A Case of Solitary Labial Porokeratosis Extending into Oral Mucosa Treated with Topical 5-Fluorouracil

Sir,

Porokeratosis is an uncommon clonal disorder of keratinisation characterized clinically by solitary or multiple annular plaques with central atrophy and a distinct ridge-like border. Classically, five clinical variants have been described: porokeratosis of Mibelli, disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis (DSAP), porokeratosis palmaris et plantaris disseminate, and linear porokeratosis.[1] The usual sites of predilection are extremities, trunk, face and can occur very rarely on mucous membranes and genitals.[2] We hereby report a case of labial porokeratosis with oral mucosa involvement.

A 40 years old female patient presented with a solitary raised lesion over upper lip since 6 months. The lesion was asymptomatic except for occasional burning sensation. The lesion started at the center of the upper lip and gradually progressed centrifugally. On examination, a single well-defined skin colored to hyperpigmented annular plaque of size approximately 4 × 3 cm with atrophic center and prominent raised margins was present over upper lip extending into mucosal surface of the lip (Figure 1). Examination of oral mucosa revealed poor oral hygiene. The patient was further evaluated with differential diagnosis of discoid lupus erythematous and labial porokeratosis of Mibelli. Dermatoscopic examination (MINI 3000 LED Dermatoscope, HEINE, Germany) showed a pale area bounded by irregular double-marginated white track border (Figure 2a and b). A punch biopsy was performed from the margin of the lesion and histopathological examination revealed hyperkeratosis, epidermal invaginations with dense parakeratotic column (cornoid lamella), underlying hypogranulosis, and mild superficial dermal perivascular infiltrate suggestive of porokeratosis [Figure 3]. The patient was started on topical 1% 5-Fluorouracil cream to be applied once daily at night. The patient showed good response to treatment after 16 weeks [Figure 4].

Porokeratosis of Mibelli usually occurs in children with a genetic predisposition and autosomal dominant inheritance but can also be secondary to risk factors including sun exposure, immune suppression, and ultraviolet exposure. It is considered that the lesions of porokeratosis result from the peripheral expansion of an abnormal, mutant clone of epidermal keratinocytes located at the base of the parakeratotic column.[1] Though porokeratosis can involve any part of the body, lesions over scalp, mucous membrane, palms and soles and genitilia are regarded uncommon.[1] Very few cases of porokeratosis of lip and oral mucosa have been reported. Tongue and buccal mucosal lesions were present as a part of disseminated porokeratosis whereas involvement of lip has been reported as a solitary lesion.[1-4] Porokeratosis is characterized by presence of column of parakeratotic cells termed as cornoid lamella and undergoing hypogranulosis. The diagnostic finding on dermoscopy is the presence of double-marginated peripheral border often described as white track-like border or lines of volcanic crater.[5] Treatment options include topical 5-fluorouracil, topical and oral retinoids, topical imiquimod, cryotherapy, photodynamic therapy, carbon dioxide laser ablation, and dermabrasion.[1] Excision may be required considering premalignant potential of...
In the present case, excellent response to treatment with topical 5-fluorouracil (5-FU) was seen. It is a fluorinated pyrimidine which disrupts DNA synthesis by disrupting thymidine synthesis. This leads to cytotoxic activity towards rapidly dividing cells in the S phase and also produces an inflammatory response due to hyperproliferative nature of porokeratosis. Both topical and systemic 5-FU has been found to be effective in porokeratosis.\[6\]

Involvement of lip and oral mucosa in porokeratosis is very rare and difficult to treat. Topical 1% 5-fluorouracil is well tolerated on lip and oral mucosa and further studies are necessary to know the tolerability, efficacy, and duration of treatment with 5-fluorouracil.

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.

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Nil.

Conflicts of Interest

There are no conflicts of interest.

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