

**Answer to Dermacase** *continued from page 1023*

**4. Progressive pigmented purpuric dermatosis**

Progressive pigmented purpuric dermatosis, also known as Schamberg disease, is the most common of the pigmented purpuric dermatoses (PPD). The PPD constitute a spectrum of chronic benign disorders characterized by a distinct symmetrical petechial rash within an orange-brown macular base, usually confined to the lower extremities.<sup>1-3</sup> Histopathology reveals superficial subepithelial and perivascular infiltration of lymphocytes and macrophages without overt vasculitis.<sup>1,2</sup> The cardinal feature differentiating PPD from other purpuric eruptions is the characteristic nonpalpable punctate purpura and orange-brown pigmentation resulting from extravasation of erythrocytes and hemosiderin deposition in the superficial dermis.<sup>1,3</sup>

The etiology of PPD has yet to be elucidated, but current evidence suggests the involvement of cytokine and cell-mediated immunity.<sup>1</sup> Familial presentation is very rare; however, there have been case reports documenting a possible autosomal-dominant inheritance pattern.<sup>4</sup> A number of precipitating factors have been reported: venous hypertension; exercise and gravitational dependency; capillary fragility; allergy to dyes, clothing, or alcohol; focal infections; and drug exposure.<sup>1</sup> Medications that are commonly linked with PPD are listed in **Box 1**. Pigmented purpuric dermatoses have also been associated with other medical conditions, including hepatic disease, malignancies, diabetes mellitus, hyperlipidemia, rheumatoid arthritis, lupus erythematosus, thyroid dysfunction, hereditary spherocytosis, hematological disorders, and porphyrias.<sup>1</sup>

**Differential diagnosis**

The differential diagnosis of PPD includes stasis dermatitis, leukocytoclastic vasculitis, purpuric contact dermatitis, and early mycosis fungoides (**Table 1**).

Stasis dermatitis is a complication of long-standing venous insufficiency and is commonly associated with hyperpigmentation, varicose veins, and dependent edema. Scaling, crusting, and ulcerations might be

**Box 1. Drugs associated with PPD**

- Acetaminophen
- Acetylsalicylic acid
- Carbromal
- Chlordiazepoxide
- Glipizide
- Glybuzole
- Hydralazine HCl
- Meprobamate
- Dipyridamole
- Reserpine
- Thiamine
- Interferon alfa
- Medroxyprogesterone acetate

HCl—hydrochloric acid, PPD—pigmented purpuric dermatoses.

present, but the characteristic punctate petechiae of PPD are not observed.

Leukocytoclastic vasculitis, also known as hypersensitivity vasculitis, is a small-vessel vasculitis named after its diagnostic histopathology on skin biopsy. It might localize to the skin or be accompanied by systemic manifestations. Cutaneous vasculitis presents as painful palpable purpura on the lower extremities, which can occasionally ulcerate.<sup>5</sup> Evidence of autoimmune diseases, acute or chronic illnesses, and ingestion of drugs with known causative associations might be suggestive of this disorder. Skin biopsy confirms the diagnosis.

Purpuric contact dermatitis, an allergic or toxic reaction to textile dyes and resins, can produce purpuric macules, papules, patches, and nodules.<sup>6</sup> Diagnosis is usually prompted by the distribution and appearance of the lesions along with a history of exposure. Patch testing can be helpful for confirmation; however, avoidance of the suspected textile can be both diagnostic and therapeutic.<sup>6</sup>

Mycosis fungoides is a cutaneous T-cell lymphoma with a potential for lymphatic and hemopoietic involvement.<sup>7</sup> It initially presents as erythematous patches

**Table 1. Differential diagnoses for PPD**

DIAGNOSIS	CLINICAL CHARACTERISTICS	CONFIRMATION
Progressive pigmentary dermatoses	Nonpalpable cayenne pepper–like purpura on an orange-brown macular base	Biopsy only if clinical diagnosis is in question
Stasis dermatitis	Hyperpigmentation, varicose veins, dependent edema; scaling, crusting, and ulcerations might be present	Underlying venous insufficiency
Leukocytoclastic vasculitis	Painful palpable purpura on lower extremities, which might ulcerate	Skin biopsy
Purpuric contact dermatitis	Purpuric macules, papules, patches, and nodules following textile distribution	Avoidance of suspected contactant
Early mycosis fungoides	Initial erythematous patches affecting lower trunk and buttocks, progressing to infiltrated plaques and tumours	Skin biopsy

PPD—pigmented purpuric dermatoses.

affecting the lower trunk and buttocks, and progresses to infiltrated plaques and tumours.<sup>7</sup> Diagnosis is confirmed by skin biopsy.

## Investigations

There are currently no established guidelines for the investigation of PPD. When diagnosis is unclear, complete blood count, peripheral blood smear, platelet count and function, prothrombin time, partial thromboplastin time, erythrocyte sedimentation rate, antinuclear antibody, and rheumatoid factor should be considered to exclude other causes of purpura.<sup>1-3</sup> A skin biopsy is useful to confirm the clinical diagnosis and to rule out other potentially serious conditions that can mimic PPD.

## Treatment

Although diagnosis is quite straightforward, PPD are slow to resolve and rather resistant to treatment. As PPD can be quite distressing, it is important for practitioners to reassure patients that PPD are benign disorders with no systemic sequelae. There is no current medical intervention shown to be beneficial; however, topical corticosteroids and antihistamines might alleviate pruritus, and the use of compression stockings is recommended.<sup>1</sup> Individual case reports have documented success with immunomodulating therapies such as psoralen photochemotherapy, griseofulvin, pentoxifylline, and cyclosporine.<sup>1</sup> Current recommendations suggest discontinuing suspected provoking factors and beginning a trial of topical corticosteroids for 4 to 6 weeks.<sup>1-3</sup> 

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### Competing interests

None declared

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