



Pigmented Purpuric Dermatoses

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ABSTRACT

Pigmented purpuric dermatoses (PPD) are a group of histopathologically similar conditions that are primarily differentiated based on morphology. The basic pathological finding is a lymphocytic perivascular infiltrate with hemorrhage limited to the papillary dermis without fibrinoid necrosis of the vessels. The etiology is unknown; they run a chronic course and are fairly resistant to treatment. We present this review for the physicians to kindle interest in this not-so-uncommon entity.

Keywords: Pigmented purpuric dermatoses, Purpura, Schamberg's disease.

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INTRODUCTION

Pigmented purpuric dermatoses (PPD) are a group of histopathologically similar conditions that are primarily differentiated based on morphology. They are generally categorized into five groups: Progressive PPD (i.e., Schamberg's disease), purpura annularis telangiectodes (i.e., Majocchi's disease), lichen aureus, PPD of Gougerot-Blum, and eczematid-like purpura of Doucas-Kapetanakis.^{1,2} A few other rarer variants have also been described in the literature (Table 1).³⁻¹² However, this classification does not change the management or prognosis.^{1,2}

Epidemiology

It is difficult to quantify the epidemiology of PPD as it is an uncommon diagnosis in India and elsewhere. Nevertheless, Sharma and Gupta found the prevalence to be 0.18% of the total patient input over a period of 1.5 years.¹³ Pigmented purpuric dermatoses appear to

be four times more frequent in males than in females.¹³ They may occur at any age,^{1,2} but most commonly in the 3rd or 4th decade.¹³ They are infrequently reported in preadolescent children and adolescents,² the commonest type being Schamberg's,¹⁴ although the pigmented purpuric lichenoid dermatoses variant is not reported in this age group.¹⁰ Lichen aureus and Majocchi disease predominantly occur children and young adults.^{1,2,10}

Etiology (Secondary PPD)

Idiopathic cases are the most common (70%) and mostly resemble Schamberg's disease. Although multiple underlying causes have been outlined (Table 2)^{1,15-22}, most of them lack specificity.²³ Drugs are responsible for 14% of the cases and comprise the most frequent among the provocative factors.^{23,24} Multiple reports citing PPD as a manifestation of mycosis fungoides occur in the literature,²⁵⁻²⁷ and few have even preceded typical mycosis fungoides. Interestingly, recent reports have also suggested the role of contact allergens as possible etiological agents in PPD. Engin et al²⁸ showed 54% of the PPD patients were positive to one or more allergens following patch test.

Pathogenesis

The pathogenesis of PPDs is not well understood. The most widely accepted mechanism is increased capillary dilation and fragility, with resultant rupture of the papillary dermis capillaries and possible aneurismal dilation of the end capillaries.^{13,29} Venous hypertension, exercise, or gravitational dependency are commonly associated findings.¹ Also, some of the previously mentioned etiologies may be the precipitating factors.

In addition, recent data suggests the role of cell-mediated immune responses.¹³ Perivascular infiltrate in Schamberg's disease consists primarily of CD3+, CD4+, and CD1a+ dendritic cells (DCs) (i.e., Langerhans cells) in a well-defined pattern, with close spatial contact between the lymphocytes and DCs.³⁰ Conditions like contact dermatitis and graft-*vs*-host disease, which are thought to have an immune/cell-mediated mechanism, share a similar pattern of inflammatory cells. The modulation of cellular adhesion molecules in dermal endothelial cells (ICAM-1, ELAM-1) and in lymphocytes (LFA-1) suggests a mechanism for lymphocyte trafficking into affected area

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Table 1: Classification of pigmented purpuric dermatoses

Sl. no.	Types	Synonyms	First description
1	Purpura annularis telangiectodes	Majocchi's disease	Majocchi ³
2	Progressive PPD	Schamberg's disease	Schamberg ⁴
3	Itching purpura	Eczematoid purpura of Doucas and Kapetanakis; disseminated pruriginous angiodermatitis	Doucas and Kapetanakis ⁵ ; Lowenthal ⁶
4	Pigmented purpuric lichenoid dermatosis	Gougerot–Blum syndrome	Gougerot and Blum ⁷
5	Lichen purpuricus	Lichen aureus	Martin ⁸ ; Calnan ⁹
6	Granulomatous PPD		Saito and Matsuoka ¹⁰
7	Linear PPD		Hersh and Shwayder ¹¹
8	Transitory PPD		Osment et al ¹²

PPD: Pigmented purpuric dermatoses

Table 2: Etiological factors for secondary pigmented purpuric dermatoses

Etiological factors	Associations
Drugs ¹⁵⁻¹⁹	Antihypertensives (CCBs, beta-blockers, ACE inhibitors, nitrites, furosemide, and other diuretics), analgesics (aspirin, paracetamol, NSAIDs), antihistamines, antidepressants, chlorthalidone, carbamazepine, glipizide, bezafibrate, medroxyprogesterone acetate, raloxifene, pseudoephedrine, vitamin B1 derivatives, interferon-alpha (in hepatitis C infection), antibiotics (ampicillin and cotrimoxazole), polyvinyl pyrrolidone, topical 5-fluorouracil
Food additives	Tartrazine, creatine supplements
Contact irritants and allergens ²⁰	Metals, dyes, clothing, alcohol ingestion
Infections ^{1,21,22}	Beta-hemolytic streptococci, toxoplasma, rickettsiae, hepatitis virus B and C, dental infections
Vascular diseases	Stasis dermatitis
Neoplasia	Mycosis fungoides
Collagen vascular disorders.	Lupus erythematosus, rheumatoid disease

CCBs: Calcium channel blockers; ACE: Angiotensin-converting enzyme; NSAIDs: Non-steroidal anti-inflammatory agents

of inflammation and their interaction with endothelial cells and DCs.¹³ Disappearance of the inflammatory infiltrate from affected areas following treatment with topical steroids and daily psoralen plus ultraviolet A (PUVA) further consolidates the immune theory.³⁰ C3 and C1q deposits on the wall of lesional blood vessels from patients with pigmented purpuras suggest that immune complexes may also play a role in the pathogenesis of these diseases.¹⁵

Clinical Features

Progressive Pigmented Purpuric Dermatitis^{1,4,31}

Patients are mostly young adult males, but PPDs may occur at any age including childhood. The lesions are most frequent on the legs but may occur anywhere on



Figs 1A and B: Patient of progressive pigmented purpuric dermatosis: (A) Multiple purpuric macules and papules interspersed among hyperpigmented macules giving the classical cayenne-pepper appearance; (B) few of them coalescing to form orange plaques

the body and may be few in number or very extensive. They consist of pinhead-sized reddish puncta resembling grains of cayenne-pepper that further form irregular plaques of orange or brown pigmentation (owing to hemosiderin) (Figs 1A and B). It is usually asymptomatic, although there may be some slight itching. The eruption is characteristically very chronic and may persist for many



years. The pattern of the eruption changes, with slow extension and often some clearing of the original lesions. Spontaneous cure may occur eventually.

Purpura Annularis Telangiectodes

It may occur at any age, but patients are more likely to be adolescents or young adults. Lesions occur at any site, often in the absence of venous stasis, and may be few in number or very numerous. Early lesions are bluish-red annular macules in which dark-red telangiectatic puncta appear. The central part of the lesion fades, with peripheral extension giving it the annular configuration, and sometimes, slight atrophy is noticed in the center.^{1,31}

Pigmented Purpuric Lichenoid Dermatitis of Gougerot and Blum

This eruption occurs especially in men aged between 40 and 60 years. It is characterized by minute, lichenoid papules that tend to fuse into plaques of various hues, in association with purpuric lesions similar to those of Schamberg's disease. Lesions are usually seen on the legs and rarely on the trunk and thighs.^{1,31}

Lichen Aureus

This is a more localized, more intensely purpuric but often asymptomatic eruption that may have rather lichenoid morphology. It presents as sudden-onset lichenoid papules in association with purpuric lesions, seen commonly on the lower limbs and occasionally on the trunk and the face. The lesions are often solitary and may be yellowish, golden, rust-colored, or purple.^{1,31}

Itching Purpura

Except for being extensive, rapid in onset, and associated with persistent, severe pruritus, it is more or less similar to Schamberg's disease. The lesions consist of erythematous and purpuric macules that may become confluent starting at the ankles and may spread to involve the entire lower limbs. Spontaneous improvement after a few months is usual, but recurrences may occur and a fluctuating but chronic course is frequent. The itching may respond to topical corticosteroids and oral antihistamines.^{1,5,6}

Miscellaneous

Granulomatous variant consisting of a PPD-like eruption on the dorsum of the feet, with the biopsy revealing a superimposed granulomatous infiltrate, has been reported. Linear¹¹ and quadrantic³² or zosteriform³³ morphology of PPD has also been described. Isolated reports of familial occurrence,^{34,35} autosomal dominant³⁶ in nature, have been seen in patients with Schamberg's disease.

Transitory PPD was described by Osment et al, includes entities like angioma serpiginosum, and is different from other PPDs, though the clinical differences between them are minor. Itching purpura of Lowenthal is considered to be a more symptomatic variant of Schamberg's disease.³¹

Dermoscopy

A dermatoscope is a noninvasive, diagnostic tool that magnifies subtle clinical surface features of skin lesions as well as unveils some subsurface skin structures not normally visible even with a magnifying lens. Dermoscopic findings in progressive PPD change according to the evolution of the lesions (Table 3, Fig. 2).³⁷

Table 3: Dermoscopic findings in progressive pigmented purpuric dermatoses³⁷

Stage	Description
Early	Red brownish or red coppery background, brighter or comparatively clearer with a few red dots, and globules.
Established	Red brownish or red coppery background, red dots, globules and patches are seen prominently. Some gray dots and a network of brownish to gray interconnected lines, which are less prominent.
Resolving	Background becomes more brownish coppery and reddish tinge reduces. Central red globules and dark brown pigment deposit indicate hemosiderin deposits in periphery. Gray dots and a network of brownish to gray interconnected lines are seen
Old or completely resolved	Pigment deposits appear blackish brown. There are no red globules and dots.



Fig. 2: Dermoscopic appearance of established lesions of progressive pigmented purpuric dermatosis: Multiple red dots and globules, majority of which are intermixed and surrounded by dark brown dots and interconnected lines

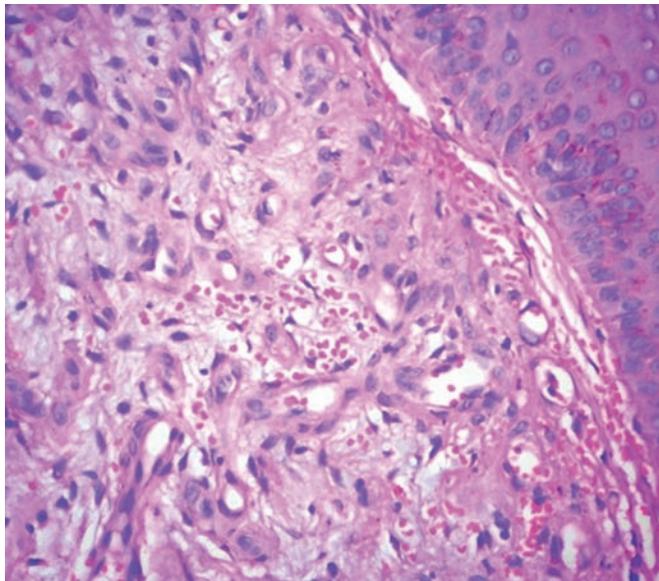


Fig. 3: High-power view of the dermis showing multiple extravasated red blood cells and few perivascular lymphocytes; no signs of leukocytoclastic vasculitis (H&E, 400 ×)

Histopathology^{23,31,38}

Despite morphological variations giving rise to a clinical classification, the basic histopathology is more or less similar. There is a perivascular infiltrate of lymphocytes and macrophages centered on the superficial small blood vessels of the skin with endothelial cell swelling and narrowing of the lumina. However, overt vasculitis is not usually observed. Extravasation of red blood cells (Fig. 3) with marked hemosiderin deposition in macrophages is typically seen, though the degree of hemosiderin deposition may be variable. Special stains, such as Perls stain and Fontana-Masson may be used to demonstrate hemosiderin, which also helps to differentiate PPD from stasis dermatitis, where a deeper deposition is seen. When lymphocytic infiltrate is lichenoid, it is diagnosed as lichenoid dermatosis of Gougerot and Blum;³⁹ when spongiosis/neutrophils are marked, the diagnosis is itching purpura;⁴⁰ and a rare variant has been described with superimposed granulomatous infiltrate.¹⁰

The cellular infiltrate in all types contains CD4+ T cells in close contact with CD1a+ Langerhans cells.^{30,41} IgA-associated lymphocytic vasculopathy has been described,⁴² but its relevance is vague, and because direct immunofluorescence is often negative, it is not recommended.

Differential Diagnoses

The most important differential diagnosis is leukocytoclastic vasculitis, but differentiation based on palpable purpura, associated pain, and features of vasculitis on histopathology is not difficult. Other conditions that present with purpuric lesions, such as purpuric clothing

Table 4: Newer therapeutic options

Therapeutic option	Proposed mechanism of action in PPD
Bioflavonoids (rutin) and ascorbic acid ⁴³	Increases capillary resistance; inhibits specific enzymes that are activated in inflammation; potent antioxidative radical scavenging activities
Pentoxifylline ⁴⁴	Reduces expression of adhesion molecules (ICAM-1 expression on keratinocytes and E-selectin expression on endothelium in upper dermis); interferes with the T-cell/keratinocyte adherence resulting in inhibition of exocytosis of lymphocytes to the epidermis
Griseofulvin ⁴⁵	Immunomodulatory effect
PUVA ^{46,47}	Immunomodulation with alteration in the activity of the T lymphocyte and the concomitant suppression of IL-2 production
Cyclosporine A ⁴⁸	Inhibitor of CD4+ T cells, which predominate in the inflammatory infiltrate in PPD

PPD: Pigmented purpuric dermatoses; PUVA: Psoralen plus ultraviolet A

dermatitis, hyperglobulinemic purpura, early mycosis fungoides, purpuric clothing dermatitis, stasis pigmentation, scurvy, and drug hypersensitivity reactions need to be considered and ruled out.³¹

Treatment

No therapy has proven benefit as PPD tends to persist for years and is very resistant to treatment. Nevertheless, reassurance, avoidance of leg dependency, and support hosiery seem the most appropriate approach. Topical corticosteroids and antihistamines may take care of associated pruritus.^{1,2}

Some evidence supports the use of topical steroids to control and/or improve PPD. A therapeutic trial may be given for 4 to 6 weeks, although their prolonged use is best avoided. Few newer therapeutic options have been suggested in some reports, but the level of evidence is poor (Table 4)⁴⁵⁻⁴⁸. More studies to outline effective treatment strategies need to be carried out.

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