History
An 86-year-old man presented with a six-year history of an asymptomatic eruption over the bilateral shins extending up both thighs. It began as a 15 cm patch on the right inner thigh that spread rapidly over one year to involve the majority of his lower extremities.

Examination
Physical examination revealed scattered 1-2 mm brown macules coalescing into patches, extending from bilateral ankles to thighs. There was no scale or induration with palpation and no associated lower extremity swelling.

Laboratory
All laboratory values were within normal limits, including CBC, CMP, UA and lipid profile.

Histopathology
Punch biopsy revealed a superficial to mid dermal perivascular lymphocyte-predominant infiltrate with associated siderophages and a focal granulomatous infiltrate comprised of histiocyes. PAS, AFB and Fite stains were negative for microorganisms. No eosinophils or leukocytoclasis was seen.

Course and Therapy
The patient showed no improvement with topical steroids.

Discussion
Granulomatous pigmented purpuric dermatosis is a rare histological variant of PPD which most commonly affects individuals from Far East Asia and presents on the distal lower extremities. Several other variants of PPD are recognized and include Schamberg’s disease, purura annularis telangiectaticum (of Majocchi), pigmented purpuric lichenoid dermatitis of Gougerot and Blum, eczematoid-like purpura of Doucas and Kapetanakis, itching purpura of Lowenthal, lichen purpuricus, lichen aureus, transitory pigmented purpuric dermatosis and linear pigmented purpuric dermatosis.

Granulomatous PPD has a total of 18 cases documented in the literature, 13 Asian and five Caucasian. It has a mean age of onset of 51 years and a female predilection. Currently the etiology is unknown; however, 10 of the reported cases have been associated with hyperlipidemia. This has led to the speculation that the two may be related. There are single case reports of associations with other systemic derangements such as hepatitis C, Sjögren syndrome, hypertension, seizure disorder, ulcerative colitis, diabetes mellitus, and chronic obstructive pulmonary disease.

Clinically, granulomatous PPD presents with asymptomatic petechiae and bronze discoloration. The clinical presentation can vary from a solitary lesion, a localized eruption typically on the lower extremities, or rarely in a widespread eruption. This variant is characterized histopathologically by ill-defined, non-necrotizing granulomas admixed with a lymphocytic infiltrate. Erythrocye extravasation and hemosiderin in the absence of vasculitis is often seen.

Granulomatous PPD, particularly when arising in the context of idiopathic inflammatory bowel disease, may be confused with cutaneous Crohn disease. PPD as a group has a propensity to simulate mycosis fungoides (MF), but there have been no reported cases of granulomatous PPD progressing into MF. However, this possibility should always be considered and clinical follow-up is advised in cases of diagnostic uncertainty. The granulomatous variant of PPD may be under-recognized, particularly when the granulomatous component is subtle. This variant is an important entity for pathologists to be aware of and consider in dermal granulomatous infiltrates showing signs of vascular injury.

Treatment with oral and topical steroids has been unsuccessful. Due to the suspected hyperlipidemia association, acquiring a lipid profile is warranted. There is no increased risk of mortality and the prognosis is excellent.

Conclusion
Granulomatous pigmented purpuric dermatosis is a rare histological variant of PPD which should be considered in patients presenting with asymptomatic petechiae and bronze discoloration of the lower extremities, especially individuals from Far East Asia. An association with hyperlipidemia is suspected and a lipid profile is suggested. Pathologists should consider this variant in granulomatous infiltrates showing signs of vascular injury. Effective treatment is currently unknown, but prognosis is excellent.

References