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Malignant Transformation in a Genitocrural Porokeratosis : A Case Report

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Abstract

Porokeratosis is a rare heterogenous group of keratinization disorder with an unclear pathogenesis, and has varied clinical presentations.

It may present with annular papules or plaques with central atrophy and clinically and histologically distinct ridge-like border called 'cornoid lamella'. Common variants include porokeratosis of Mibelli (PM), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, punctate porokeratosis and porokeratosis palmaris et plantaris disseminata (PPPD). Here we report the case of a 79 year old female, who developed malignancy of porokeratosis over the groin region which had been mismanaged as eczema for many months and has been eventually diagnosed as Squamous cell carcinoma.

Keywords: Malignancy; Porokeratosis; Squamous cell carcinoma

Introduction

Porokeratosis is a rare, acquired heterogeneous group of keratinization disorders.[1] It is an enigmatic skin condition in terms of etiopathogenesis, clinical presentation, histopathology, as well as treatment options.[2] Porokeratosis has been majorly classified as: porokeratosis of mibelli (PM), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, punctate porokeratosis, porokeratosis palmaris et plantaris disseminata (PPPD) ,porokeratosis ptychotropica, and eruptive disseminated porokeratosis.[3]

Besides these, a few rare variants like giant, hyperkeratotic, and verrucous lesions have also been described, with malignant degeneration associated with almost all forms of porokeratosis.[4] Histologically, cornoid lamella is the hallmark of all porokeratosis variants.[1]

Case Report

A 79 year old female patient presented with a single dark coloured lesion over the right groin fold for the last 2 years.

The lesion had been asymptomatic initially, but since the last few months, she had developed complaints of pain and bleeding from the lesion on manipulation. The patient has been applying a topical corticosteroid and salicylic acid combination on and off for the last 1 year on the advice of a local doctor who had diagnosed it as eczema.

There was no significant past history other than pulmonary tuberculosis around 50 years ago for which she had taken treatment for one year.

Cutaneous examination revealed a single, well defined, discrete dark brown coloured annular plaque of size 3*5 cm with raised, slightly scaly borders present over the right groin and 2 nodules of approximately 1*1*1 cm present over the superior border and one of these nodules which were present over the medial aspect,has a hyperkeratotic, verrucous surface, while the other

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one, present laterally, had a smooth overlying surface. The surrounding skin showed atrophy with striae and telangiectasia. (Figure 1) There was no palpable inguinal or femoral lymphadenopathy.

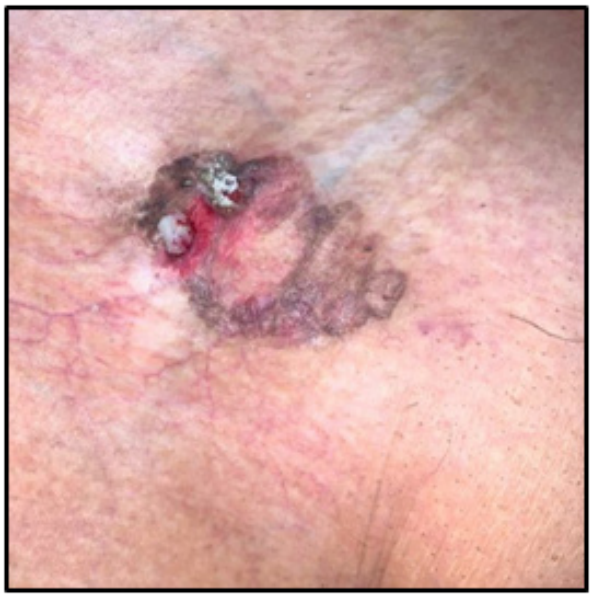

Figure 1: single annular plaque of size 3*5 cm with a slightly raised scaly erythematous border over the right groin, also over one margin of the plaque, an erosion of approximately 1*0.5 cm in size was noted, and 2 nodules of approximately 1*1*1 cm were seen at the superior border of the lesion.

Dermoscopic (10x) evaluation of the lesion showed a single, hyperpigmented plaque with yellowish coloured adherent crusting present over the lateral aspect of the lesion, and with fine white scaling over the medial aspect of the lesion, with telangiectasia in the surrounding skin. (Figure 2)

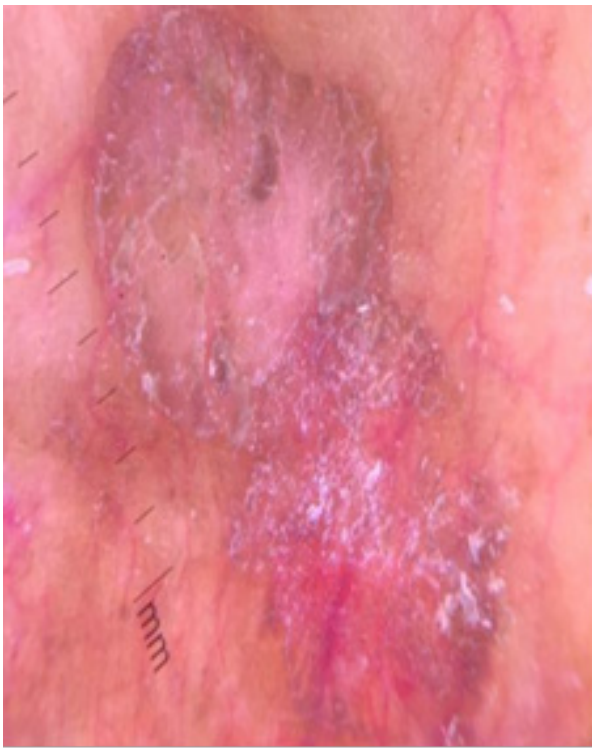

Figure 2: single, well defined hyperpigmented plaque with yellowish coloured adherent crusting present over the lateral aspect of the lesion, having fine white scaling over the medial aspect with telangiectasia in the surrounding skin.

Dermoscopy (20x) showed dark brown pigmentation with the keratin rim having shiny white structures over the superior border (Figure 3)

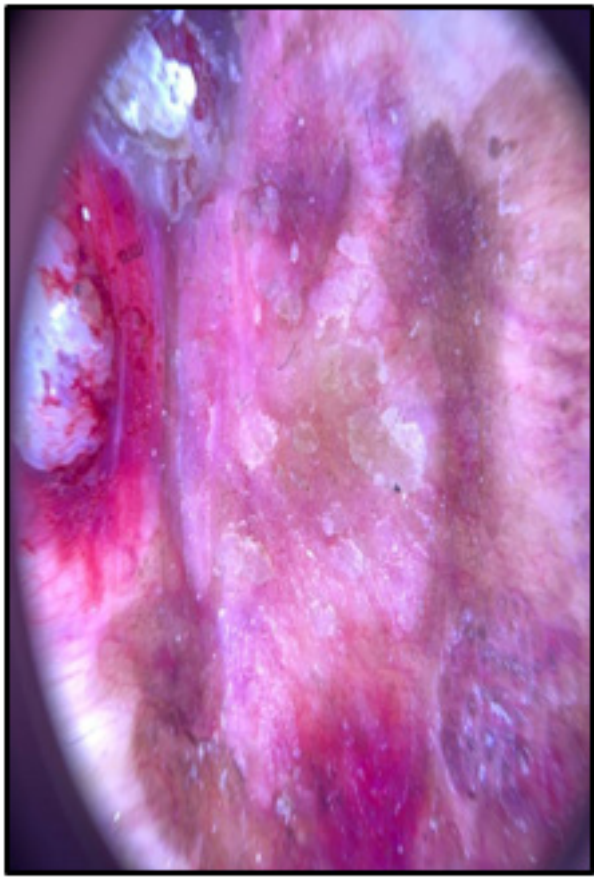

Figure 3: -Dark brown to black coloured nodules with whitish scaling present over the superior border of the hyperpigmented plaques with multiple telangiectasia seen over the skin surrounding it.

All haematological investigations were normal. The Mantoux test was positive, with radiological consolidation in the upper zone of the right lung. Sonography of the localized part was done, which was suggestive of a hypoechoic lesion with internal hyperechoic (air foci) and surrounding echogenic subcutaneous tissue seen in the right inguinal region. It also showed a few subcentimeter sized lymph nodes in the right inguinal region.

Based on clinical evaluation and non-specific dermoscopy findings, a wide array of differential diagnoses were considered, which included porokeratosis with malignant transformation, basal cell carcinoma, extramammaryPaget's disease, Bowen's disease, and lupus vulgaris.

For further evaluation, a 4 millimetre punch biopsy was taken from the nodule that was present over the superior border, which showed epidermis with parakeratosis and characteristic cornoid lamella with

focal atypically large hyperchromatic multinucleated cells, and few mitotic cells at the stroma, along with keratin pearls as demonstrated in the histopathology (Figure 4), which was suggestive of squamous cell carcinoma in situ with stromal invasion.

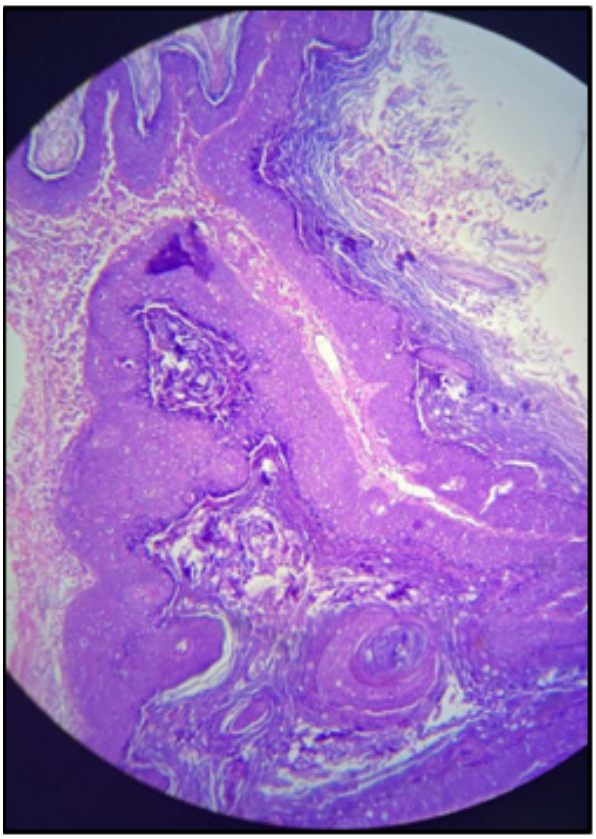

Figure 4: 10x magnification of histopathology showed epidermis with parakeratosis and characteristic cornoid lamella with focal atypically large hyperchromatic multinucleated cells, and few mitotic cells at the stroma, along with keratin pearls, which was suggestive of squamous cell carcinoma in situ with stromal invasion.

Considering the option of complete excision, the patient was referred to an onco-surgeon for expert management, following which the patient was lost to follow-up.

Discussion

Porokeratosis is a dermatosis of clonal expansion and abnormal development and differentiation of keratinocytes and is diversified clinically on the basis of localized and disseminated forms. Localized forms include porokeratosis of Mibelli, linear, punctate, giant, palmoplantar, and genitocrural porokeratosis, while generalized forms include disseminated superficial actinic, systemized linear, disseminated superficial, disseminated palmoplantar porokeratosis.[5] Recently few new variants like eruptive bullous, pruriginous, lichen planus like, follicular variants, and porokeratoma have also been described.[2]

Dermoscopy of porokeratosis usually shows red to brown coloured dots and globules, which is suggestive of melanophages and dilated capillaries in the dermis.[6] There are central white areas of scarring, with the characteristic finding of a white coloured double marginated peripheral border; on UV light dermoscopy, these findings are known as “white track”, “lines of volcanic crater” and “diamond necklace” appearances.[7]

In histopathology, most of the forms show hallmark cornoid lamella (suggestive of an active border), which is composed of a thin column of parakeratosis with keratinized invagination of epidermis through the stratum corneum, and an absent or reduced stratum granulosum (result of faulty maturation or acceleration of epidermopoiesis).[5] In dermis perivascular and lymphocytic infiltrates, having areas of epidermal atrophy centrally.[5]

It is estimated that 7.5 -11% of patients with porokeratosis have malignant transformations, of which squamous cell carcinoma is the most common. Malignant transformation can be seen in all forms of porokeratosis, with the highest incidence in linear porokeratosis which is 19%, followed by 7.6% in the porokeratosis of Mibelli and 3.4 % in disseminated superficial actinic porokeratosis.[1]

Management of porokeratosis is an equally challenging situation, and patients with genitocrural, disseminated superficial actinic, and disseminated superficial porokeratosis are often the most difficult to manage.[2] The treatment for this condition is difficult; however, various modalities have been tried for the same, which include electrochemotherapy, radiation therapy and complete excision.[8]

In our case, the elderly female showed malignant changes over the preexisting genitocrural porokeratotic lesions, which had been modified by the use of various combinations of multiple topical applications. As the patient belonged to a remote area with no easy availability of an expert dermatologist, her condition remained undiagnosed for a few months.

CONCLUSION

The incidence of malignant transformation in a premalignant condition like porokeratosis can be curbed by a proper timely clinical diagnosis with further confirmation provided by laboratory investigations like dermoscopic and histopathological evaluation.

Even with a colossal increase in the number of new dermatologists getting qualified every year, there are still areas in our country with limited access to an expert dermatologist, as a result, these people fall prey to unqualified doctors and are often misdiagnosed and improperly treated.

The telemedicine approach is also not feasible in remote areas due to a lack of tech-awareness and telecom facilities. Community dermatology with organisation of medical camps is a way forward to help people take better care of their skin related ailments.

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