A Case of Linear Porokeratosis

Dear Editor,

Porokeratosis is a disorder of keratinization characterized clinically by hyperkeratotic papules or plaques surrounded by a thread-like elevated border and histologically cornoid lamella. Linear porokeratosis is an uncommon variant that presents in early childhood, although congenital presentations have been reported. Here, we report a case presented with an atrophic plaque with hyperpigmented border over the left palm involving fingers in a linear pattern without any lesion over other parts of the body. An 18-year-old male, born out of a non-consanguineous marriage, presented to the outpatient department of a tertiary hospital with gradually progressive atrophic plaque over the left palm for two years of age. There was no history of itching, pain, irritation, or worsening of the lesion on sun exposure, and consultation was primarily sought for cosmetic reasons. There was no history of trauma, vesicle, or pustule, and family history was non-contributory.

On examination, an atrophic plaque with hyperpigmented border of size 12.5 × 1.5 cm was noted in a linear distribution over the left palm, extending from the middle of the palm proximally to the tip of the third and fourth digits distally [Figure 1]. No nail, hair, mucous membrane, or trunk involvement was observed. Systemic examination revealed no abnormality.

Our differential diagnoses were localized linear porokeratosis, linear verrucous epidermal nevus, and nevoid psoriasis. Biopsy was taken from the peripheral hyperpigmented border, which showed the presence of cornoid lamella with a parakeratotic column overlying a small vertical zone of dyskeratotic and vacuolated cells within the epidermis [Figure 2a and b]. Focal loss of the granular layer was seen. A mild lymphocytic infiltrate was seen around an increased number of capillaries in the underlying dermis. A diagnosis of localized linear porokeratosis was established based on clinical and histological features.

He has been prescribed topical 5% 5-fluorouracil (5-FU) cream twice a day, which resulted in a very good clinical response within three months [Figure 3]. The patient was informed about the malignant risk associated with his condition and instructed to follow up every six months to monitor the development of any malignant changes.

Subtypes of porokeratosis include classic porokeratosis of Mibelli, disseminated superficial porokeratosis (DSP), disseminated superficial actinic porokeratosis (DSAP), linear porokeratosis, porokeratosis palmaris et plantaris disseminata, and punctate porokeratosis.

There is a higher risk of malignant change in linear porokeratosis (20%) than in other forms of porokeratosis such as porokeratosis of Mibelli (7.5%), porokeratosis palmaris et plantaris, and DSAP (3.4%).[1] Early identification of linear porokeratosis is important due to its malignant transformation to basal cell carcinoma or, more commonly, squamous cell carcinoma.[2] It is a chronic disease with no tendency for spontaneous resolution. Treatment with topical 5-FU,[3] potent topical steroids, tacrolimus 0.1%, imiquimod 5%, cryotherapy, carbon dioxide laser, pulsed dye laser, topical or systemic retinoids,[4] and surgical excision may be considered.

5-FU is a structural analog of uracil, and its metabolites block the DNA synthesis of rapidly proliferating cells. Other indications of 5-FU include actinic keratosis,

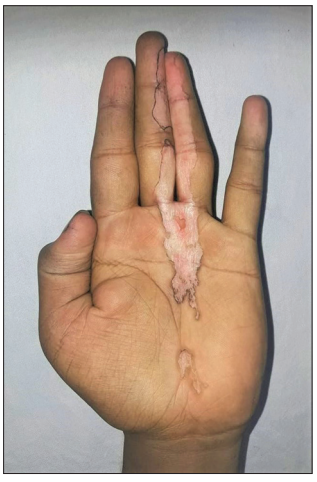

Figure 1: An atrophic plaque with hyperpigmented border in a linear distribution over the left palm

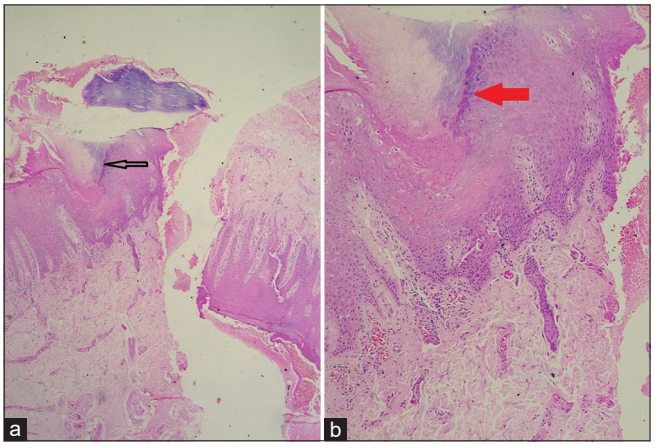

Figure 2: (a) Histopathology (H and E stain, 10×) showed the presence of cornoid lamella (black arrow) with a parakeratotic column overlying a small vertical zone of dyskeratotic and vacuolated cells within epidermis. (b) Histopathology (H and E stain, 40×) showed the presence of cornoid lamella (red arrow)

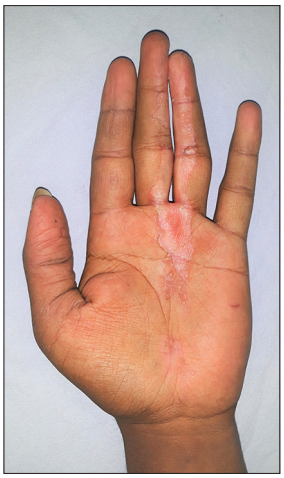

Figure 3: After three months of topical 5% 5-fluorouracil application

superficial basal cell carcinoma, keratoacanthoma, squamous cell carcinoma *in situ*, Bowen's disease, and actinic cheilitis. Till now, 5-FU has been used in six case reports, and none of the cases were linear porokeratosis.[5] To the best of our knowledge, 5-FU has not been used in cases of linear porokeratosis, and our case shows a very good clinical response. Closer disease surveillance (six monthly) is needed in cases of linear porokeratosis lesions because it has more chances of malignant transformation.

Declaration of patient consent

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

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Access this article online

Website: http://journals.lww.com/IDOJ

DOI: 10.4103/idoj.idoj_408_22

Quick Response Code

How to cite this article: Rai T, Ansari MH. A case of linear porokeratosis. Indian Dermatol Online J 2023;14:552-3.

Received: 28-Jul-2022. **Revised:** 11-Oct-2022.
Accepted: 18-Oct-2022. **Published:** 25-May-2023.