

THE CLINICAL PICTURE

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The Clinical Picture

Palpable purpura

A HEALTHY 47-YEAR-OLD WOMAN presents with a 3-day history of widespread asymptomatic lesions in the extremities, fever, arthralgias, and mild abdominal pain. A physical examination reveals a symmetric rash in the lower legs with erythematous purple papules. These papules do not blanch when pressure is applied and so are clinically compatible with palpable purpura (FIGURE 1). Skin biopsy shows leukocytoclastic vasculitis with immunoglobulin A (IgA) deposits in the vascular wall.

Q: Which is the most likely diagnosis?

- Idiopathic thrombocytopenic purpura
- Vitamin C deficiency (scurvy)
- Kaposi sarcoma not related to human immunodeficiency virus (HIV) infection
- Henoch-Schönlein purpura
- Polyarteritis nodosa

A: The correct answer is Henoch-Schönlein purpura.

Idiopathic thrombocytopenic purpura is an autoimmune disease caused by specific antibodies against platelet-membrane glycoproteins. It is characterized by thrombocytopenia not explainable by contact with toxic substances or by other causes. Along with nonpalpable purpura, other common signs are epistaxis, gingival bleeding, menorrhagia, and retinal hemorrhage.

Scurvy is an uncommon deficiency of ascorbic acid (vitamin C). The elderly and alcoholics are at higher risk, as they do not take in enough vitamin C in the diet. Patients usually show perifollicular hemorrhages of the skin and mucous membranes, typically petechial hemorrhage or ecchymosis of the gums around the upper incisors. Other cutaneous signs are follicular hyperkeratosis on the

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FIGURE 1. Erythematous purple papules on the lower legs.

forearms, small corkscrew hairs, and sicca syndrome, which is more common in adults.

Non-HIV Kaposi sarcoma usually affects elderly patients, with pink, red, or brown papules or nodules on the legs and, less commonly, on the head and neck. Histopathologic examination shows newly formed irregular blood vessels with an inflammatory infiltrate of plasma cells and lymphocytes; immunohistochemical human herpes virus staining is usually positive.

As Henoch-Schönlein purpura is less common in adults, biopsy plays an important diagnostic role

Polyarteritis nodosa is a systemic vasculitis that affects medium or small arteries with necrotizing inflammation; renal glomeruli and arterioles, capillaries, and venules are unaffected. Skin manifestations include palpable purpura, livedo reticularis, ulcers, and distal gangrene. The condition also usually affects the kidneys, the heart, and the musculoskeletal and nervous systems.

■ A SYSTEMIC VASCULITIS

Henoch-Schönlein purpura is a systemic vasculitis affecting the skin, gastrointestinal tract, kidneys, and joints. Palpable purpura and joint pain are the most common and consistent presenting symptoms. The kidneys are affected in about one-third of children and in 60% of adults, and this is the major factor determining the long-term outcome.¹

In our patient, laboratory testing that included a complete blood cell count, biochemical testing (including IgA levels), urinary sediment, and coagulation studies showed no abnormalities except elevations of the erythrocyte sedimentation rate and the concentration of C-reactive protein (an acute-phase reactant). These can be normal in some patients. Renal involvement was also not present.

■ DIAGNOSIS

The diagnosis relies on clinical manifestations. Because Henoch-Schönlein purpura is less common in adults, biopsy plays a more important role in establishing the diagnosis in this age group, and it does this by demonstrating leukocytoclastic vasculitis with a predominance of IgA deposition under immunofluorescence. Recent studies in children showed that an elevated IgA concentration along with reduced IgM levels was associated with a higher rate of severe complications.² However, depending on the age of the biopsied lesion, IgA may not be detected.

■ TREATMENT DIRECTED AT SYMPTOMS

Our patient received oral corticosteroids 0.5 mg/kg per day for 20 days, and the lesions resolved by 4 weeks.

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Management of Henoch-Schönlein purpura is mainly directed at the symptoms, with oral hydration and non-steroidal anti-inflammatory drugs. For severe cases, a short course of corticosteroids (0.5–1 mg/kg) may be used. Although no controlled clinical trial has proven that Henoch-Schönlein purpura responds to corticosteroids, colchicine, or other drugs, corticosteroids are used most often, especially in patients with renal disease. Patients with severe renal insufficiency, abdominal pain, joint involvement, or bleeding should be hospitalized. Plasmapheresis³ has been used in severe cases.

■ HENOCH-SCHÖNLEIN PURPURA AND MALIGNANCY

During a follow-up evaluation 1 month later, our patient was diagnosed with adenocarcinoma of the breast. This highlights the value of a workup for cancer in adults with cutaneous vasculitis.

Cutaneous vasculitis can represent a paraneoplastic syndrome associated with a malignant tumor. The pathophysiology of this association is unclear, but one proposed mechanism is the exaggerated production of antibodies that react against tumor neoantigens, leading to the formation of immune complexes, or that occasionally recognize endothelial cells because of similarities with tumor antigens. Another theory is that abnormally high levels of inflammatory cytokines are produced by neoplastic cells or in response to decreased immune complex clearance.

Yet another theory is that hyperviscosity of the blood, seen in some cancers, increases the contact time for deposition of immune complexes and causes endothelial damage. Drugs used to treat cancer have also been reported to produce Henoch-Schönlein purpura.⁴

Although hematologic malignancy is three to five times more common than solid tumors in patients with small-vessel vasculitis, the disease has been associated with solid tumors of the liver, skin, colon, and breast in adults over age 40.⁵ An evaluation for neoplasm is therefore reasonable in adults with Henoch-Schönlein purpura, as is an evaluation for tumor recurrence or metastasis if the patient has been previously treated for a malignant tumor. ■

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