Verrucous Linear Porokeratosis with Dermal Amyloid Deposits – A Rare Presentation

Dear Editor,

Porokeratosis is a disorder of keratinization in which there is clonal expansion of keratinocytes with abnormal differentiation due to unknown etiopathological factors. Clinical presentation of this condition is variable, but in histopathology, all forms of porokeratosis show a vertical column of parakeratosis, known as the cornoid lamella. Porokeratosis is now considered a non-malignant clonal proliferation of epidermal keratinocytes due to a mutation in the gene coding for mevalonate kinase (MVK).[1] Linear porokeratosis is characterized by a linear arrangement of papules and plaques with an elevated peripheral rim, usually seen unilaterally on the limbs. Secondary cutaneous amyloidosis is associated with pre-existing inflammatory disorders, hamartomas, or neoplastic skin disorders. Disseminated superficial actinic porokeratosis (DSAP) is reported to be rarely associated with localized cutaneous amyloidosis.

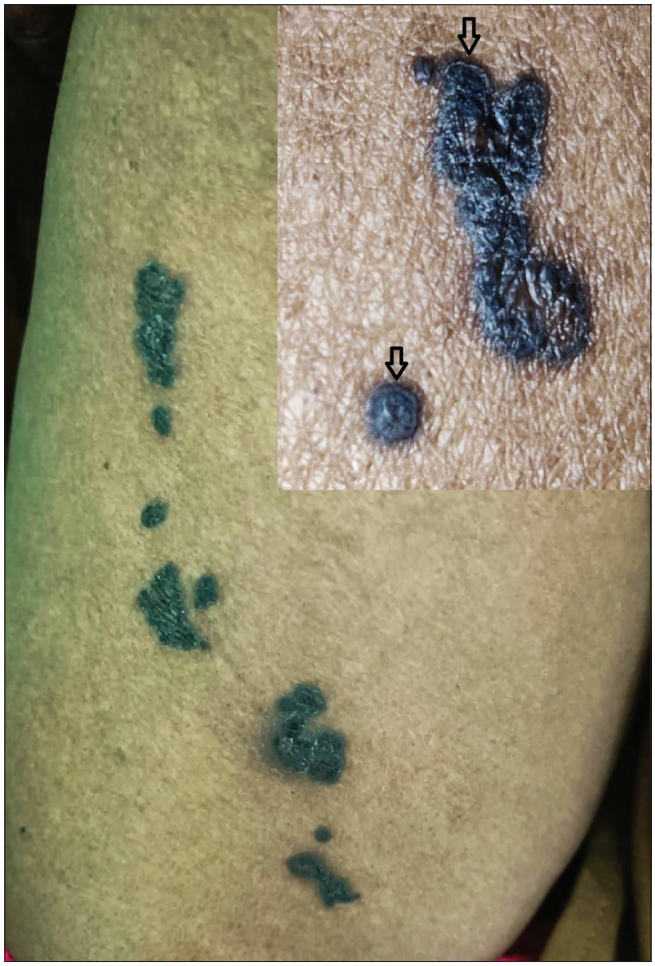

Figure 1: Verrucous linear papules on anterior thigh, inset showing close-up of peripheral raised keratotic rim (black arrow)

A 43-year-old woman presented with multiple hyperpigmented raised lesions on her right thigh of 10 years duration. It started as a single elevated lesion and then gradually increased in number and size. Newer lesions developed in a linear pattern. The lesions were mainly asymptomatic, but there was occasional pruritus. On examination, there were multiple discrete and confluent hyperpigmented verrucous papules and plaques, with some of them showing an elevated keratotic rim and well-demarcated border, distributed in a linear pattern on the anterior aspect of the right thigh [Figure 1]. We had a differential diagnosis of linear porokeratosis and a linear verrucous epidermal nevus. Dermoscopy showed irregularly pigmented areas with a double-marginated white border [Figure 2].

The hemogram, liver, and renal function tests were normal. Serological tests for syphilis and viral markers were negative. Skin biopsy specimens showed epidermis with invaginations filled with tiers of parakeratosis suggestive of cornoid lamellae [Figure 3], and the papillary dermis showed an inflammatory infiltrate admixed with melanophages and abundant amorphous eosinophilic deposits [Figure 3]. A special stain with Congo red confirmed the presence of salmon-pink amyloid deposits in the papillary dermis [Figure 4]. No atypical cells or nevoid cells were seen. We made a final diagnosis of verrucous linear porokeratosis with dermal amyloid deposits. The patient did not have any clinical signs or symptoms of

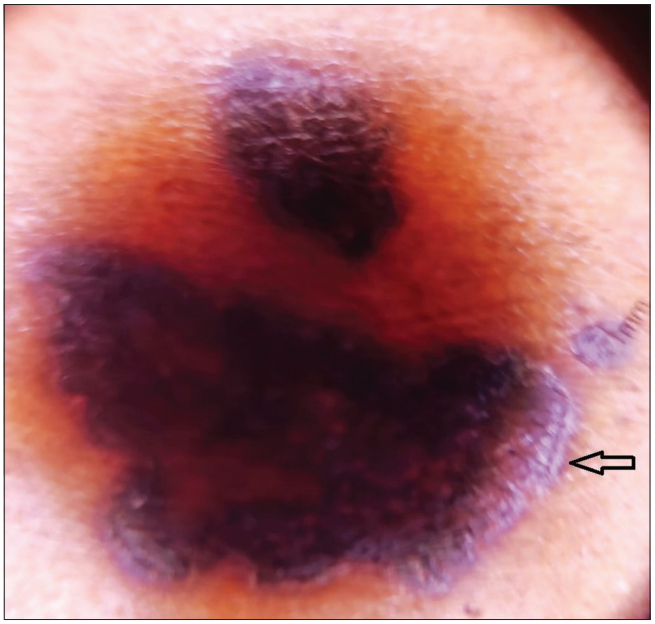

Figure 2: Irregular pigmented areas with double-marginated white border (black arrow), (Dermalite, polarised, 10x)

systemic amyloidosis. The patient was treated with topical tretinoin cream and is being planned for complete excision as the lesions are localized.

Our patient presented with verrucous linear papules and plaques with a peripheral keratotic rim; dermoscopy showed a double-marginated white border; and a skin biopsy showed the classical cornoid lamella with amyloid deposits in the papillary dermis, confirmed by Congo red stain. Therefore, we made a diagnosis of verrucous linear porokeratosis with amyloid deposits. Verrucous linear papules and plaques of long duration are also the presentation of verrucous epidermal nevus, but the presentation is at birth or soon after birth, and there are definite histopathology features to diagnose it. Porokeratoma may present with linear verrucous papules but have distinct histological features. Linear porokeratosis is a localized form of porokeratosis characterized by a linear arrangement of papules and plaques with an elevated peripheral rim, usually seen unilaterally on limbs. There is a higher risk of malignant change in linear porokeratosis (20%) than in the other forms, even though our case did not show any histological features of malignancy.[2] The histological hallmark of all types of porokeratosis in the cornoid lamella is a vertical elongated column of parakeratosis in the stratum corneum. However, cornoid lamella has been described in actinic keratosis and verruca vulgaris. The cornoid lamella represents an abnormal clone of cells that is usually not malignant. This increased chance of malignancy is attributed to the loss of alleles due to post-zygotic mutation.[3]

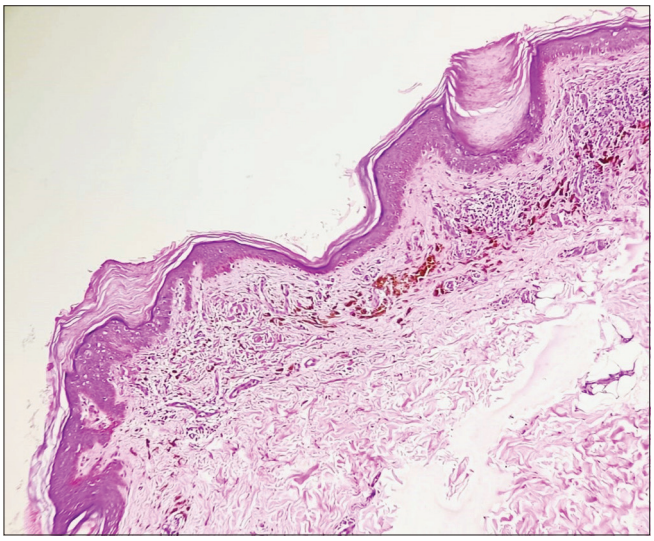

Figure 3: Epidermis showing invaginations filled with parakeratosis forming cornoid lamella, while dermis showing inflammatory infiltrate with melanophages, (H&E, 100x)

There are a few case reports of DSAP associated with dermal amyloid deposits.[4,5] The exact mechanism by which porokeratosis induces amyloid deposits is not known. Piamphongsant *et al.* suggested that the amyloid in DSAP might be derived from degenerated epidermal keratinocytes.[6] There have been reports that the amyloid deposits in porokeratosis were more evident near the cornoid lamellae, indicating that the deposition was secondary to epidermal defects, and immunohistochemical studies using anti-cytokeratin antibodies have supported this.[7,8] In our case, the duration of porokeratosis was 10 years, and the patient reported occasional pruritus in the lesion. The chronic nature of the lesion and mechanical trauma due to scratching might have contributed to secondary amyloid deposition in the present case due to epidermal keratinocyte

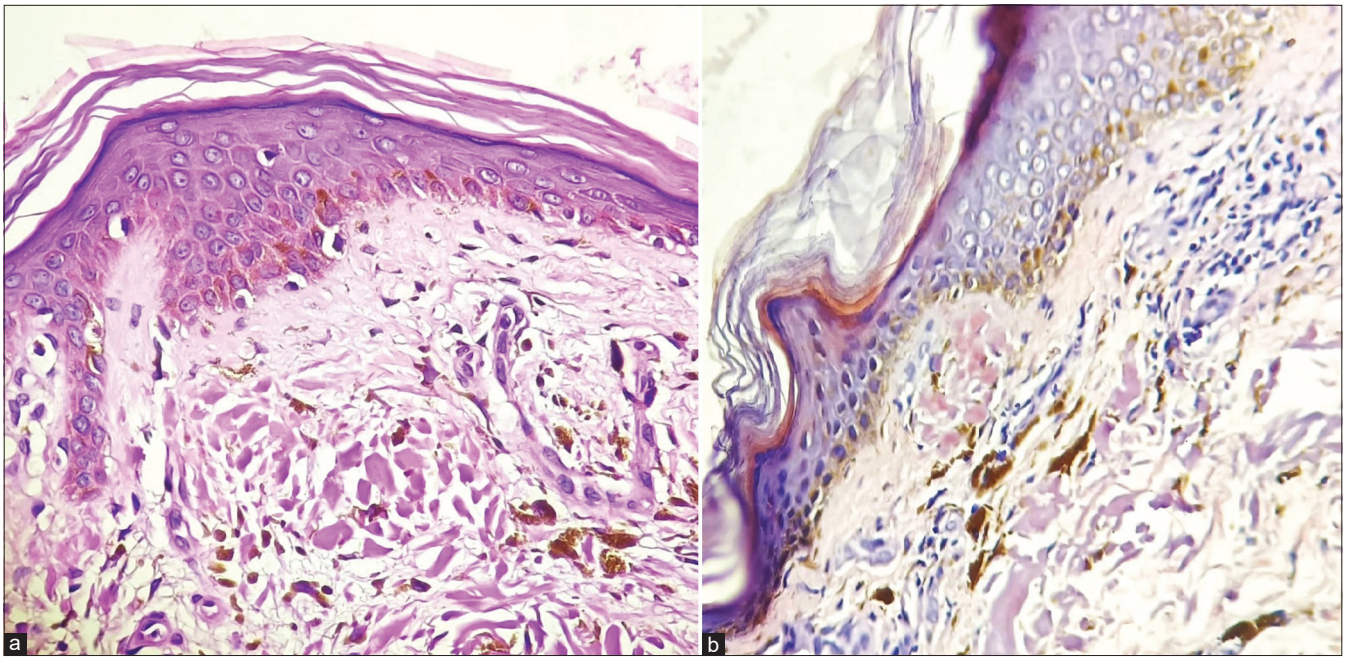

Figure 4: (a) Upper dermis showing amorphous eosinophylic amyloid deposits, (H&E, 400x), (b) Congo red stain highlighting salmon pink amyloid deposits in the upper dermis, (400x)

damage.[9] The dermoscopic findings in porokeratosis are central brown area, blue grey dots, surrounding hypopigmented bands and peripheral white tracks.

There are no definite treatment modalities for porokeratosis. The topical therapies tried are 5-florouracil, imiquimod, calcipotriol, tacrolimus, topical steroids, and tretinoin. The destructive therapies tried are dermabrasion, cryotherapy, CO2 ablative laser, Nd Yag laser, pulse dye laser, and surgical excision.[10] Acitretin is the systemic drug tried, which can also protect against malignancy. The present case is unique due to its presentation as verrucous papules and plaques, which is usually not a presentation of porokeratosis. The amyloid deposits in the dermis are another highlight of this case. Even though amyloid deposits have been reported in DSAP, we could not come across any report of linear porokeratosis with amyloid deposits to the best of our knowledge. Hence, we are reporting this case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In this form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Athira Sudhesan, Pradeep N. Sukumaran, Rashmi Ratnakaran[1], Aswathy S. Lazar

Departments of Dermatology and Venereology, [1]Pathology, Government TD Medical College, Alappuzha, Kerala, India

Address for correspondence:
Dr. Pradeep N. Sukumaran, Department of Dermatology and Venereology, Government TD Medical College, Vandanam, Alappuzha - 688 005, Kerala, India.
E-mail: dvmchtvm@yahoo.co.in

References

1. Zeng K, Zhang QG, Li L, Duan Y, Liang YH. Splicing mutation in MVK is a cause of porokeratosis of Mibelli. Arch Dermatol Res 2014;306:749-55.
2. Das A, Vasudevan B, Talwar A. Porokeratosis: An enigma beginning to unravel. Indian J Dermatol Venereol Leprol 2022;88:291-99.
3. Sasaki S, Urano Y, Nakagawa K, Nagae H, Nakanishi H, Arase S. Linear porokeratosis with multiple squamous cell carcinomas: Study of p53 expression in porokeratosis and squamous cell carcinoma. Br J Dermatol 1996;134:1151-3.
4. Husein HE, Inmaculada RM, Vicente CA, Eduardo SG. Disseminated superficial porokeratosis with dermal amyloid deposits in an elderly man: A rare entity. J Dtsch Dermatol Ges 2015;13:331-3.
5. Ramírez-Santos A, Suárez-Amor O, Pérez-Pérez L, Loureiro M, Peteiro C, Toribio J. [Localized cutaneous amyloidosis secondary to porokeratosis: A retrospective histopathologic study of 30 patients]. Actas Dermosifiliogr 2008;99:639-43.
6. Piamphongsant T, Sittapairoachana D. Localized cutaneous amyloidosis in disseminated superficial actinic porokeratosis. J Cutan Pathol 1974;1:207-10.
7. Amantea A, Giuliano MC, Balus L. Disseminated superficial porokeratosis with dermal amyloid deposits: Case report and immunohistochemical study of amyloid. Am J Dermatopathol 1998;20:86-8.
8. Jee MS, Chang SE, Suh HS, Choi JH, Sung KJ, Moon KC, *et al.* Porokeratosis ptychotropica associated with dermal amyloid deposits. Clin Exp Dermatol 2003;28:450-2.
9. Kim JH, Yim H, Kang WH. Secondary cutaneous amyloidosis in disseminated superficial porokeratosis: A case report. J Korean Med Sci 2000;15:478-81.
10. Dereli T, Ozyurt S, Ozturk G. Porokeratosis of Mibelli: Successful treatment with cryosurgery. J Dermatol 2004;31:223-7.

Access this article online

Website: https://journals.lww.com/idoj

DOI: 10.4103/idoj.idoj_297_23

Quick Response Code

How to cite this article: Sudhesan A, Sukumaran PN, Ratnakaran R, Lazar AS. Verrucous linear porokeratosis with dermal amyloid deposits – A rare presentation. Indian Dermatol Online J 2024;15:515-7.

Received: 19-Apr-2023. **Revised:** 08-May-2023.
Accepted: 11-Jun-2023. **Published:** 07-Nov-2023