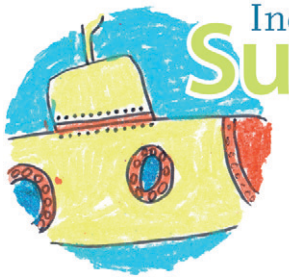

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1 Abdominal Pain and Seizure in a 4-year-old Boy

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PRESENTATION

A 4-year-old boy presents with a 2-week history of intermittent abdominal pain and vomiting. He also has poor oral intake and decreased urine output. Yesterday he developed left ankle pain and redness and refused to walk. He also had a staring spell that developed into a brief tonic seizure this morning.

Physical examination reveals temperature of 96.8°F (35.9°C), heart rate of 165 beats/min, respiratory rate of 16 breaths/min, and blood pressure of 145/107 mm Hg. His weight is 17 kg (45th percentile) and height is 104 cm (34th percentile). He has dry oral mucosae. He has no rashes but a warm, erythematous, swollen area over the superior aspect of the left ankle. He has mild tenderness all over the abdomen, although it is soft and without ascites, masses, or organomegaly. He has 2+ pitting edema on both legs. His lungs are clear to auscultation.

Laboratory tests upon admission demonstrate:

- Sodium 123 mEq/L (123 mmol/L)
- Potassium 4 mEq/L (4 mmol/L)
- Chloride 89 mEq/L (89 mmol/L)
- Carbon dioxide 28 mEq/L (28 mmol/L)
- Blood urea nitrogen 12 mg/dL (4.3 mmol/L)
- Creatinine 0.4 mg/dL (35.4 μmol/L)
- Aspartate aminotransferase 29 U/L (0.5 μkat/L)
- Alanine aminotransferase 15 U/L (0.3 μkat/L)
- Alkaline phosphatase 106 U/L (1.8 μkat/L)
- Total bilirubin 0.5 mg/dL (8.6 μmol/L)
- Albumin 3.3 g/dL (33 g/L)
- White blood cell count 32,000/μL (32 $\times$ 10^9/μL) with 71% neutrophils, 16% lymphocytes, and 10% monocytes
- Hemoglobin 16.4 g/dL (164 g/L)
- Hematocrit 46% (0.46)
- Platelets 688 $\times$ 10^3/μL (688 $\times$ 10^9/L)

Urinalysis reveals a specific gravity of 1.030, large ketones, large blood, 35 to 50 red blood cells per high-power field, and 100 mg/dL of protein.

Soon after admission, the boy develops melena and a raised petechial rash over his upper and lower extremities. Further evaluation reveals the diagnosis.

EDITORS NOTE

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AUTHOR DISCLOSURE Drs Sanders and Huber have disclosed no financial relationships relevant to this article. This commentary does not contain a discussion of an unapproved/investigative use of a commercial product/device.

DISCUSSION

The differential diagnoses included an evolving nephrotic syndrome and glomerulonephritis. After hospitalization, a urine protein-creatinine ratio of 7.3 was reported. A renal biopsy revealed mesangial and segmental endocapillary hypercellularity with positive immunoglobulin (Ig)A, C3, and kappa and lambda light chains (Fig 1), consistent with proliferative and necrotizing IgA nephropathy. The clinical manifestations and biopsy results led to the diagnosis of Henoch-Schönlein purpura (HSP).

The boy had a complicated hospital course. Due to the melena and severe abdominal pain, abdominal ultrasonography was obtained, which was concerning for an intestinal wall hematoma leading to bowel obstruction. He was taken to the operating room and found to have significant hemorrhagic and edematous bowel with multiple intramural hematomas but no bowel ischemia or necrosis (Fig 2). Following surgery, he developed pulmonary hemorrhage that required high-frequency oscillatory ventilation. Subsequently he developed significant intermittent bradycardia with a heart rate in the thirties and significant sinus pauses on telemetry. This was consistent with conduction delay, which improved with immunosuppressive therapy. Echocardiography showed normal coronary anatomy and cardiac function. He had no further seizures, although electroencephalography (EEG) revealed focal, left-sided abnormality, but findings on brain magnetic resonance imaging (MRI) were normal. Focal abnormalities on his EEG suggested that his seizure was likely due to central nervous system vasculitis exacerbated by hyponatremia. This was an unusual case of HSP, with the patient demonstrating severe, multiorgan involvement related to systemic vasculitis.

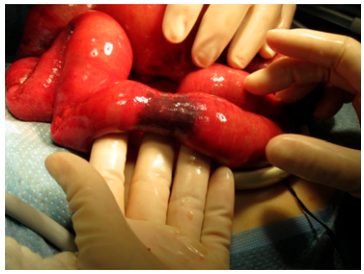


Figure 2. Surgery revealed a large intramural hematoma in the small intestine.

The Condition

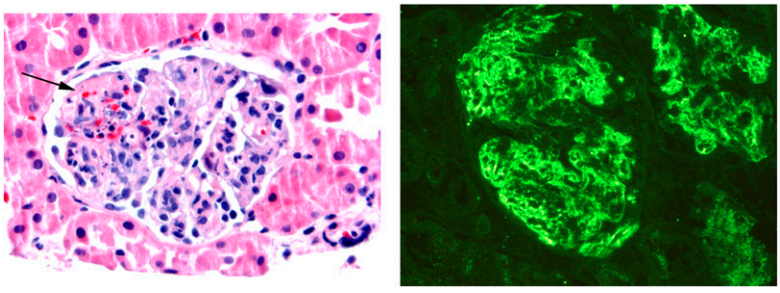
HSP is the most common vasculitis in children and is typically a self-limited disease. The estimated annual incidence is 20 per 100,000 in children younger than age 17 years. HSP most commonly affects children 4 to 6 years of age, with the incidence peaking in the winter and spring. Caucasian males are more commonly affected.

HSP is an IgA-mediated small-vessel vasculitis that commonly presents as lower-extremity purpura, arthritis, abdominal pain, and renal disease (glomerulonephritis). IgA immune complexes consisting of antigens bound to IgA deposit in small vessels. HSP has been postulated to be triggered by infections, vaccinations, or medications, although the exact cause is uncertain. Diagnostic criteria developed by the EULAR/PRINTO/PReS task force in 2010 include petechiae/purpura plus one of the following four criteria: arthritis/arthralgias, abdominal pain, histopathology demonstrating IgA deposition, or renal involvement in the form of nephritis or nephrotic syndrome.

Gastrointestinal involvement is very common in HSP, affecting approximately two thirds of patients. Severe intra-abdominal complications are less common; intussusception is the most frequent. Abdominal pain can be severe enough to warrant exploratory laparotomy, as occurred with this patient.

Neurologic symptoms are less frequent in HSP, occurring in 1% to 8% of children. Symptoms include headache, seizures, cranial nerve deficits, hemiplegia, and encephalopathy. Posterior reversible encephalopathy syndrome is likely due to hypertension. HSP vasculitis is very difficult to diagnose by imaging because it typically involves small vessels; magnetic resonance angiography (MRA), MRI, and single-photon emission computed tomography (SPECT) imaging may be helpful.

Figure 1. Renal biopsy showing endocapillary hypercellularity with deposition of immunoglobulin A.



Pulmonary hemorrhage is very rare in HSP; fewer than 10 cases have been reported in the literature. It typically presents as tachypnea, dyspnea, hemoptysis, cough, and anemia and is associated with high morbidity and mortality rates. Most patients require mechanical ventilation. Bronchoscopy with lung biopsy is recommended to confirm the diagnosis. Biopsy typically shows IgA deposition along alveolar spaces. Aggressive immunosuppressive therapy such as pulse-dose corticosteroids, cyclophosphamide, and plasma exchange are recommended due to the high morbidity and mortality associated with pulmonary hemorrhage.

Cardiac involvement is also very rare in HSP, reported in fewer than 10 cases. This severe expression of HSP can be fatal. Manifestations include conduction abnormalities, myocardial infarction, cardiac dysfunction/dilation, and coronary vasculitis. Autopsy findings in published case reports demonstrated myocardial necrosis with deposition of IgA and C3 in myocardial vessel walls, consistent with HSP.

Management

HSP is typically managed with supportive care. Corticosteroids are used primarily for severe abdominal pain but do not appear to prevent renal dysfunction.

This patient received multiple immunosuppressive therapies due to the severity of his illness and multiorgan involvement. Initially, he was given oral corticosteroids but later transitioned to pulse corticosteroids (six doses) and intravenous immunoglobulin on two occasions due to worsening symptoms. After he developed pulmonary hemorrhage, he was started on cyclophosphamide. When the bradycardia and conduction abnormalities occurred, he received three consecutive daily plasmapheresis treatments.

Lessons for the Clinician

- Henoch-Schönlein purpura is the most common vasculitis in children and is typically a self-limited disease that resolves in 3 to 8 weeks. Approximately one third of patients have recurrences within 4 months.
- Diagnosis involves petechiae/purpura in dependent areas in addition to one of the following: arthritis/arthralgias, abdominal pain, histopathology demonstrating immunoglobulin A deposition, or renal involvement (nephritis or nephrotic syndrome). Diagnosis may not be clear upon presentation
- Treatment is typically supportive and may involve oral corticosteroids for severe abdominal pain. Severe vasculitis requires immunosuppressive therapy such as cyclophosphamide and pulse-dose corticosteroids.

Suggested readings for this article are at http://pedsinreview.aappublication.org/content/37/2/78.