A 10-Year-Old Girl with Gastrointestinal Hemorrhage

Ilaria Possenti, MD; Elena Borali, MD; Paola Longaretti, MD; Lorenzo A. Bassi, MD; Federico Cattaneo, MD; Lucia Bianchi, MD; Stella Boghen, MD; and Antonietta Marchi, MD

A 10-year-old girl presented to our hospital with arthralgia in the lower extremities and abdominal pain. Laboratory tests were normal. Urinalysis was negative for hematuria and proteinuria. Stool for occult blood test was negative. She was given oral ibuprofen for joint pain. On the same day she also presented with fever and sore throat, so she was given oral clavulanic-amoxicillin. After a few days she presented to our hospital with advanced polyarthalgias (affecting her wrists and ankles in particular) and two episodes of vomiting. She was admitted to our unit, at which point she suddenly presented with severe abdominal pain localized in the epigastric and the lower quadrant of her abdomen, leading to hematemesis and melena. On physical examination there were discreet general conditions, the abdomen was soft, and only deep palpation evoked diffuse moderate pain. Cardiac and lung findings were normal. The girl also presented with swollen and painful wrists and ankles, and a few purpuric lesions appeared on her lower extremities and eyelid.

We suspended anti-inflammatory oral therapy and began omeprazole intravenously. We positioned nasogastric and rectal tubes. Laboratory tests revealed normal values of hemoglobin and coagulation parameters, C-reactive protein of 2.1 mg/dL (normal value <.5 mg/dL), and normal renal and liver function. Urinalysis was negative for hematuria and proteinuria. Abdominal ultrasound showed thickening of the duodenal wall (Figure 1). Esophagogastroduodenoscopy showed multiple erosions in the duodenum with hyperemic gastric mucosa, and the biopsies were compatible with leukocytoclastic vasculitis (Figure 2). Steroid therapy with intravenous methylprednisolone at 2 mg/kg was started. The patient showed progressive improvement of her general condition in the first 24 hours, and 2 days later we removed the nasogastric tube and she began a liquid diet and then began to eat lighter foods. The patient was discharged from the hospital, and her laboratory and radiological follow-up results showed complete regression of the lesions after 3 weeks of steroid therapy (Figure 3).

For diagnosis, see page 98

Editor’s note: Each month, this department features a discussion of an unusual diagnosis. A description and images are presented, followed by the diagnosis and an explanation of how the diagnosis was determined. As always, your comments are welcome via email at pedann@Healio.com.
Diagnosis:
Duodenitis in Henoch-Schönlein Purpura

DISCUSSION
Henoch-Schönlein purpura (HSP) is the most common vasculitis of childhood. It is an immune-mediated systemic vasculitis of the small vessels with an estimated incidence in children of 10.5-20.4/100,000 per year, with peak incidence between ages 2 and 6 years.1-3 There is a slight male preponderance (1.2:1), with a lower incidence in black children as compared with white or Asian children.4 The pathogenesis of HSP is not yet clearly understood, although it is known to be an immune complex-mediated disease.1 It can be associated with infections (such as group A beta-hemolytic streptococcus, mycoplasma, adenovirus, parvovirus B19, varicella, and herpes simplex), medications, vaccinations, tumors, alpha-1-antitrypsin deficiency, and familial Mediterranean fever.5-7 Antigen and antibody complexes (mostly immunoglobulin A) form as a result of bacterial and viral infections, vaccinations, drugs, and autoimmune mechanism.8 These antigen antibody complexes deposit in small vessel walls and activate the alternate complement pathway, which leads to neutrophil accumulation, resulting in inflammation and vasculitis without a granulomatous reaction. This can involve multiple systems, including skin, gastrointestinal tract, kidney, and joints, and it can also involve any organ system. Vasculitis causes extravasation of blood and its components into the interstitial spaces, resulting in edema and hemorrhage.9

In 2006, the European League Against Rheumatism and Paediatric Rheumatology European Society published a new classification of vasculitis (Table 1).10 Characteristic symptoms and signs include palpable purpuric rash, abdominal pain, arthralgia, and nephritis.11 Gastrointestinal symptoms and disease occurs in up to 85% of patients and can be of varying intensity. Symptoms include colicky abdominal pain, nausea, vomiting, diarrhea, or bleeding and can occur in 30% or more of patients, although serious complications such as intussusception, perforation, or obstruction are unusual.12,13 Most cases of HSP are self-limited, with good prognosis and 5-year survival rate of 95%.7,14-16 One-third of patients have a relapse, but these are milder and shorter in duration, usually within 4 months of the original onset of symptoms, and involve the same organs.7 The prognosis depends on the age of onset, extent of renal involvement and its course, extent of skin involvement (particularly above the waist line), immunoglobulin imbalance, and neurological involvement. The differential diagnosis of HSP includes conditions such as Crohn’s disease, small vessel vasculitis with granulomatosis, infective endocarditis, immunoglobulin A nephropathy, and hemolytic uremic syndrome. Symptomatic treatment will be sufficient for symptoms such as rash and arthritis. Oral steroids are indicated in patients with severe rash, edema, severe colicky abdominal pain (without nausea, vomiting), and renal, scrotal, or testicular involvement.9

TABLE 1.
European League Against Rheumatism and Paediatric Rheumatology European Society Criteria for Vasculitis

<table>
<thead>
<tr>
<th>Mandatory criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Palpable purpura</td>
</tr>
<tr>
<td>Plus at least one of the following criteria</td>
</tr>
<tr>
<td>• Diffuse abdominal pain</td>
</tr>
<tr>
<td>• Immunoglobulin A deposition in any biopsy</td>
</tr>
<tr>
<td>• Arthritis/arthralgias</td>
</tr>
<tr>
<td>• Renal involvement (hematuria and/or proteinuria)</td>
</tr>
</tbody>
</table>

Adapted from Ozen et al.10

Figure 3. Abdominal ultrasound showing complete regression of lesions after 3 weeks of treatment with steroids.
Second-line therapy includes immunosuppressive drugs, intravenous immunoglobulins, and plasmapheresis. There are some case reports showing that dapsone or colchicine may be useful in cases of chronic HSP.17

Our patient presented with severe gastrointestinal bleeding and hematemesis and melena after oral ibuprofen therapy for arthralgia that was taken without gastric protection. We suspected use of nonsteroidal anti-inflammatory drugs and started intravenous gastroprotection but there was no improvement. The patient continued to have gastrointestinal bleeding and severe abdominal pain. Esophagogastroduodenoscopy showed multiple erosions in the duodenum with hyperemic gastric mucosa. Immunoglobulin A deposition on mucosal vessels with concomitant skin and joint involvement is diagnostic of HSP.11,12

Our patient did not present with renal involvement, and this case of HSP was atypical in that there was predominant gastrointestinal involvement followed by a mild rash that developed later.18-23 Because our patient had severe duodenal bleeding, we at first ruled out the use of steroids and considered using intravenous immunoglobulin, which has been suggested in the literature to be effective.24-28 However, we changed our minds and decided to use steroids (2 mg/kg of methylprednisolone intravenously) as first-line therapy. Our patient showed complete progressive improvement of her general condition in the first 24 hours of steroid therapy, with complete remission after 3 weeks.

CONCLUSION

Although HSP is uncommon in the preadolescent/adolescent age group, nonthrombocytopenic palpable purpura with multiorgan involvement (gastrointestinal, skin, joints) should make one consider the diagnosis. Prompt diagnosis and multidisciplinary intervention can lead to appropriate management and mitigate potential complications.

REFERENCES