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**Case report**

**Granulomatous pigmented purpuric dermatosis: an unusual variant associated with hyperlipidemia**

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**Abstract**

Granulomatous pigmented purpuric dermatosis (PPD) is a rare subtype of PPD that is typically seen in women of Far East Asian descent on the distal lower extremities and feet. Granulomatous PPD is a benign condition that does not typically require treatment and tends to follow a pattern of exacerbations and remissions. Hyperlipidemia has been observed in over half of the eighteen cases reported in the literature. We report an unusual presentation of granulomatous pigmented purpuric dermatosis seen in a 71 year-old woman with hyperlipidemia.

**Introduction**

The pigmented purpuric dermatoses are a group of eruptive dermatoses that are often characterized by brown to orange petechial macules or plaques on the lower extremities. These disorders are benign and often follow a waxing and waning course with no initial cause identified. Granulomatous PPD is a rare subtype of PPD that has been reported in the literature, occurring frequently on the lower legs of women of East Asian descent. Recently it has been observed that many of these patients have abnormal lipid studies, with hyperlipidemia being present in over half of the reported cases in the literature. We report an additional patient with granulomatous PPD, hyperlipidemia, and hypertension.

**Case synopsis**

A 71 year-old Caucasian woman presented to our clinic with a 3-month history of “bruises” on the right anterior thigh. The areas on the right thigh were asymptomatic and she noted no preceding trauma to the area. She was not on any anticoagulation besides 81-mg aspirin daily and did not have any history of injections to the area. The patient had a history of hyperlipidemia, for which she was taking atorvastatin. She had no personal or family history of clotting disorders. She was unsure of exactly how the lesions began, but had noticed the areas of bruising growing over the initial few months; the areas stabilized in the month prior to the appointment.

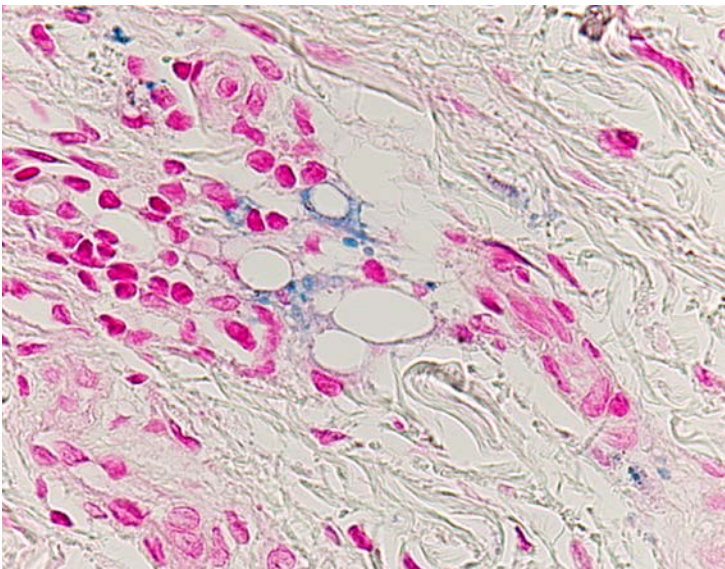
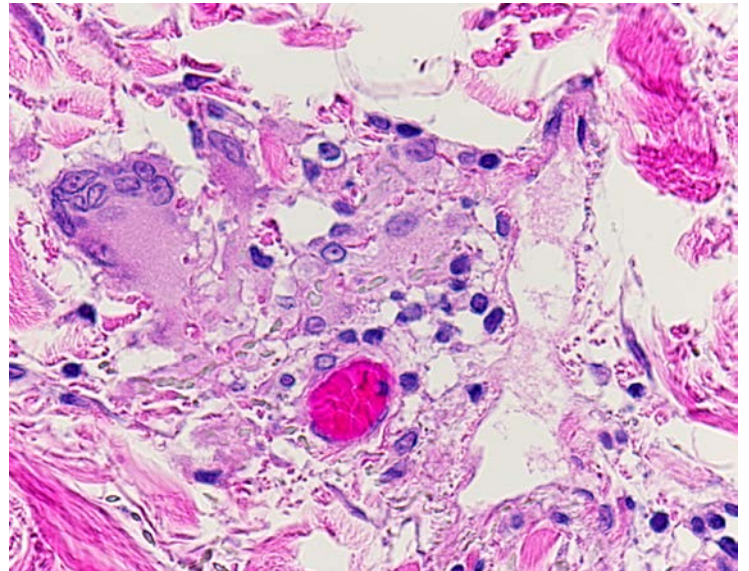
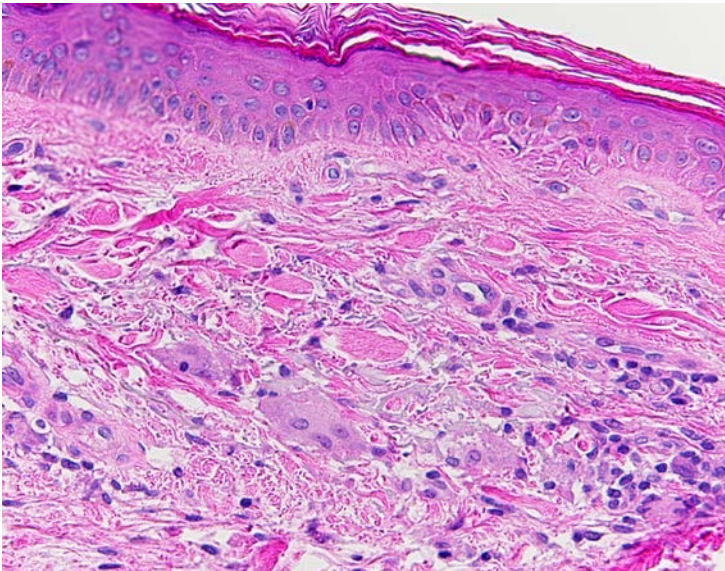
Physical examination revealed several 1-7cm patches of non-blanching, well-circumscribed purpura clustered on the right anterior thigh (Figure 1). Dermoscopy showed scattered red dotted vessels overlying a diffuse brawny red background pigmentation. A 4-mm punch biopsy was obtained from the edge of the largest lesion. Clinical differential diagnosis included small vessel vasculitis, actinic purpura, and trauma induced purpura.

**Figure 1** Purpuric well-circumscribed patches clustered on the right anterior thigh.



Pertinent laboratory findings include several prior readings of triglycerides above 300 mg/dL (normal = <150mg/dL) and a history of an increased low-density lipoprotein level at 194 mg/dL (normal = <100mg/dL) on atorvastatin.

The biopsy demonstrated a mixed granulomatous and lymphocytic perivascular dermatitis. Granulomas were comprised of epithelioid histiocytes and multinucleated giant cells (Figures 2a, 2b). Perls histochemistry highlighted perivascular siderophages (Figure 3).



**Figure 2a** Superficial lympho-histiocytic inflammation with poorly-formed granulomas comprised of epithelioid histiocytes and multinucleated giant cells (haematoxylin and eosin; original magnification x 200).

**Figure 2b** Perivascular lympho-histiocytic inflammation with multinucleated giant cells (haematoxylin and eosin; original magnification x 400).

**Figure 3** Siderophages with blue granules highlighted by PERLS (Prussian Blue histochemistry; original magnification x 400).

Our final diagnosis based on histopathology and clinical presentation was granulomatous pigmented purpuric dermatosis.

**Table 1** Clinical features of patients with reported PPD

Age	Sex	Race	Location	Morphology	Lipid Studies	Associated Diseases	Reference
42	F	Caucasian	Lower legs, ankles	Cluster of purpuric papules, petechiae	Hyperlipidemia	Ulcerative colitis	Kerns et al <sup>5</sup>
48	F	Asian	Lower legs, dorsum of feet	Multiple erythematous to purpuric papules	Hyperlipidemia	None	Lee et al <sup>6</sup>
49	F	Asian	Legs, knees, dorsum of feet	Brown macules, patches, papules	Normal	Hypertension	Wang et al <sup>7</sup>
53	F	Caucasian	Bilateral feet	Multiple purpuric papules	Normal	Seizure disorder	MacQuarrie et al <sup>9</sup>
53	F	Japanese	Dorsum of both feet	Hemorrhagic papules and brown pigmentation	Normal	Hepatitis C	Saito et al <sup>1</sup>
54	F	Asian	Legs, dorsum of feet	Purpuric lesions, lower legs, and dorsum of feet	Hyperlipidemia	Hypertension	Wang et al <sup>7</sup>
56	F	Caucasian	Lower back	Solitary brown patch	Normal	None	MacKenzie et al <sup>2</sup>
60	F	Caucasian	Lower back, legs, wrists, forearms	Cayenne pepper-like spreading rash	Hyperlipidemia	Hypothyroidism	Kaplan et al <sup>8</sup>
61	F	Japanese	Dorsum of both feet	Hemorrhagic papules and brown pigmentation	Normal	Not available	Saito et al <sup>1</sup>
66	F	Asian	Medial aspect of ankles	Purpuric macules and patches	Hyperlipidemia	None	Wang et al <sup>7</sup>
67	F	Asian	Dorsum of Both feet	Erythematous to brown macules	Hyperlipidemia	COPD	Wong et al <sup>3</sup>
68	F	Japanese	Lower legs	Multiple pigmented macules	Normal	Sjogren syndrome	Wakusawa et al <sup>10</sup>
71	F	Taiwanese	Legs, feet	Brown to purpuric papules	Hyperlipidemia	Hypertension, diabetes mellitus	Lin et al <sup>4</sup>
<b>71</b>	<b>F</b>	<b>Caucasian</b>	<b>Right thigh</b>	<b>Multiple purpuric papules and macules</b>	<b>Hyperlipidemia</b>	<b>Hypertension, hypothyroidism, seizure disorder</b>	<b>Current case</b>
22	M	Taiwanese	Legs	Purpuric papules	Normal	Hypertension	Lin et al <sup>4</sup>
37	M	Taiwanese	Wrist	Brown macules and papules	Normal	Not available	Lin et al <sup>4</sup>
47	M	Taiwanese	Legs, feet, dorsum of hands	Erythematous, brown papules, and plaques	Hyperlipidemia	Hypertension, hepatitis C, hyperuricemia	Lin et al <sup>4</sup>
57	M	Asian	Dorsum of both feet	Brown papules and plaques	Hyperlipidemia	Hypertension	Wong et al <sup>3</sup>
65	M	Caucasian	Lower legs	Multiple purpuric to brown papules	Hyperlipidemia	Hypertension	Tato et al <sup>11</sup>

PPD, pigmented purpuric dermatosis; COPD, chronic obstructive pulmonary disease

## Discussion

The pigmented purpuric dermatoses are a group of conditions that present commonly on the lower extremities with brown/orange petechial macules or plaques. Histological features of the various PPDs include erythrocyte extravasation and perivascular infiltration with T-lymphocytes. Hemosiderin deposits in macrophages are also seen, giving the lesions their characteristic color.

Pigmented purpuric dermatoses have historically been divided into 5 clinical entities; Schamberg's disease, Purpura annularis telangiectodes (Majocchi's disease), Gougerot-Blum purpura, Lichen aureus, and Eczematid-like purpura of Doucas and Kapetanakis. There are also rare additional subtypes of pigmented purpuric dermatosis that have been recognized, such as itching purpura, unilateral linear capillaritis, and granulomatous pigmented purpura. In this case report, we present an unusual patient who has granulomatous PPD.

The granulomatous subtype of PPD is a rare variant that was first described in 1996 in two Japanese women with pruritic lesions on the feet bilaterally [1]. Granulomatous PPD presents most commonly with multiple erythematous to brown purpuric macules or papules on the dorsum of the feet and the lower legs. To our knowledge, a total of 18 cases of granulomatous PPD have been reported in the literature and although most have involved the distal lower extremities in patients of Far East Asian descent, there has been one case report of a 58-year-old Caucasian woman who presented with a solitary lesion located on the lower back [2]. Only 5 of the 18 documented cases have presented in Caucasian individuals prior to this report [1-11].

Dermoscopy is being increasingly used in evaluation of various non-pigmented skin disorders and dermoscopic findings documented in cases of granulomatous PPD include irregular, round to oval red dots, globules, and patches in a background red

brown or coppery diffuse and homogenous pigmentation [2,12]. Histopathology has shown a superficial lichenoid granulomatous dermatitis with palisading lymphocytes and an abundance of extravasated erythrocytes. A true vasculitis is absent in granulomatous PPD, differentiating it from a variety of other conditions in which a granulomatous vasculitis is present [5].

The most commonly associated systemic disease seen in patients with granulomatous PPD appears to be hyperlipidemia, which has been reported in 10 of the 18 cases in the literature [1-11]. Although the association between lipid levels and granulomatous PPD has not been determined, some authors suggest that these patients should all have lipid profiles drawn to assess cardiovascular risk [11]. Other studies have shown an association between other granulomatous diseases such as granuloma annulare and dyslipidemia, with the granulomatous reaction thought to be associated with the inflammation related to the dyslipidemia [13]. Previous studies have also suggested the possibility of an association between an individual's race and a hyperlipidemia-related granulomatous reaction, but more recent studies suggest that this may not be the case [3,7,11].

## Conclusion

Our case report adds evidence to the observation that granulomatous pigmented purpuric dermatosis tends to occur in patients with hyperlipidemia. It may be useful to evaluate lipid levels in patients who have been diagnosed with this rare subtype of pigmented purpuric dermatosis. Granulomatous pigmented purpuric dermatosis should also be considered as a possible diagnosis for any female patient who has hyperlipidemia and brown/orange macules or plaques on the lower extremities.

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