

> THE PATIENT

8-year-old boy

> SIGNS & SYMPTOMS

- Palpable purpura on the legs with arthralgia
- Absence of coagulopathy
- Upper respiratory infection

> THE CASE

An 8-year-old boy presented to his family physician (FP) with pharyngitis, nasal drainage, and a dry cough of 3 days' duration. He denied any fever, chills, vomiting, or diarrhea. He had no sick contacts or prior history of streptococcal pharyngitis, but a rapid strep test was positive. No throat culture was performed at this time. The patient was started on amoxicillin 250 mg 3 times daily for 10 days.

On Day 7 of symptoms, the patient presented to the emergency department with elbow and knee pain, as well as mild swelling and purpura of his legs of 3 days' duration. He was normotensive and reported no abdominal pain. A laboratory workup, including a complete blood cell count and differential, prothrombin time, partial thromboplastin time, comprehensive metabolic panel, creatinine kinase test, urinalysis, and chest radiograph, was normal, but his erythrocyte sedimentation rate (ESR) was mildly elevated at 22 mm/h (reference range, 0–20 mm/h). The patient was discharged on acetaminophen 15 mg/kg every 4 hours as needed for pain.

THE DIAGNOSIS

Based on the distinctive palpable purpura on the legs, arthralgia, upper respiratory infection, and lack of thrombocytopenia and coagulopathy, a presumptive diagnosis of Henoch-Schönlein purpura (HSP) was made.

On Day 9 of symptoms, the patient returned to his FP's office because the arthralgia persisted in his ankles, knees, and hips. He had developed lower back pain, but the pharyngitis and upper respiratory symptoms had resolved. On physical examination, he was normotensive with a normal abdominal exam. The patient reported that it hurt to move his wrists, hands, elbows, shoulders, knees, and ankles. He also had mild swelling in his left wrist, hand, and ankle. The paraspinal muscles in the lower thoracic and lumbar back were mildly tender to palpation. A complete metabolic panel and urinalysis were normal. Dermatologic examination revealed discrete purpuric lesions ranging from 1 to 8 mm in diameter on the child's shins, thighs, and buttocks. Urinalysis, blood urea nitrogen, and creatinine kinase were normal. His ESR remained mildly elevated at 24 mm/h. Since there was no evidence of glomerulonephritis, ibuprofen 10 mg/kg every 8 hours as needed was added for pain management.

The child was brought back to his FP on Day 18 for a scheduled follow-up visit. The parents reported that his arthralgia was improved during the day, but by the evening, his knees and ankles hurt so much that they had to carry him to the bathroom. On physical examination, he still had palpable purpura of the legs. There was no swelling, but his joints were still tender to palpation. His parents were reminded to give him ibuprofen after school to control evening pain. Over the next 2 weeks, the patient showed gradual improvement,

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FIGURE

The distinctive nonthrombocytopenic purpuric rash of Henoch-Schönlein purpura



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and by Day 33 the rash and all of the associated symptoms had resolved.

DISCUSSION

Clinical presentation. HSP is an IgA immune complex vasculitis in which abnormal glycosylation of IgA creates large immune complexes that are deposited in the walls of the skin capillaries and arterioles. The primary clinical finding in HSP is a distinctive nonthrombocytopenic purpuric rash that is not associated with coagulopathy and is characterized by reddish purple macules that progress to palpable purpura with petechiae (FIGURE). Lesions generally are distributed on the legs or buttocks but also may appear on the torso or arms. Flu-like symptoms, such as fever, runny nose, and cough, are common.

A preceding upper respiratory infection has been found in 37% of patients,¹ and in patients with renal complications, 20% to 50% have been found to have a group A *Streptococcus* infection.² Other associations include food allergies, cold exposure, insect bites, and drug allergies.

HSP vasculitis causes abdominal pain in 50% to 75% of patients due to proximal small-bowel submucosal hemorrhage and bowel wall edema.³ In children with HSP, 20% to 55% have been shown to develop renal disease,⁴ which can range in severity from microscopic

hematuria to nephrotic syndrome.³ To ensure prompt treatment of renal manifestations, renal function should be monitored regularly via blood pressure and urinalysis during the course of HSP and after resolution. Renal disease associated with HSP can be acute or chronic.

■ **This case was different** because our patient did not exhibit all elements of the classic tetrad of HSP, which includes the characteristic rash, abdominal pain, renal involvement, and arthralgia.

■ **Incidence.** HSP is more common in children than adults, with average annual incidence rates of 20/100,000 and 70/100,000 in children in the United States and Asia, respectively.⁵ While 90% of HSP cases occur in children < 10 years, the peak incidence is at 6 years of age.⁶ Complications from HSP are more common in adults than in children.⁷ Caucasian and Asian populations have a 3- to 4-times higher prevalence of HSP than black populations. The male-to-female ratio is 2 to 1.⁶

■ **The diagnosis of HSP is usually made clinically,** based on the distinctive rash, which typically is symmetrical, involving the buttocks, lower legs, elbows, and/or knees. HSP also can be confirmed via skin biopsy and/or direct immunofluorescence, which can identify the presence of IgA in the vessel walls.

The presence of 3 or more of the following criteria also suggests HSP: palpable purpura, bowel angina, gastrointestinal (GI) bleeding, hematuria, ≤ 20 years of age at onset, and no medications prior to presentation of symptoms (87% of cases correctly classified). Fewer than 3 of these factors favor hypersensitivity vasculitis (74% of cases correctly classified).⁸

■ **The differential diagnosis** for HSP includes polyarteritis nodosa, a vasculitis with a different characteristic rash; acute abdomen, distinguished by the absence of purpura or arthralgia; meningococcemia, in which fever and meningeal signs may occur; hypersensitivity vasculitis, which arises due to prior exposure to medications or food allergens; and thrombocytopenic purpura, which is characterized by low platelet count.⁹

CONTINUED

Treatment focuses on pain management

In the absence of renal disease, HSP commonly is treated with naproxen for pain management (dosage for children < 2 years of age: 5-7 mg/kg orally every 8-12 hours; dosage for children ≥ 2 years of age, adolescents, and adults: 10-20 mg/kg/d divided into 2 doses; maximum adolescent and adult dose is 1500 mg/d for 3 days followed by a maximum of 1000 mg/d thereafter).

For patients of all ages with severe pain and those with GI effects limiting oral intake of medication, use oral prednisone (1-2 mg/kg/d [maximum dose, 60-80 mg/d]) or intravenous methylprednisolone (0.8-1.6 mg/kg/d [maximum dose, 64 mg/d]). Glucocorticoids may then be tapered slowly over 4 to 8 weeks to avoid rebound since they help with inflammation but do not shorten the course of disease. Steroids can ease GI and joint symptoms in HSP but will not improve the rash.

THE TAKEAWAY

The classic tetrad of HSP includes the characteristic rash, abdominal pain, renal involvement, and arthralgia. Diagnosis usually is made clinically, but skin biopsy and direct

immunofluorescence can confirm small vessel vasculitis with IgA deposits. More severe manifestations of HSP such as renal disease, hemorrhage, severe anemia, signs of intestinal obstruction, or peritonitis require rapid subspecialty referral.

JFP

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