

Observation Letters

Acral porokeratosis associated with anonychia

Sir,

Among dermatological disorders, porokeratosis remains an enigma owing to its uncertain pathogenesis, varied clinical presentations, and unpredictable outcome. Porokeratosis is characterized by marginate scaling lesions, histologically showing a column of parakeratotic keratinocytes (the cornoid lamella). Various morphological patterns include classical porokeratosis of Mibelli, disseminated superficial actinic porokeratosis, disseminated superficial porokeratosis of immunosuppression, childhood forms, linear porokeratosis, porokeratosis palmaris et plantaris disseminata, and punctate porokeratosis.¹

While a few cases involving distal extremities with nail changes have been described in the literature, complete loss of nail (anonychia), with exclusive involvement of the acral area, is very rare.

A 35-year-old female patient, an agricultural laborer, presented with a history of raised asymptomatic lesions, over the right hand and bilateral feet, of one-year duration. She had noticed a ring-like lesion over her feet, involving the toes, with associated nail discoloration and subsequent dystrophy. Subsequently, she noticed similar lesions involving the right hand. The skin lesions preceded the involvement of nails. There was no family history of similar illness.

On cutaneous examination, a well-defined, hyperpigmented plaque with a prominent peripheral keratotic ridge and central atrophy and depigmentation was seen over the right hand involving the thumb, and the medial border of the second finger and first interdigital space [Figure 1]. Anonychia of the right thumb nail was seen. Bilaterally, the dorsa of the feet showed similar plaques of size 5 × 5 cm over the first and second toes. There was anonychia over both the great toe nails [Figure 2]. The right second toe nail showed dystrophy. There was no involvement of the oral or genital mucosa. The patient had no features of immunosuppression.



Figure 1: A well-defined, hyperpigmented plaque with a prominent keratotic ridge at the periphery and central atrophy and depigmentation seen over the right hand

Skin biopsy, taken from the edge of the lesion, showed acanthosis, hyperkeratosis, papillomatosis, and a keratin-filled invagination with a central parakeratotic column, suggestive of cornoid lamellae. The epidermis underlying the cornoid lamella showed absence of granular layer [Figure 3]. The dermis showed minimal perivascular lymphocytic infiltration. All the other laboratory investigations were found to be within normal limits. Patient chose treatment with electrocautery; however, the response was poor.

Porokeratosis presents itself in a fascinating array of morphological variants. The morphology of porokeratosis depends upon the site and the triggers. Based on the site, facial porokeratosis, genital porokeratosis, and porokeratosis ptychotropica have been described in the literature.^{1,2} However, porokeratosis of the acral areas, particularly the thumbs, with nail involvement is very rare. Two case reports describing nail involvement are compared with this case in Table 1. This case presented with anonychia of the affected digits and it could be due to the disease process involving the nail matrix and nail bed.

Apart from the genetic causes, certain environmental triggers such as irradiation/ultraviolet light, mechanical trauma, infective agents have been proposed as etiological factors. Immunosuppression has also been implicated. Certain drugs, such as thiazide diuretics, and immunosuppressive drugs such as prednisolone, azathioprine, used in organ transplantation, have also been reported as etiological factors in elderly patients. This case is being presented for the unusual involvement of acral areas, along with anonychia, giving rise to the intriguing possibility of a new clinical variant of porokeratosis i.e., acral porokeratosis with anonychia.

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Conflicts of interest

There are no conflicts of interest.



Figure 2: Bilateral dorsa of feet shows well defined plaques of size 5 cm × 5 cm with peripheral raised margins over the first and second toes. There was anonychia over both the great toe nails

Table 1: Comparison of case reports with our case

Features	Report by Robati <i>et al.</i> ³	Report by Karthikeyan <i>et al.</i> ²	Present report
Age of the patient (years)	30	26	35
Sex	Male	Female	Female
Morphological type of porokeratosis	Porokeratosis of Mibelli	Porokeratosis of Mibelli	Porokeratosis of Mibelli
Sites involved	Radial aspect of left hand; dorsal surface of right hand involving second and third fingers; face	Bilateral thumbs and great toes	Right hand involving thumb Bilateral dorsa of feet involving great toes
Mucosal involvement	Oral mucosa	No	No
Nail involvement	Dystrophy of left thumb nail	Dystrophy of nails	Anonychia of both great toe nails; Near total loss of nail over right thumb
Pathology	Typical features of porokeratosis were seen	Pathological hall mark cornoid lamella seen	Cornoid lamella seen
X-ray features	Not done	No bony abnormality	Not done
Treatment given	2 months of calcipotriol and tretinoin 0.05; cryotherapy	Topical retinoids	Electrocautery for keratotic border

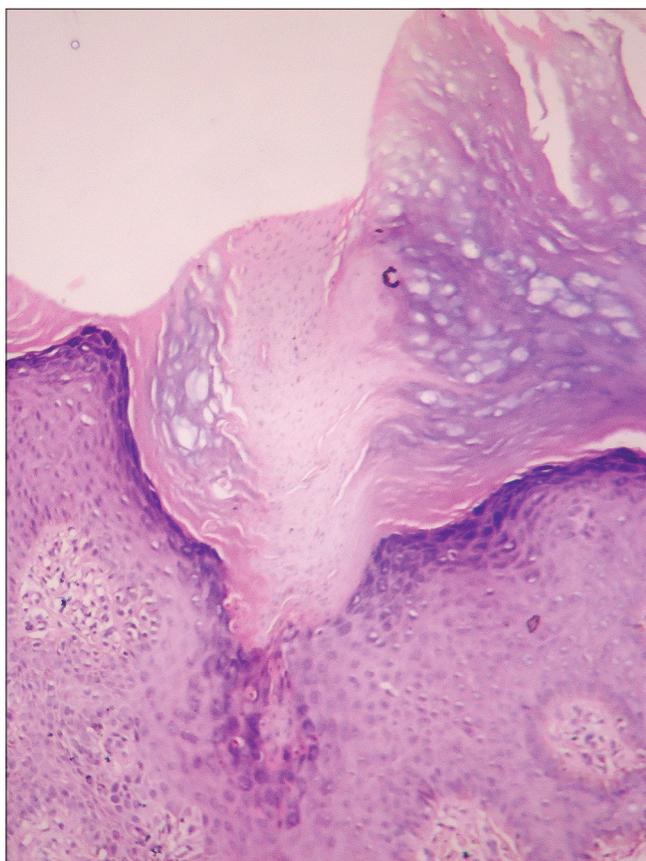


Figure 3: Hyperkeratosis, papillomatosis and a keratin filled invagination. From the center of the keratin filled invagination raises a parakeratotic column suggestive of cornoid lamellae (H and E, ×100)

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