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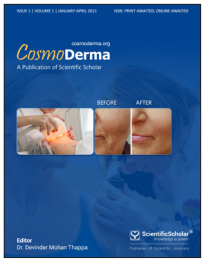

Letter to the Editor

A case of mucosal porokeratosis in a middle-aged female in a tertiary health-care center in South India: A case report

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Dear Sir

Porokeratosis represents a disorder of unknown etiology with several clinical variants having been described.[1] Although described first by Neumann in 1875, it was an Italian dermatologist Mibelli who is credited with naming the lesion as porokeratosis due to the involvement of eccrine ostia in his patient.[2] Being rare in presentation, accurate data regarding its prevalence are lacking.[1] Although the prevalence of certain subtypes may have a geographical variation [disseminated superficial actinic porokeratosis (DSAP) being more common in the white population of Australia, as they are subjected to more ultraviolet radiation], no racial or ethnic predilection has been noted in general.[1] Photoexposed sites like face and extremities are most commonly affected, followed by trunk, genitalia, and mucosa.[3] The classically described morphology is that of an atrophic plaque rimmed by an elevated ridge and having characteristic histopathological appearance of cornoid lamellation.[1] In spite of a theoretical risk of malignant transformation, the actual incidence varies between the subtypes. Treatment options include topical retinoids and immunomodulators, and systemic retinoids, or even surgical procedures for small, localized lesions.[2]

We report here the case of a 48-year-old female, a farmer by occupation, who had complaints of white discoloration over her lips for the past 8 years associated with dysphagia and occasional burning sensation over the lips for the past 8 months. She was a habitual tobacco chewer for several years. Intermittent treatment taken from nearby health facility was of no help to her. She did not give any history of ulceration. There were no other lesions elsewhere or any history to suggest connective tissue disorder.

Physical examination revealed a large depigmented, atrophic plaque over the full extent of upper lip (and extending over to the philtrum) and left half of the lower lip [Figure 1]. There were smaller discrete circular plaques above the right upper lip [Figure 2]. A thread-like ridge bordered the plaque [Figure 3]. Closer inspection also revealed intraoral extension of the atrophic plaque [Figure 4]. She was noted to have marked poor oral hygiene with dental caries and nicotine staining of teeth.

The differentials considered were discoid lupus erythematosus, porokeratosis, actinic cheilitis, and lupus vulgaris. A punch biopsy was performed from the edge of the lesion and routine histopathological examination revealed the classical cornoid lamellation – which is basically a tall, wedge-shaped column of parakeratotic cells. Just beneath the mound of parakeratosis, the granular layer was sparse compared to the surrounding epidermis. A mild perivascular chronic inflammatory infiltrate in the dermis was noted [Figure 5].

Thus, a diagnosis of mucosal porokeratosis was made, bearing the clinical observation of a lacy ridge-like rim around an atrophic plaque and the histopathological presence of cornoid

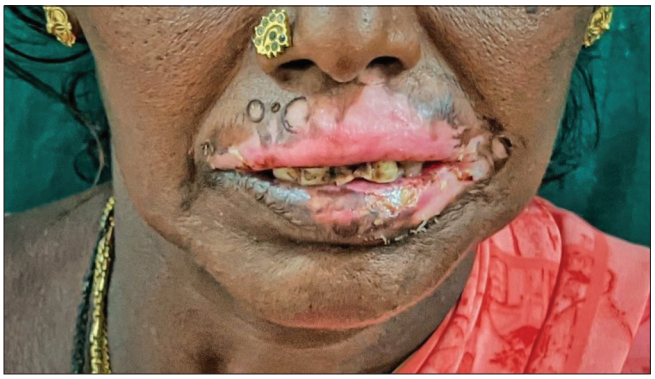

Figure 1: Front profile of the patient – with the externally visible extent of the atrophic plaque.

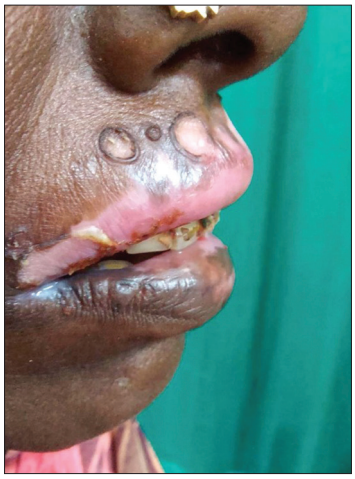

Figure 2: Side-on view of the lesion – smaller discrete plaques in close proximity to the main plaque.

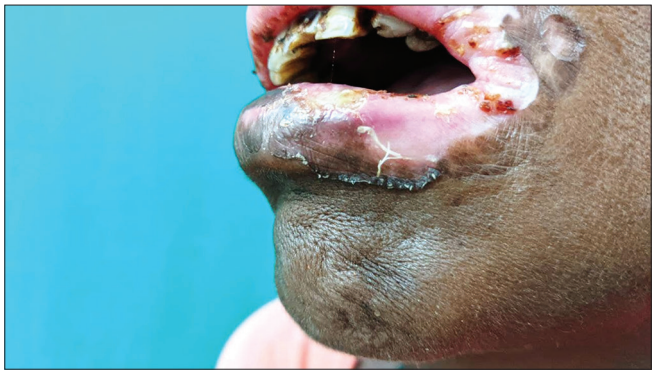

Figure 3: Typical ridge like rim of the lesion as visible externally.

lamellation. She was prescribed topical 5-fluorouracil (an immunomodulator) and symptomatic treatment for burning (with lignocaine-containing gel). However, the patient was lost to follow-up.

Porokeratosis is a disorder of keratinization most commonly occurring due to UV exposure or immunosuppression.[1]

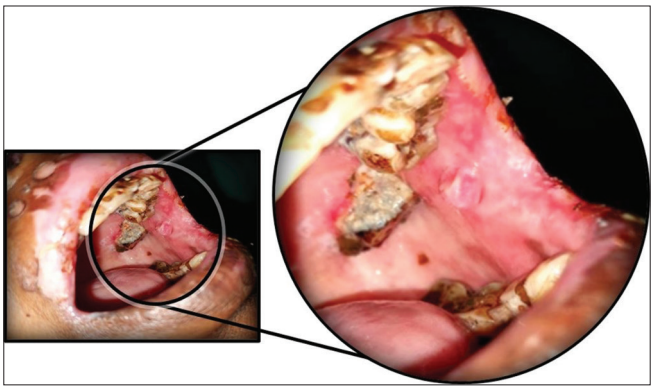

Figure 4: Intraoral extension of the plaque.

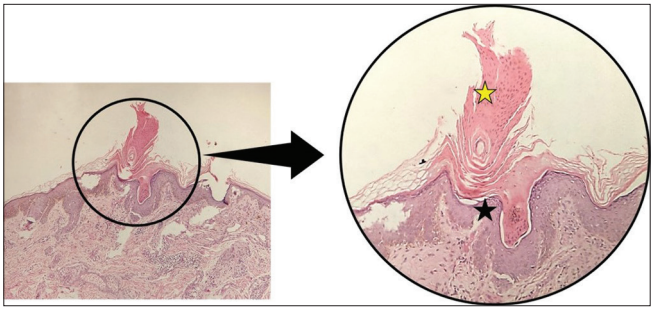

Figure 5: Hematoxylin and Eosin staining of the biopsy specimen (taken from the edge of the discrete plaque [Figure 2] – showing column of parakeratosis (yellow star) overlying area of hypogranulosis (black star) – ×10 magnification and ×40 magnification (inset).

While the Porokeratosis of Mibelli is the most commonly described variant, other entities like DSAP, disseminated superficial porokeratosis, and linear porokeratosis are also described. Rarer presentations include the likes of porokeratosis ptychotropica, punctate porokeratosis, genital porokeratosis, and mucosal porokeratosis. Porokeratosis with mucosal involvement has been reported only in few case reports – either as part of disseminated disease,[4] or solitary labial lesion[5] or involving both lip and oral mucosa.[3] Such lesions may be confused with discoid lupus erythematosus plaque or with atrophic annular lichen planus[5] and are difficult to treat. The treatment found useful in one such case report of mucosal porokeratosis was topical 5-fluorouracil[3] – similar to what we had prescribed. However, because the patient was lost to follow-up, the response could not be assessed. It is advisable to keep this diagnosis in the differentials if an atrophic plaque with mucosal extension/involvement and raised margin is encountered and a biopsy study should be carried out wherever possible to rule out differentials and due to the risk of malignant transformation that has been reported occasionally.

Declaration of patient consent

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

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Nil.

Conflicts of interest

There are no conflicts of interest.

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