

Pigmenting Purpuric Dermatoses: Striking But Not a Manifestation of COVID-19 Infection

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PRACTICE POINTS

- Dermatologists should be aware of the clinical presentations of pigmenting purpuric dermatoses (PPDs).
- Certain PPDs may resemble the thromboembolic events seen in COVID-19. Clinicians should especially be aware of how to differentiate these benign pigmentary disorders from other serious conditions.
- Teledermatology is widely utilized, but caution may be prudent when evaluating erythematous or purpuric dermatoses, especially those of the lower extremities.
- Pigmenting purpuric dermatoses generally are benign and do not require immediate treatment.

Purpuric or erythematous eruptions of the lower extremities can be perplexing, especially because this clinical sign has been associated with COVID-19 infection. Since the start of the pandemic, COVID-19 often has been the first differential diagnosis for many clinical findings; however, pigmented purpuric dermatoses (PPDs) are distinctive, reassuring, eponymous, and not linked to this virus. In this article, we differentiate the PPDs and delineate them from potentially serious differential diagnoses, including inflammatory retiform purpura, leukocytoclastic vasculitis, stasis dermatitis, and cellulitis.

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Pigmented purpuric dermatoses (PPDs) are characterized by petechiae, dusky macules representative of postinflammatory hyperpigmentation and dermal hemosiderin, and purpura generally localized to the lower extremities. They typically represent a spectrum of lymphocytic capillaritis, variable erythrocyte extravasation

from papillary dermal blood vessels, and deposition of hemosiderin, yielding the classic red to orange to golden-brown findings on gross examination. Clinical overlap exists, but variants include Schamberg disease (SD), Majocchi purpura, Gougerot-Blum purpura, eczematoid purpura of Doucas and Kapetanakis (DK), and lichen aureus.¹ Other forms are rarer, including linear, granulomatous, quadrant, transitory, and familial variants. It remains controversial whether PPD may precede or have an association with cutaneous T-cell lymphoma.² Dermoscopy usually shows copper-red pigmentation in the background, oval red dots, linear vessels, brown globules, and follicular openings. Although these findings may be useful in PPD diagnosis, they are not applicable in differentiating among the variants.

Pigmented purpuric dermatoses can easily be mistaken for stasis dermatitis or cellulitis, as these may occur concomitantly or in populations at risk for all 3 conditions, such as women older than 50 years with recent trauma or infection in the affected area. Tissue biopsy and clinical laboratory evaluation may be required to differentiate between PPD from leukocytoclastic vasculitis or the myriad causes of retiform purpura. Importantly, clinicians also should differentiate PPD from the purpuric eruptions of the lower extremities associated with COVID-19 infection.

Pigmented Purpuric Dermatoses

Schamberg Disease—In 1901, Jay Frank Schamberg, a distinguished professor of dermatology in Philadelphia, Pennsylvania, described “a peculiar progressive pigmentary disease of the skin” in a 15-year-old adolescent boy.³ Schamberg disease is the most common PPD, characterized

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by pruritic spots resembling cayenne pepper (Figure 1) with orange-brown pigmented macules on the legs and feet.⁴ Although platelet dysfunction, coagulation deficiencies, or dermal atrophy may contribute to hemorrhaging that manifests as petechiae or ecchymoses, SD typically is not associated with any laboratory abnormalities, and petechial eruption is not widespread.⁵ Capillary fragility can be assessed by the tourniquet test, in which pressure is applied to the forearm with a blood pressure cuff inflated between systolic and diastolic blood pressure for 5 to 10 minutes. Upon removing the cuff, a positive test is indicated by 15 or more petechiae in an area 5 cm in diameter due to poor platelet function. A positive result may be seen in SD.⁶

Histologically, SD is characterized by patchy parakeratosis, mild spongiosis of the stratum Malpighi, and lymphoid capillaritis (Figure 2).⁷ In addition to CD3⁺, CD4⁺, CD8⁺, CD1a⁺, and CD36⁺ lymphocytes, histology also may contain dendritic cells and cellular adhesion molecules (intercellular adhesion molecule 1, epithelial cell adhesion molecule 1) within the superficial perivascular infiltrate.⁸ There is no definitive therapy, but first-line interventions include emollients, topical steroids, and oral antihistamines. Nonpharmacologic management includes compression or support stockings, elevation of the lower extremities, and avoidance of offending medications (if identifiable).¹

Majocchi Purpura—Domenico Majocchi was a renowned Italian dermatologist who described an entity in 1898 that he called *purpura annularis telangiectodes*, now also known as *Majocchi purpura*.⁹ It is more common in females, young adults, and children. Majocchi purpura has rarely been reported in families with a possible autosomal-dominant inheritance.¹⁰ Typically, bluish-red annular macules with central atrophy surrounded by hyperpigmentation may be seen on the lower extremities, potentially extending to the upper extremities.¹ Treatment of Majocchi purpura remains a challenge

but may respond to narrowband UVB phototherapy. Emollients and topical steroids also are used as first-line treatments. Biopsy demonstrates telangiectasia, pericapillary infiltration of mononuclear lymphocytes, and papillary dermal hemosiderin.¹¹

Gougerot-Blum Purpura—In 1925, French dermatologists Henri Gougerot and Paul Blum described a pigmented purpuric lichenoid dermatitis known as *Gougerot-Blum purpura*,¹² a rare PPD characterized by lichenoid papules that eventually coalesce into plaques of various colors, along with red-brown hyperpigmentation.⁴ As with other PPD variants, the legs are most involved, with rare extension to the trunk or thighs. The plaques may resemble and be mistaken for Kaposi sarcoma, cutaneous vasculitis, traumatic purpura, or mycosis fungoides. Dermoscopic examination reveals small, polygonal or round, red dots underlying brown scaly patches.¹³ Gougerot-Blum purpura is found more commonly in adult men and rarely affects children.⁴ Histologically, a lichenoid and superficial perivascular infiltrate composed of lymphocytes and macrophages is seen. Various therapies have been described, including topical steroids, antihistamines, psoralen plus UVA phototherapy, and cyclosporin A.¹⁴

Eczematoid Purpura of Doucas and Kapetanakis—In 1949, Greek dermatologists Christopher Doucas and John Kapetanakis observed several cases of purpuric dermatosis similar in form to the “pigmented purpuric lichenoid dermatitis” of Gougerot-Blum purpura¹² and to the “progressive pigmentary dermatitis” of Schamberg disease.³ After observing a gradual disappearance of the classic yellow color from hemosiderin deposition, Doucas and Kapetanakis described a new bright red eruption with lichenification.¹⁵ Eczematoid purpura of Doucas and Kapetanakis is rare and predominantly seen in middle-aged males. Hyperpigmented or dark brown macules may develop bilaterally on the legs, progressing to the thighs



FIGURE 1. Schamberg disease. Dusky, red-brown, nonscaling macules resembling cayenne pepper on the legs.

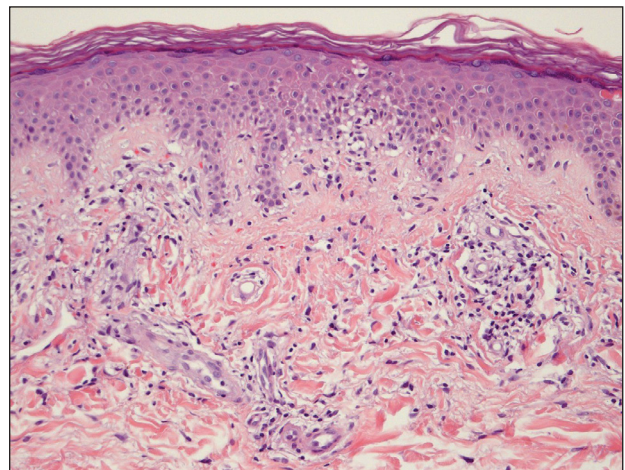


FIGURE 2. Histopathology of pigmented purpuric dermatoses. Orthokeratosis and focal spongiosis overlying a superficial perivascular lymphocytic infiltrate with occasional extravasated erythrocytes (H&E, original magnification ×20).

and upper extremities. Unlike the other types of PPD, DK is extensive and severely pruritic.⁴

Although most PPD can be drug induced, DK has shown the greatest tendency for pruritic erythematous plaques following drug usage including but not limited to amlodipine, aspirin, acetaminophen, thiamine, interferon alfa, chlorthalidone, and isotretinoin. Additionally, DK has been associated with a contact allergy to clothing dyes and rubber.⁴ On histology, epidermal spongiosis may be seen, correlating with the eczematoid clinical findings. Spontaneous remission also is more common compared to the other PPDs. Treatment consists of topical corticosteroids and antihistamines.¹⁶

Lichen Aureus—Lichen aureus was first observed by the dermatologist R.H. Martin in 1958.¹⁷ It is clinically characterized by closely aggregated purpuric papules with a distinctive golden-brown color more often localized to the lower extremities and sometimes in a dermatomal distribution. Lichen aureus affects males and females equally, and similar to Majocchi purpura can be seen in children.⁴ Histopathologic examination reveals a prominent lichenoid plus superficial and deep perivascular lymphocytic infiltrate, extravasated erythrocytes, papillary dermal edema, hemosiderophages, and an unaffected epidermis. In rare cases, perineural infiltrates may be seen. Topical steroids usually are ineffective in lichen aureus treatment, but responses to psoralen plus UVA therapy also have been noted.¹⁷

Differential Diagnosis

COVID-19–Related Cutaneous Changes—Because COVID-19–related pathology is now a common differential diagnosis for many cutaneous eruptions, one must be mindful of the possibility for patients to have PPD, cutaneous changes from underlying COVID-19, or both.¹⁸ The microvascular changes from COVID-19 infection can be variable.¹⁹ Besides the presence of erythema along a distal digit, manifestations can include reticulated dusky erythema mimicking livedoid vasculopathy or inflammatory purpura.¹⁹

Retiform Purpura—Retiform purpura may occur in the setting of microvascular occlusion and can represent the pattern of underlying dermal vasculature. It is nonblanching and typically stellate or branching.²⁰ The microvascular occlusion may be a result of hypercoagulability or may be secondary to cutaneous vasculitis, resulting in thrombosis and subsequent vascular occlusion.²¹ There are many reasons for hypercoagulability in retiform purpura, including disseminated intravascular coagulation in the setting of COVID-19 infection.²² The treatment of retiform purpura is aimed at alleviating the underlying cause and providing symptomatic relief. Conversely, the PPDs generally are benign and require minimal workup.

Leukocytoclastic Vasculitis—The hallmark of leukocytoclastic vasculitis is palpable purpura, often appearing as nonblanchable papules, typically in a dependent distribution such as the lower extremities (Figure 3). Although it

primarily affects children, Henoch-Schönlein purpura is a type of leukocytoclastic vasculitis with lesions potentially similar in appearance to those of PPD.²³ Palpable purpura may be painful and may ulcerate but rarely is pruritic. Leukocytoclastic vasculitis represents perivascular infiltrates composed of neutrophils, lymphocytes, and occasionally eosinophils, along with karyorrhexis, luminal fibrin, and fibrinoid degeneration of blood vessel walls, often resulting from immune complex deposition. Leukocytoclastic vasculitis may affect blood vessels of any size and requires further clinical and laboratory evaluation for infection (including COVID-19), hypercoagulability, autoimmune disease, or medication-related reactions.²⁴

Stasis Dermatitis—Stasis dermatitis, a chronic inflammatory condition stemming from retrograde venous flow due to incompetent venous valves, mimics PPD. Stasis dermatitis initially appears as demarcated erythematous plaques, fissures, and scaling of the lower legs bilaterally, usually involving the medial malleolus.²⁵ With time, the affected region develops overlying brawny hyperpigmentation and fibrosis (Figure 4). Pruritus or pain are common features, while fissures and superficial erosions may heal and recur, leading to lichenification.

Although both commonly appear on the lower extremities, duplex ultrasonography may be helpful to distinguish PPDs from stasis dermatitis since the latter occurs in the context of chronic venous insufficiency, varicose veins, soft tissue edema, and lymphedema.²⁵ Additionally, pruritus, lichenification, and edema often are not seen in most PPD variants, although stasis dermatitis and PPD may occur in tandem. Conservative treatment involves elevation of the extremities, compression, and topical steroids for symptomatic relief.

Cellulitis—The key characteristics of cellulitis are redness, swelling, warmth, tenderness, fever, and leukocytosis. A history of trauma, such as a prior break in the skin, and pain in the affected area suggest cellulitis. Several skin conditions present similarly to cellulitis, including PPD, and thus approximately 30% of cases are



FIGURE 3. Palpable purpura of the lower extremities with nonblanching, dusky, erythematous papules in a patient with leukocytoclastic vasculitis.



FIGURE 4. Stasis dermatitis with hyperpigmentation, induration, and edema of the legs.

misdiagnosed.²⁶ Cellulitis rarely presents in a bilateral or diffusely scattered pattern as seen in PPDs. Rather, it is unilateral with smooth indistinct borders. Variables suggestive of cellulitis include immunosuppression, rapid progression, and previous occurrences. Hyperpigmented plaques or thickening of the skin are more indicative of a chronic process such as stasis dermatitis or lipodermatosclerosis rather than acute cellulitis. Purpura is not a typical finding in most cases of soft tissue cellulitis. Treatment may be case specific depending on severity, presence or absence of sepsis, findings on blood cultures, or other pathologic evaluation. Antibiotics are directed to the causative organism, typically *Streptococcus* and *Staphylococcus* species, although coverage against various gram-negative organisms may be indicated.²⁷

Caution With Tele dermatology

COVID-19 has established the value of telemedicine in providing access to health care services for at-risk or underserved individuals. The PPDs are benign, often asymptomatic, and potentially identifiable with tele dermatology alone; however, they also can easily be mistaken for COVID-19-related eruptions, vasculitis, other types of purpura, stasis dermatitis, or other complications of lower extremity stasis and lymphedema, especially in an aging population. If tissue biopsy is required, as in the workup of vasculitis, the efficacy of telemedicine becomes more questionable. It is important to delineate the potentially confusing PPDs from other potentially dangerous or life-threatening inflammatory dermatoses.²⁸

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