Giant Verrucous Porokeratosis with Psoriasiform Alteration: A Rare Entity with Atypical Clinico-histopathologic Features

Sir,

Porokeratosis is an inherited, clonal disorder of epidermal keratinization, classically characterized by the development of single or multiple annular lesions with raised, sharply marginated, keratotic ridge, and central atrophy.\(^1\) This clinical presentation may vary with different types of porokeratosis but histopathologically unified by the consistent finding of “cornoid lamella.” Here, we document an atypical and rare case of “giant verrucous porokeratosis” which showed psoriasiform histopathological changes at the center of the lesion.

A 25-year-old male, with no premorbid illness, presented with multiple verrucous plaques over the right side of the back, left lateral aspect of chest and abdomen, nape of the neck, right arm, and abdomen for the past 8 years. The lesions were mildly pruritic and scaly. Cutaneous examination revealed a total of five lesions characterized by well-defined, whitish, scaly, verrucous plaque without any apparently raised margin. Two of such lesions were giant in size involving the whole right half of the back and left lateral aspect of trunk and one was over the nape of neck [Figures 1 and 2]. No other sites were affected including scalp, mucosa, and nails. There was no family history of psoriasis or porokeratosis, and the patient had not received any prior allopathic treatment for this condition. Routine laboratory parameters were within normal limits.

For histopathological evaluation, two samples of punch biopsy were taken, one from the center of plaque and other from the edge. The sample taken from the edge incidentally demonstrated cornoid lamella as vertical column of poorly stained parakeratotic cells with pyknotic nuclei, running through the normally stained surrounding cells along with absence of granular layer in its floor and hypergranulosation of wall. Dyskeratotic keratinocytes in the spinous layer were also present along with perivascular patchy lichenoid lymphocytic infiltrates and focal basal vacuolar changes. All these findings were in favor of porokeratosis [Figure 3a and b]. Other sample taken from the center of the lesion revealed characteristic psoriasiform changes such as orthohyperkeratosis, foci of parakeratosis, attenuated granular layer, and moderate papillomatosis. In dermis, dilated tortuous capillaries at the tip of clubbed dermal papilla and moderately dense superficial perivascular lymphocytic infiltrate were seen supporting the psoriasiform changes [Figure 3c and d].

The presence of typical cornoid lamella at the margin of verrucous plaque and other histopathologic findings led us to diagnose it as “verrucous porokeratosis” with psoriasiform changes. The patient was kept on oral isotretinoin (30 mg daily) and topical salicylic acid, but he did not turn up further.

There are many well-recognized clinical variants of porokeratosis: porokeratosis of Mibelli, disseminated superficial porokeratosis, disseminated superficial actinic porokeratosis (DSAP), giant porokeratosis, palmaris et plantaris disseminata, linear porokeratosis, and punctate porokeratosis.\(^2\) An another variant was reported by Helfman and Poulos in 1985, but first described in detail by Lucket et al. in 1995 who termed it as “porokeratosis ptychotropica” (ptyche means fold) owing to its characteristic flexural involvement in genitogluteal region.\(^3\) Due to its verrucous morphology, it was also known as hyperkeratotic or “verrucous porokeratosis.” Since its first description, <40 cases have been reported worldwide to the best

Figure 1: Giant scaly verrucous plaque over (a) the right half of the back and (b) left lateral aspect of chest and abdomen extending to the right scapular area

Figure 2: Similar verrucous plaque over the nape of neck
Gene expression profiling of porokeratosis demonstrates alterations at the center of the lesion which can be treated with topical 5% imiquimod, Vitamin D analogs, oral retinoic acid derivatives, or can be surgically excised if feasible.\textsuperscript{3,6}

of our knowledge. Of note, we were unable to find a single case of “verrucous porokeratosis” which affected sites other than genitogluteal region, buttocks, and extremities (as in our case).

The association of psoriasis and porokeratosis is long known, but definitely rare and their pathogenetic evolution is still an enigma. Recently, in a study, a similar pattern of upregulation of certain proteins such as keratins (keratins 16, 6A, 6B, and 17), specific calcium-binding proteins (psoriasin 1, calgranulin A, calgranulin B, and calgizarin), and gap junction proteins (connexin 26 and 30) was noted in psoriasis and porokeratosis which favors a genetic relationship between these two dermatoses.\textsuperscript{4} In 2006, a classification was proposed to categorize this association into three groups.\textsuperscript{5} The first group includes cases of porokeratosis mimicking psoriasis, wherein verrucous porokeratotic plaques are usually localized to the natal cleft and buttocks known as “porokeratosis pschytropica.”\textsuperscript{13,6} The second group includes patients presenting with classical clinical lesions of porokeratosis along with psoriasiform changes at the center of the porokeratotic plaque.\textsuperscript{7} The third group consists of patients with psoriasis and DSAP induced by topical corticosteroid. J Cutan Pathol 2006;33:516-8.

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Conflicts of interest
There are no conflicts of interest.

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