# Coexistence of multiple variants of porokeratosis

Sir,

Porokeratosis is a group of hereditary or acquired disorders of epidermal keratinization characterized by keratotic lesions with an atrophic center and a prominent peripheral ridge, the histological hallmark of which is the cornoid lamella.[1] Clinically, seven variants have been identified: plaque type, disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis, linear porokeratosis, giant porokeratosis, porokeratosis palmaris et plantaris disseminata and punctate porokeratosis. Although different types of porokeratosis have been previously reported to coexist in a single individual, such combinations are rare.[1-4] We report coexistence of linear porokeratosis, disseminated superficial porokeratosis, and giant porokeratosis in a Chinese man.

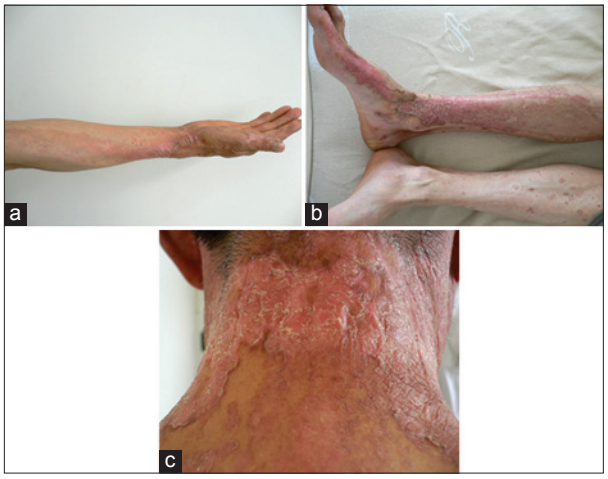


**Figure 1: Clinical variants: (a) Linear porokeratosis on the right upper limb; (b) linear porokeratosis on the right lower limb and disseminated superficial porokeratosis on the left lower limb; (c) giant porokeratosis on the nape of neck**

A 45-year-old man presented with multiple, asymptomatic, brownish and keratotic plaques in a linear array on his right upper and lower limbs since the age of 25 years. Similar lesions had recently developed over his left upper limb, face, nape of neck, trunk and thighs. Cutaneous examination revealed unilateral, scaly and well-defined erythematous plaques with central atrophy and a distinct keratotic ridge arranged in linear patterns over the extensor surface of both his upper limbs and right lower limb. Two large lesions of size 14 × 26 cm and 16 × 18 cm with central atrophy and a peripheral hyperkeratotic ridge were observed on his neck and back respectively. Multiple annular, coin-sized, scaly plaques with central atrophy and thin grooves were found on both covered and exposed sites [Figure 1a–c]. Complete blood count, urinalysis, fasting blood sugar, liver and renal function, CD4 count, CD8 count, CD4/CD8 ratio, complement and immunoglobulin levels were within normal limits and serologic tests for syphilis and HIV were negative. A chest radiograph, ultrasonography of the abdomen and computed tomography scan of the right lower extremity were all normal. His family history was unremarkable.

Biopsies were performed on the borders of three lesions with different morphology on the right upper limb, neck and left thigh. All three biopsies showed a cornoid lamella, absent granular layer below the invagination and mild inflammatory infiltrate in the upper dermis [Figure 2]. Based on the clinical manifestations and histological features, diagnosis of coexistent linear porokeratosis, disseminated superficial porokeratosis and giant porokeratosis was made.

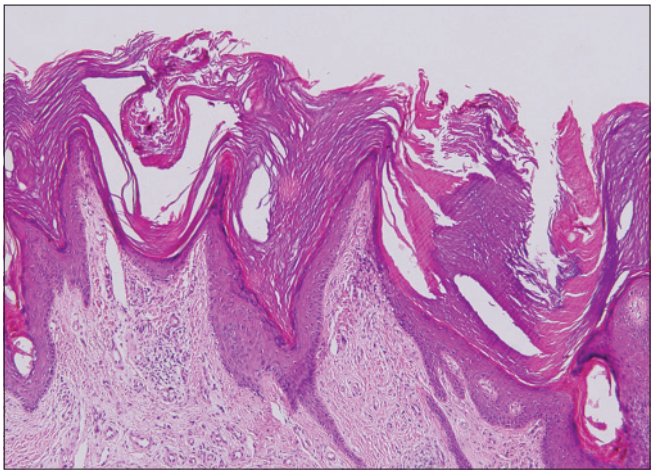

**Figure 2: Photomicrograph showing cornoid lamella with inflammatory infiltrate in the dermis (H and E, ×100)**

Till date seven variants of porokeratosis have been described.[1] Since Welton[5] first described an association of linear porokeratosis and disseminated superficial actinic porokeratosis in a woman, along with disseminated superficial actinic porokeratosis in her three sisters in 1972, some cases of coexisting linear porokeratosis and disseminated superficial actinic porokeratosis in a single individual have been reported.[1,4] Porokeratosis of Mibelli has also been described to coexist with disseminated superficial actinic porokeratosis.[5]

Our patient had linear, giant porokeratotic, and widely disseminated flat and small annular lesions on different anatomic sites. As the disseminated lesions were predominantly localized to unexposed areas rather than on the face and there was no history of prolonged outdoor work, the term disseminated superficial porokeratosis was preferred instead of disseminated superficial actinic porokeratosis. We were unable to find any previous reports of the co-existence of linear porokeratosis, disseminated superficial porokeratosis and giant porokeratosis.

The exact pathogenesis of the co-existence of different variants of porokeratosis in a single individual remains largely unknown. It is suggested that the similarities of clinical appearance and histological findings among different variants of porokeratosis in one patient may indicate different phenotypic expressions of a common genetic abnormality.[1,4] Alternatively, genes of different variants of porokeratosis may be present on the same or closely linked loci.[3] The coexistence of variants of porokeratosis could be explained based on simultaneous expression of closely linked genes.

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