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Prurigo Pigmentosa: An under-recognized inflammatory dermatosis characterized by an evolution of distinctive clinicopathological features

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Cover Quizlet

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Figures 1 and 2 are depicted on the journal cover.

Figure 3.

Figure 4.

Figure 5.

Figure 6.

Your diagnosis?

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Your diagnosis?

Discussion follows on page 811
Prurigo Pigmentosa: An under-recognized inflammatory dermatosis characterized by an evolution of distinctive clinicopathological features

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Prurigo pigmentosa (PP) was originally described 45 years ago, yet because of its variable histopathological manifestations it remains an under-recognized inflammatory dermatosis clinically characterized by recurrent pruritic, erythematous macules, urticarial papules and papulovesicles that eventuate in symmetric reticulated pigmentation.1 Although the majority of cases have been reported in women of Asian descent, it also has been described in various ethnicities worldwide. Of the approximately 400 reported cases of PP, fewer than 10 have been reported from the United States.1–7

Due to the relative rarity of this condition, we present two cases from the United States to illustrate the clinicopathological spectrum of PP. Both patients were healthy young women who presented with recurrent episodes of pruritic erythematous macules and subsequently developed urticarial plaques, papules and/or papulovesicles which eventuated in reticulated pigmentation. The clinical and histological findings for these patients are summarized in Table 1.

Clinically PP presents as a recurrent intensely pruritic, symmetrically distributed eruption located on the neck, chest, upper back, lumbar-sacral region and abdomen, and more rarely, the forehead or arms. To date, there are no reports of mucous membrane involvement.5,7 The lesions tend to progress through several stages of development, initially starting as erythematous macules which then evolve to urticarial papules and papulovesicles. Subsequently the lesions become crusted or scaly and within a few weeks spontaneously resolve, leaving behind reticulated pigmentation. Due to the recurrent nature of the eruption, patients often have multiple lesions at various stages of development.1 The clinical differential diagnosis is broad, but the distribution of the lesions and the reticulated pattern helps narrow the possibilities.
Table 1. Clinicohistopathological presentations of patients 1 and 2

<table>
<thead>
<tr>
<th></th>
<th>Patient 1</th>
<th>Patient 2</th>
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<tbody>
<tr>
<td><strong>Clinical Findings</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ethnicity</td>
<td>Palestinian descent</td>
<td>Mexican descent</td>
</tr>
<tr>
<td>Duration</td>
<td>24-36 months</td>
<td>29-months</td>
</tr>
<tr>
<td>Anatomic location</td>
<td>Chest (Figs. 1 and 3)</td>
<td>Central chest and breast (Fig. 6), flank, sacrum,</td>
</tr>
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<td></td>
<td></td>
<td>buttock (Fig. 7) and focally on left elbow</td>
</tr>
<tr>
<td>Exacerbating factors</td>
<td>Pregnancy</td>
<td>Heat and exercise</td>
</tr>
<tr>
<td>Serological findings</td>
<td>None</td>
<td>Elevated WBC with left shift</td>
</tr>
<tr>
<td>Treatments</td>
<td>Minocycline-improvement</td>
<td>Topical steroids-minimal benefit.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Minocycline-resolution</td>
</tr>
<tr>
<td><strong>Histopathological findings</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Evolving lesion</td>
<td>Mild spongiosis, sparse perivascular infiltrate and dermal edema. (Fig. 4)</td>
<td>Epidermal spongiosis associated with a perivascular lymphocytic infiltrate and focal vascular degeneration.</td>
</tr>
<tr>
<td>Early lesions</td>
<td>Superficial dermal mixed inflammatory infiltrate associated with neutrophilic spongiosis. (Fig. 2)</td>
<td>Brisk vascular degeneration with numerous apoptotic keratinocytes often concentrated near acrosyringium. Superficial perivascular lymphocytic infiltrate with increased eosinophils. (Fig. 8)</td>
</tr>
<tr>
<td>Fully developed lesions</td>
<td>Dense mixed dermal infiltrate associated with combined balloon degeneration and spongiosis, and clusters of necrotic keratinocytes. (Fig. 5)</td>
<td>Parakeratosis, focal apoptotic keratinocytes, and a superficial perivascular lymphocytic infiltrate containing melanophages. (Fig. 9)</td>
</tr>
<tr>
<td>Resolving/late lesions</td>
<td>Mild spongiosis and a sparse perivascular lymphocytic infiltrate.</td>
<td></td>
</tr>
</tbody>
</table>

Although the histopathological findings were originally felt to be nonspecific, distinctive histological features have now been identified for each clinical stage.\(^1,8-11,13,17\) Early lesions present as pruritic erythematous macules or urticarial papules, and a biopsy from lesions at this stage shows a sparse, perivascular and interstitial, predominantly neutrophilic infiltrate involving the superficial dermis with focal epidermal exocytosis and variable papillary dermal edema. Fully developed lesions manifest as excoriated papules, vesicles, and papulovesicles which corresponds histologically to varying degrees of spongiosis accompanied by ballooning associated with epidermal necrosis and focal collections of neutrophils. The dermal inflammatory infiltrate at this stage is denser, and often mixed, with increased numbers of lymphocytes, neutrophils and scattered eosinophils. In late lesions, there is slight epidermal spongiosis and focal parakeratosis associated with a mild superficial perivascular lymphocytic infiltrate and variable vacuolar degeneration. Often scattered apoptotic keratinocytes and melanophages are readily found. Therefore, the findings in any given biopsy are reflective of the clinical stage of the lesion, and represent only a single point on an evolving continuum of clinical and histologic features. Depending on the stage of development, the histopathological differential diagnosis includes dermatitis herpetiformis, irritant contact dermatitis, linear IgA disease, neutrophilic dermatoses, erythema multiforme, adult onset Still’s disease, as well as various spongiotic and interface dermatitides.\(^1,3,6\) Although the reticulated pigmentation can be clinically impressive, histologically it is non-distinctive, showing only a superficial perivascular lymphocytic infiltrate associated pigment-laden macrophages, which is indistinguishable from other inflammatory dermatoses resulting in postinflammatory hyperpigmentation.\(^1\) Therefore, accurate diagnosis requires detailed clinicopathological correlation with attention to the dynamic clinical and histopathological course of the disease.

The etiopathogenesis is unknown, but various hypothesizes have been postulated and include friction, contact allergens, photosensitivity, infectious agents, and metabolic disorders such as diabetes mellitus, fasting, anorexia and ketosis.\(^1,3,5,9,11,14\) To date, none have shown a consistent causal relationship; however, symptoms are often exacerbated by hot weather and/or sweating.\(^1,5,15\) There have been a few case reports of PP associated with other diseases, including autoimmune conditions, adult-onset Still’s disease, atopy and pregnancy; however,
these associations may merely be coincidental rather than causative.1,5,12-15 Ilkovitch and Patton recently posited PP might represent an inflammatory variant of confluent and reticulated papillomatosis,16 but that hypothesis is not supported histologically and those conditions are quite distinct.

Most patients with PP experience spontaneous resolution, followed by recurrent episodes of exacerbations that can last months to years.1,9,11 The condition responds well to minocycline, doxycycline and dapsone, presumably due to the anti-inflammatory properties of these medications, as well as inhibition of the migration and function of neutrophils.1,5,9,18,19 Yet notably, the condition does not respond to treatment with corticosteroids or anti-histamines.1,5 Lastly, there has been one report of 2 patients who responded well to weekly Jessner’s peels and irradiation with an 830-nm light emitting diode.20

In conclusion, although PP is an uncommon inflammatory dermatoses it can be readily recognized by its distinctive clinicopathological features. Awareness of the dynamic changes that occur over the course of the eruption is essential to make an accurate diagnosis.
References