

Porokeratosis ptychotropica without genitogluteal involvement – A rare presentation

Dear Editor,

Porokeratosis ptychotropica (PP) is a rare variant of porokeratosis. Very few cases of PP are reported worldwide. We present a case of PP with no genitogluteal involvement which has been reported only once before. An 18-year-old boy visited the dermatology outpatient department with the complaint of a single lesion over his left ankle for one year. The lesion was slightly itchy and had gradually increased in size. On examination, a single well-defined hyperkeratotic verrucous plaque of size 3×3 cm² was present on the left medial malleolus [Figure 1]. Clinical differential diagnoses were lichen simplex chronicus, deep mycosis, cutaneous tuberculosis, and squamous cell carcinoma. An incisional biopsy from the lesion was done. Histopathology revealed an acanthotic epidermis with elongation of rete ridges and focal parakeratosis, multiple lamellae composed of parakeratotic cells with loss of granular layer beneath the parakeratotic zone, and moderate perivascular lymphocytic infiltrates in the superficial dermis. The histopathological findings confirmed the diagnosis of PP [Figure 2]. The patient was prescribed topical tazarotene 0.1% cream once daily for two months. On follow-up, the lesion had improved with no recurrences thereafter [Figure 3].

Porokeratosis is a keratinisation disorder due to abnormal clonal expansion and differentiation of keratinocytes. The prevalence of porokeratosis was found to be 24.2 per 100,000 population.¹ Germline mutation of mevalonate pathway genes has been postulated to be the underlying cause of porokeratosis.¹ PP is a rare hyperkeratotic variant of porokeratosis. The term porokeratosis ptychotropica is derived from the words ‘ptyché’ meaning folds and ‘tropé’ meaning a turning which was coined by Lucker *et al.* in 1995.² PP most commonly affects the gluteal cleft with or without involving the extremities. There is only one case reported of PP not involving genito-gluteal region, similar to our case.³ It is clinically characterised by pruritic thick hyperpigmented verrucous plaques in a butterfly configuration



Figure 1: Initial presentation with a verrucous plaque on the medial malleolus.

that can simulate viral warts, hypertrophic lichen planus, lichen simplex chronicus, psoriasis, perforating disorders, squamous cell carcinoma, deep mycosis, and tuberculosis verrucosa cutis. The classical histopathologic feature of PP is the presence of multiple layers of cornoid lamellae throughout the lesion unlike other variants of porokeratosis with cornoid lamellae only at the periphery.⁴ There are also some reports of the presence of dermal amyloid deposits

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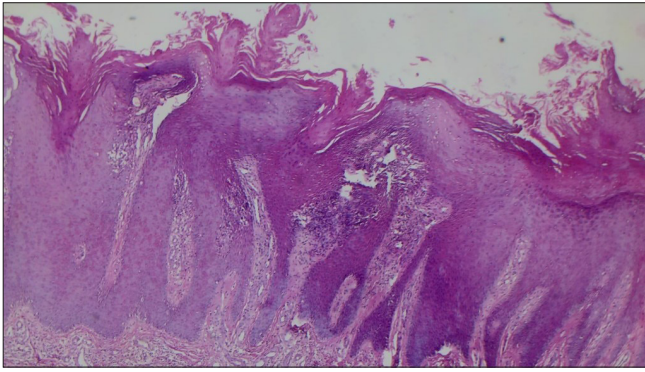


Figure 2: Histopathology of the plaque showing acanthosis, elongated rete ridges, and multiple cornoid lamellae [Haematoxylin and Eosin, 40x].



Figure 3: Near complete clearance of the lesion after two months of treatment with topical tazarotene.

noted on histopathology.⁵ PP is usually resistant to treatment. Various treatment modalities include topical retinoids, imiquimod, 5-fluorouracil, isotretinoin, photodynamic therapy, cryotherapy, carbon dioxide laser, and excision.⁶ Around 7.5% of porokeratotic lesions undergo malignant transformation. There is a case report of PP developing into squamous cell carcinoma after 18 years.⁷ Therefore, long-term follow-up of PP is essential.

In conclusion, though most of the cases of PP are present in the genitogluteal region, these regions can rarely be spared. So, PP should always be considered in the differential diagnosis of a hyperkeratotic verrucous lesion to avoid misdiagnosis and a delay in diagnosis, thus preventing long-term complications and malignant transformation.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

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