



Chronic lesions on legs

The rash was asymptomatic and had developed over the past few years. The patient wasn't taking any prescriptions and hadn't changed detergents.

A 62-YEAR-OLD MAN came into our facility for a skin exam at the urging of his wife. They were both concerned about a rash on his legs that was asymptomatic and had gradually developed over the past few years.

The patient, who worked as a laborer, was not taking any prescription medications, vitamins, or herbal products. He didn't smoke, drink alcohol, or use recreational drugs and denied having any fevers, chills, arthralgias, or night sweats. He indicated that his father had a basal cell carcinoma.

On physical exam, we noted some seborrheic keratoses and mild acne rosacea in addition to the rash on his legs. The rash itself consisted of nonpalpable, nonblanchable macules on the lower shins, ankles, and dorsal feet with sparing of the soles (FIGURE).

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE
Nonpalpable rash



The rash on this 62-year-old man's legs developed over several years. The nonblanchable macules on the lower shins, ankles, and dorsal feet spared the soles.

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PHOTO ROUNDS FRIDAY

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➤ Although these lesions have a vasculitic appearance, there is no hematologic or other internal disease association.

**Diagnosis:
Schamberg’s disease**

Schamberg’s disease is a form of progressive pigmented purpuric dermatosis. This type of nonpalpable purpura consists of reddish-brown macules that are usually located on the lower extremities and ankles, and spare the soles of the feet. These petechiae appear due to capillary leakage and breakdown of blood near the skin surface, leaving behind hemosiderin deposits.

Clinically, the lesions are asymptomatic and may persist for months or years. They are characteristically referred to as punctate “cayenne pepper” spots, and can vary in size and shape. Occasionally, mild erythema and scaling can cause slight itching.

Although these lesions have a vasculitic appearance, there is no hematologic or other internal disease association. Schamberg’s purpura may resemble stasis dermatitis, because both processes include inflammation, dilation of capillaries, extravasation of erythrocytes, and hemosiderin deposits. However, lesions due to venous insufficiency extend deep into the dermis and have more pronounced epidermal changes and dermal fibrosis.

■ **An unknown cause.** Although no one knows what causes Schamberg’s disease, a cellular immune reaction may be at work. Early endothelial expression of certain intracellular adhesion receptors has shown a common pericapillary infiltrate pattern in cases of Schamberg’s disease.¹ Recent viral infection and allergic reactions to medications, such as aspirin, have also been associated with Schamberg’s disease.²

One of several pigmented purpuric dermatoses

Schamberg’s disease fits under the inclusive term of pigmented purpuric dermatoses (PPD), as a macular purpuric reddish-brown dermatosis. Other subtypes of PPD can be distinguished from Schamberg’s disease clinically. These subtypes include lichen aureus, which appears as golden patches; Majocchi’s purpura, which forms an annular pattern with telangiectasias; Gougerot-Blum purpura, which is associated with lichenoid dermatitis; and Doucas and Kapentanakis purpura, which is an eczematous variant. Other forms of purpura can be distinguished from Schamberg’s disease histologically (TABLE).³

■ **Platelet abnormalities** can also produce red, flat, nonblanchable petechiae, and may occur in patients who have taken certain prescribed drugs, over-the-counter medications, or herbal remedies; those who have received recent vaccinations; and in those who have (or have had) severe viral infections, beta-hemolytic streptococcal infections, leukemia, lupus erythematosus, or idiopathic thrombocytopenic purpura.

■ **Scurvy** can also cause perifollicular petechiae that are distributed primarily on the lower extremities and are symmetrical. Gums may be swollen or hemorrhagic, and patients will have a history of vitamin C deficiency, myalgia, and fatigue. Children may have bone tenderness, epistaxis, and hematuria. A diagnosis can be made by looking for low ascorbic acid in the serum. Prompt correction with vitamin C supplementation is also diagnostic.

TABLE
Histologic spectrum of purpura³

Histologic pattern	Diseases
Erythrocyte extravasation without perivascular inflammation or fibrin deposition	Thrombocytopenic purpura, senile purpura, steroid purpura, scurvy
Erythrocyte extravasation with lymphocytic perivascular infiltrate; no fibrin deposition	Lichen aureus; Schamberg’s, Majocchi’s, Gougerot-Blum, and Doucas and Kapentanakis purpuras
Erythrocyte extravasation with vascular damage, neutrophilic infiltrate, and fibrin deposition	Leukocytoclastic vasculitis, mixed cryoglobulinemia

■ **Dysproteinemia** can cause crops of petechiae and occasionally ecchymoses. Lesions appear on the lower extremities and sometimes on the ears and the tip of the nose. Cryoglobulinemia is associated with Raynaud's phenomenon, cold urticaria, dizziness, and epistaxis; it is also associated with hepatitis C infection in >90% of cases.⁴ Macroglobulinemia is associated with dilated vessels and hemorrhage in the optic fundi, mental confusion, anemia, weight loss, hepatosplenomegaly, and lymphadenopathy. Hyperglobulinemic purpura may be associated with arthritis, xerostomia, anemia, and hepatosplenomegaly. These various dysproteinemias can be diagnosed by finding abnormal proteins through serum electrophoresis. Skin biopsy of the petechiae reveals thrombi in dermal vessels, which may be associated with leukocytoclastic vasculitis.⁴

■ **Many viral infections** may result in a petechial rash, including measles, rubella, hepatitis, cytomegalovirus, Coxsackievirus, and respiratory syncytial virus. Petechial rashes associated with eczema or seborrheic dermatitis-like lesions can also be seen in children with Langerhans cell histiocytosis or Wiskott-Aldrich syndrome.

Biopsy confirms diagnosis

Platelet and clotting studies are usually normal with Schamberg's purpura. The definitive diagnosis can be confirmed through skin biopsy, which shows capillaritis of dermal vessels. Perivascular inflammatory infil-

trates with extravasations of blood cells and hemosiderin-laden macrophages are seen on histologic evaluation.

Treat mild itching with corticosteroids

These lesions pose mostly a cosmetic problem, but mild itching and scaling occasionally occur. These symptoms can be treated with topical corticosteroid therapy.

Treatment with an 8-week course of pentoxifylline (Trental)—400 mg daily—has shown resolution of lesions in some patients, but no benefit in others.^{5,6} In some patient's, psoralen plus ultraviolet light therapy has been shown to provide modest improvement.⁷ Treatment with aminaphtone 75 mg bid for 4 weeks has recently shown improvements in patients with longstanding Schamberg's purpura.⁸ A review of medications may point to a possible etiology for some patients.⁹

Although lesions can persist for years, the lesions may eventually clear and patients should be assured that there is no definitive underlying systemic disease associated with Schamberg's disease.¹⁰

Educating our patient

Our patient required no treatment, because his rash was asymptomatic. We told him that the pigmentation can last for years, but could be covered with cosmetics. We also reassured him that no underlying systemic disease had caused his rash.

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Treatment with aminaphtone may help patients with longstanding Schamberg's purpura.

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