

Porokeratotic Variant of Lichen Planus

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Abstract

Porokeratotic Variant of Lichen Planus: Lichen planus (LP) is a well-known dermatologic disorder with various different clinical presentations. We present a case of patient who was found to have porokeratotic lichen planus, a previously unreported variant of LP.

Keywords: Porokeratosis; Lichen planus

Case Report

A 64 year-old female presented with a 6-month history of a suddenly appearing scaly rash on her back, chest, and upper extremities. She denied pruritus or discomfort associated with the rash. The patient endorsed a 16-pound intentional weight loss over the past two years, but otherwise review of systems was negative. Physical examination revealed scattered pink annular papules and plaques with elevated rim of scale on the trunk and upper extremities (Figure 1A). A clinical diagnosis of eruptive porokeratosis was made. Histopathologic examination of a lesion from the back revealed a disruption of the stratum corneum with a small aggregate of parakeratotic cells forming a column, focal hypergranulosis and acanthosis, basal cell vacuolar change and colloid bodies (Figure 2). The papillary dermis showed a lichenoid inflammatory infiltrate; these findings were consistent with lichen planus with porokeratosis (Figure 2).

Upon follow up the patient had developed new lesions and complained that her existing lesions had become pruritic and had spread to her mouth and genital region. She also endorsed a recent biopsy of an oral lesion performed by her dentist. Examination revealed red-purple papules on the inner lip, superior buccal mucosa, hard palate and right labia majora. The pathology of an oral biopsy revealed vacuolar change in the basal layer of the mucosa, and a band-like infiltrate of lymphocytes and occasional plasma cells in the upper submucosa. There were many annular atrophic purple and hyperpigmented papules and plaques with rim of scale over the back (Figures 1A and 1B), bilateral shoulders and right groin. Punch biopsy of the lesion from the right labia majora revealed hyperkeratosis, hypergranulosis, irregular acanthosis and focal vacuolar change in the basal cell layer; marked edema was present in the upper dermis with an infiltrate of lymphocytes and extravasated erythrocytes confirming the diagnosis of LP. (Figure 3) The patient was treated with fluocinonide 0.05% ointment and tacrolimus 0.1% ointment with subsequent improvement. Hepatitis panel was found to be negative.

Discussion

Porokeratoid lichen planus (PLP) is a clinical variant of LP that to our knowledge has not been previously described in the literature. While lesions became pruritic and spread to involve the oral mucosa

and genitalia, locations classic for LP, our patient initially had an asymptomatic eruption on her trunk clinically consistent with eruptive porokeratosis. Individual lesions had a strikingly consistent peripheral rim of scale and were located primarily on the upper trunk and upper extremities (Figures 1A and 1B). While the clinical presentation was similar to porokeratosis, histologically our patient's lesions demonstrated an imperfect cornoid lamella. Instead, the tissue showed a disruption of the stratum corneum with a poorly defined column of cells, and no dyskeratotic cells were noted at the base of the column.

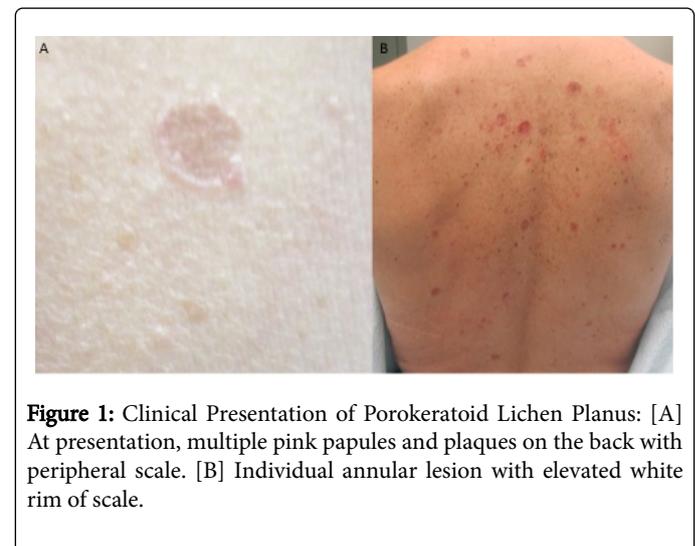


Figure 1: Clinical Presentation of Porokeratoid Lichen Planus: [A] At presentation, multiple pink papules and plaques on the back with peripheral scale. [B] Individual annular lesion with elevated white rim of scale.

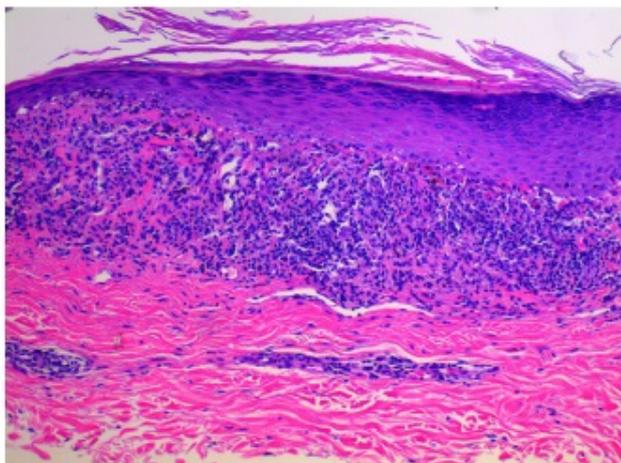


Figure 2: Histologic Findings of Porokeratoid Lichen Planus Hematoxylin-eosin (H+E) stain reveals focal basal cell vacuolization and exocytosis of lymphoid cells. 10X.

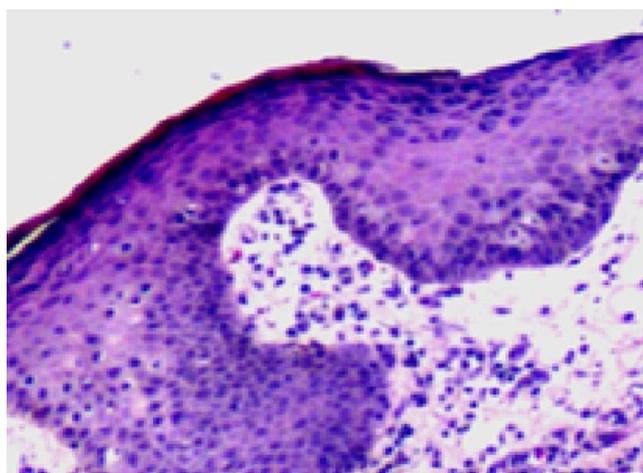


Figure 3: Histologic findings of R labia majora biopsy.H+E stain reveals hypergranulosis, marked upper dermal edema with an infiltrate of lymphocytes and occasional extravasated erythrocytes. (Bulla not present clinically; likely this was an artefact of processing.) 40X.

Of the well-known variants of LP, our patient's rash most resembled the annular variant characterized by annular violaceous plaques classically involving the male genitalia, axilla, groin, and extremities [1,2]. This particular variant of LP is asymptomatic especially when arising in the groin, which differs clinically from our patient's pruritic rash.

There are a few case reports of a rare annular atrophic variant of LP (AALP) that looks similar on physical exam to PLP variant found in our patient. With this AALP variant, lesions begin as purple violaceous papules that grow peripherally to form lesions with an atrophic and hyperpigmented center and raised borders [3]. However, our case was different from the reported cases of AALP because our patient had mucosal involvement. Such involvement was not found in any patients with reported AALP. In addition, the annular rim of both annular LP and AALP has been described as red-brown or violaceous and hyperpigmented [1,3] whereas the lesions in our patient had a scaly white rim evocative of those occurring in porokeratotic lesions. Finally, AALP shows elastolytic activity histologically which was not appreciated in our patient's pathology.

Of note, eruptive porokeratosis has been associated with several internal malignancies particularly those of the gastrointestinal system but including ovarian carcinoma and lymphoma as well [4,5]. One report shows that 30% of patients with eruptive porokeratosis have a recently diagnosed malignancy with a mean onset of rash 2.7 months prior to or after diagnosis [5]. In light of this evidence, our patient underwent an age-appropriate cancer screening which came back negative. It is unknown whether this porokeratotic variant of LP carries a similar association with malignancy and perhaps, remains a question warranting further investigation.

References

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