Prurigo Pigmentosa: A Case Report in a Caucasian Female

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Background

Prurigo pigmentosa is a rare inflammatory disease that is uncommon in Caucasian women. It is typically seen in young Japanese women and has been proposed to be due to genetic and environmental factors they are exposed too. We present a case of a 36-year-old Caucasian female with one year history of pruritic eruption on her anterior chest. Treatment included doxycycline, however it is discussed whether to work up associated systemic diseases such as Still's disease, Helicobacter pylori infection, and Sjögren's syndrome in such cases where genetics and environmental factors may not have a major influence. Prurigo pigmentosa is a rare inflammatory disease otherwise known as Nagashima's disease, first described in 1971 in eight young Japanese women by dermatologist Masaharu Nagashima. 1 It presents as a pruritic, erythematous papulo-vesicular rash most commonly found on the back, chest, and neck. 2 Prurigo pigmentosa is typically seen in young Japanese women. The pathogenesis is still relatively unclear; however, it has been described as having three distinct clinical and histopathological stages. Prurigo pigmentosa has been linked to several systemic conditions including Still's disease, Helicobacter pylori infection, and Sjögren's syndrome. The condition typically resolves with oral antibiotics such as minocycline, doxycycline, or macrolide antibiotics.

Methods

Retrospective review of the clinical and pathologic findings of a case of prurigo pigmentosa and review of related cases published in the literature

Objectives

1) Present a unique case of a prurigo pigmentosa in a patient that is being closely monitored and evaluated for underlying conditions
2) Raise awareness among clinicians of clinical features most suggestive of prurigo pigmentosa
3) Inform providers about the importance of both underlying factors screening at time of presentation and Treatment options for prurigo pigmentosa

Clinical Case Description

We present a 36-year-old Caucasian female with a one year history of pruritic eruption on her upper anterior chest. Prior treatments included over-the-counter antibacterial soap and aveeno cream, which provided no relief of the itching and no improvement in rash. Patient does not have any other known dermatologic history and does not endorse other rashes, fevers, chills, or weight loss. On physical examination, there was a reticulated erythematous scaly plaque with some areas of hyperpigmentation on the anterior chest. Initial clinical impression was consistent with late stage prurigo pigmentosa. Other differentials included CARP and Grover's. Patient was prescribed doxycycline 100 mg twice daily as well as Triamcinolone acetonide 0.1% cream to use as needed for pruritus up to three times daily. Patient followed up, by telephone one week later reporting significant improvement in both the cutaneous eruption and pruritus.

Clinical Figure

Clinical Findings: Reticulated erythematous scaly plaque with some areas of hyperpigmentation on the anterior chest

Histologic Features

Histological findings: confluent parakeratotic crust, mild spongiosis, and scattered dyskeratotic cells. In the dermis, there is a moderate perivascular lymphocytic infiltrate

Stages Of Prurigo Pigmentosa

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<thead>
<tr>
<th>Stage</th>
<th>Clinical Differential</th>
<th>Histological Differential</th>
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<tbody>
<tr>
<td>Early</td>
<td>Contact dermatitis, dermatis herpetiformis, linear IgA dermatosis, psoriasis vulgaris, systemic lupus erythematosus, urticaria</td>
<td>Dermatitis herpetiformis, dermatophytosis, leukocytoclastic vasculitis, linear IgA dermatosis, psoriasis vulgaris, systemic lupus erythematosus</td>
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<tr>
<td>Fully-developed</td>
<td>erythema multiforme, mucha habermann disease (PLEVA)</td>
<td>Erythema multiforme, mucha habermann disease (PLEVA)</td>
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<tr>
<td>Late</td>
<td>Confluent and reticulated papillomatosis; Dowling-Degos disease; erythema ab igne; erythema dyschromicum perstans; lichen planus pigmentosus; macular amyloidosis; medication-induced pigmentation</td>
<td>Post-inflammatory hyperpigmentation</td>
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Discussion and Conclusions

Fewer than 50 cases have been described in non-Japanese patients. 2 It is worth discussing whether clinicians should work up the associated systemic conditions in a case where hereditary or genetic factors may not predominate. It is hypothesized that ketosis-induced neutrophil-mediated inflammation contributes to the development of this eruption. There have been several patients where elevated urine and blood ketone levels were reported, including patients with past medical history of diabetes mellitus, anorexia nervosa, or bariatric surgery. This helps explain why minocycline and doxycycline that affect neutrophil chemotaxis are an effective treatment for prurigo pigmentosa.

Additional Work-Up

Autoimmune
- Still's Disease
- Sjögren's

Medical Workup
- Helicobacter Pylori infection
- Diabetes mellitus
- Anorexia nervosa
- History of Bariatric Surgery

Treatment

Previous Treatment Regimen:
- Refractory to antibacterial soap, aveeno cream

New Treatment Regimen:
- Triamcinolone acetonide .1% cream applied up to three times daily
- Doxycycline- 100 mg twice daily

References