bowen's disease
4. Squamous cell carcinoma

A skin biopsy from the lesion showed dense band-like inflammatory infiltrate composed mainly of lymphocytes and histiocytes with scattered melanophages. The epidermis showed hyperkeratosis and acanthosis with elongation of rete ridges (Fig. 3). In focal area, there was an abrupt change in the stratum corneum with appearance of a column of parakeratosis. The granular layer was almost absent under the parakeratotic column while

the Malpighian layer showed scattered vacuolated and necrotic keratinocytes (Fig. 4).

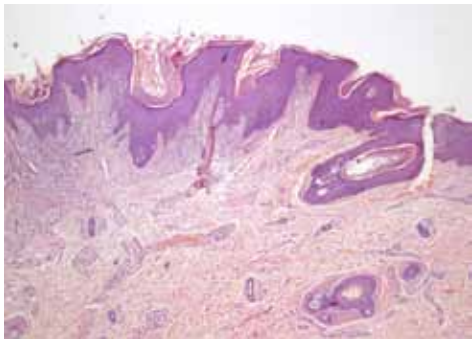

Fig. 3 The epidermis showed hyperkeratosis and acanthosis with dense band-like inflammatory infiltrate.

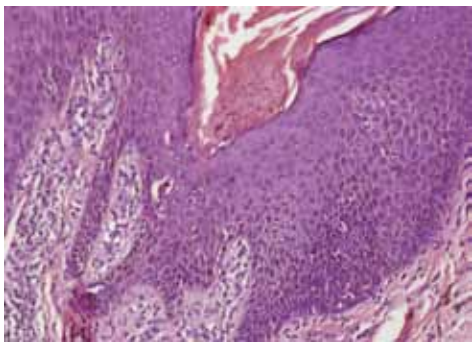

Fig. 4 A column of parakeratosis with absent granular layer and scattered vacuolated and necrotic keratinocytes, characteristic of cornoid lamella.

The diagnosis is

Porokeratosis of Mibelli.

DISCUSSION

Porokeratosis is an inherited dermatosis with an autosomal dominant pattern. There are five different common forms that can be distinguished clinically, including the plaque type of Mibelli, disseminated superficial actinic porokeratosis (some lesions of this type are distributed mainly in areas not exposed to the sun or appear in patients on immunosuppressants), linear porokeratosis, porokeratosis plantaris palmaris et disseminata and punctuate porokeratosis.[1,2]

Classic porokeratosis or porokeratosis of Mibelli (PM) is a chronic, progressive dermatosis that may rarely evolve with spontaneous remission. Regarding the mode of inheritance, it may arise in an autosomal dominant form, more commonly in a random form.[3] Multiple etiologies are proposed in the clonal proliferation of keratinocytes, like chronic sun exposure, hepatitis B and C virus infection, HIV and immunosuppresion.[4]

PM consists of one or more plaques, usually a small number of them, which may occur anywhere in the body, more frequently in extremities, especially hands and feet, with unilateral distribution. Other areas, such as neck, shoulders and genitals may also be affected. The occurrence of facial and mucosal lesions is rare.[5] Genital porokeratosis is extremely rare, but classical lesions have been reported on the penis and scrotum. It seems that genital porokeratosis is more common in Asian populations.[6]

The classic lesion of porokeratosis is usually seen during childhood as one or multiple annular plaques with central atrophy and elevated keratotic borders usually greater than 1 mm in height that have a longitudinal furrow typically seen in the center of the ridge. This ridge expands over a period of time. It affects men twice as often as women. The lesion may be hypopigmented or hyperpigmented, scaly, atrophic, hairless and anhidrotic.[7]

Giant porokeratosis is considered to be a morphological variant of porokeratosis of Mibelli with a diameter of up to 20 cm and surrounding wall of 1cm. As there is risk of development of squamous cell carcinoma in giant porokeratosis (10%), early diagnosis and treatment is necessary.[8]

Porokeratosis involving the genital region occurs in generalized forms or as a localized lesion confined to the genital area, latter being the extremely rare presentation is generally classified as

a plaque-type porokeratosis of Mibelli, with less than 30 cases reported in the literature.[9] Based on published reports, the skin lesions are typically characterized by one or more well-defined brownish erythematous keratotic papules and annular plaques. The center of the lesion can be hyperpigmented or hypopigmented and atrophic. They can be asymptomatic or associated with severe itching.[10]

A variant called porokeratosis ptychotropica, was initially described as a pruritic eruption of the gluteal cleft, typically involves the buttocks and gluteal cleft mimicking an inflammatory disorder and is an important differential diagnosis in genitogluteal porokeratosis. Although affecting similar locations, with clinical features that can be histologically superimposable, the ptychotropic form presents a distinctive feature of the presence of multiple cornoid lamellae.[11]

Porokeratoma is a recently described type of acanthoma containing the characteristic feature of porokeratosis, which is known as the cornoid lamella. It usually appears as a tumor-like lesion in patients without personal or family history of porokeratosis, and without immunosuppressive conditions. The most frequent locations are extremities, followed by head and neck, chest, buttocks, and intergluteal cleft.[12] Clinically, porokeratoma may be present as scaling plaques, papules, or nodules with central hyperkeratosis, but lesions can occasionally have verrucous appearance. Although porokeratomas share a similar histological feature of cornoid lamellae, they are clinically and morphologically distinct from porokeratosis and its variants.[13]

Treatment modalities for genital porokeratosis are same as that of porokeratosis occurring on other sites. These include cryotherapy (liquid N2), surgical excision, CO_2 laser ablation, topical 5% 5-fluorouracil, Vitamin D3 analogues, retinoids and imiquimod cream 3%, and diclofenac gel have been found to be useful for the symptomatic relief and stabilizing evolution of lesions of genital porokeratosis.[14] Malignant degeneration has been reported in all forms of porokeratosis. Large lesions, long-standing lesions, and linear lesions were found to be at greatest risk. There is no such change reported till date, in lesions of genital porokeratosis.[15]

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