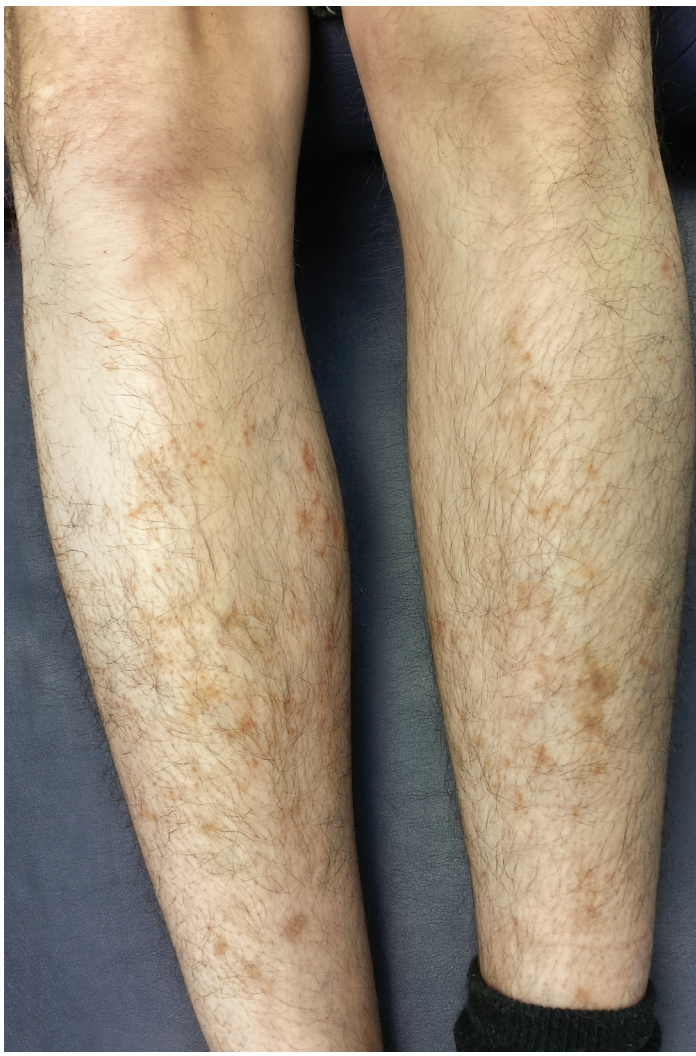
As Problem Spreads, Man Seeks Help



For several months, a 30-year-old man has had an asymptomatic rash on his legs. The lesions first appeared on his lower legs and ankles; over the subsequent months, they have spread upward. Now, the rash reaches to just below his knees.

During this time, he has had two bouts of strep throat, both adequately treated. He denies any other skin problems and has no relevant family history. The patient denies alcohol or drug abuse and is not taking any prescription medications.

Prior to referral to dermatology, he was seen in two urgent care clinics; at one, he received a diagnosis of fungal infection and at the other, of "vitamin deficiency." He was given a month-long course of terbinafine (250 mg/d) that produced no change in his rash. He achieved the same (non) result from an increased intake of vitamins.

Examination reveals annular reddish brown macules, measuring 1 to 3 cm, sparsely distributed from the knees to just above the ankles on both legs. The lesions are a bit more densely arrayed on the anterior legs. There is no palpable component to any of them and no discernable surface scale. Digital pressure fails to blanch the lesions.

The hairs and follicles on the patient's legs appear normal. There are no notable skin changes elsewhere, and the patient is alert, oriented, and in no distress.

Given the facts of the case as presented, the most likely diagnosis is

a) Scurvy
b) Schamberg disease
c) Cutaneous T-cell lymphoma
d) Thrombocytopenia

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ANSWER

The correct answer is Schamberg disease (choice "b"), a benign form of capillaritis; see Discussion for more information.

Scurvy patients can present with ecchymosis (among other findings that were missing in this case). But scurvy (choice "a") is rare, and by the time the disease is evident, the patient is typically quite ill.

Cutaneous T-cell lymphoma (choice "c") can manifest as purpuric annular lesions. However, these would be unlikely to take the distributive pattern seen in this case, and they usually have an atrophic surface.

Thrombocytopenia (choice "d") and other coagulopathies, although rightly considered, would probably manifest in other ways as well (ie, not just cutaneously).

DISCUSSION

Schamberg disease is typical of a whole class of conditions in which red blood cells (RBCs) are extravasated from slightly damaged capillaries. This results in nonblanchable purpura and sub-

sequent hemosiderin staining caused by phagocytosis of the RBCs by macrophages. Clinically, this family of diseases present as cayenne pepper–colored macules, most of which are annular in configuration.

Schamberg is, by far, the most common of these conditions. This presentation was typical: manifestation on the knees and ankles followed by upward spread (hence the condition's other name, *progressive pigmentary purpura*). Usually resolving on their own within months, these lesions are almost always asymptomatic—but nonetheless alarming to the patient.

Other equally benign, self-limited forms of capillaritis include those in which lesions are pruritic (eg, purpura of Doucas and Kapetanakis) or lichenoid (purpura of Gougerot-Blum). Another example is lichen aureus, in which only one or two lesions, more gold than brown, appear on the legs of younger patients.

There are many theories as to these conditions' cause, the most common of which is increased intravascular pressure secondary to dependence. However, if this were so, we'd likely see a great deal more cases, since many patients have problems related to venous insufficiency.

In some cases, skin biopsy (usually 4-mm punch) is necessary to rule out more serious diseases, such as an early form of cutaneous T-cell lymphoma. When a coagulopathy is suspected, blood work is necessary to confirm or rule out the diagnosis. In this case, there was no reason to suspect coagulopathy (or scurvy), since no other signs were seen.

This patient was educated about his diagnosis and provided Web-based resources he could consult. Various treatments—topical steroids, increased vitamin C intake, and increased exposure to UV light—have been tried but with disappointing results. **CR**