CASE STUDY IN
GASTROENTEROLOGY & HEPATOLOGY

Henoch-Schonlein Purpura With Gastrointestinal Involvement in an Adult Patient

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A 23-year-old woman with no significant medical history presented to a community hospital with abdominal pain and an erythematous, nonitching rash on the dorsal aspect of both feet for 3 weeks. She did not have drug or food allergies and was not taking any prescribed medications. Her family history was significant for celiac disease (in her mother) and cardiovascular disease and diabetes (in her father). The patient was of northern European heritage, and she was a current smoker with 7 pack years of cigarette smoking and occasional alcohol and marijuana use. Her physical examination showed a petechial rash affecting both lower extremities and also revealed tenderness in the epigastric region on deep palpation. Her laboratory studies, including complete blood count and serum chemistries (with liver tests), were normal. She underwent a computed tomography (CT) scan of the abdomen, which was unremarkable. The patient also underwent an upper endoscopy, along with biopsies of the duodenum, which showed gastritis and normal mucosa of the duodenum.

The next day, the patient was discharged on omeprazole, acetaminophen with codeine, and levofloxacin. However, she was readmitted the following day with nausea, vomiting, diarrhea, melenic stool, worsening abdominal pain, and skin rash spreading to her thighs. Repeat laboratory studies showed a white blood cell count of 25,500/μL (normal, 3800-10,800/μL), hemoglobin level of 11.8 g/dL (normal, 13-17 g/dL), platelet count of 423,000/μL (normal, 150,000-350,000/μL), serum creatinine level of 0.4 mg/dL (normal, 0.8-1.2 mg/dL), aspartate aminotransferase level of 12 U/L (normal, 10-35 U/L), alanine aminotransferase level of 26 U/L (normal, 10-35 U/L), total bilirubin level of 0.2 mg/dL (normal, 0.2-1.2 mg/dL), and alkaline phosphatase level of 51 U/L (normal, 40-115 U/L). A repeat CT scan of the abdomen showed a thickened small bowel (Figure 1). The patient's ultrasound of the abdomen was normal, while magnetic resonance angiography of the abdomen showed gallbladder sludge.

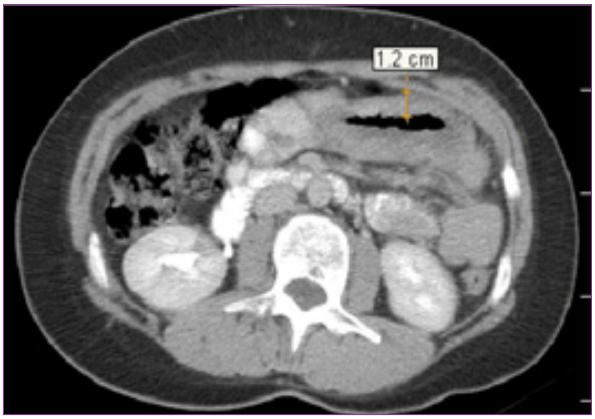

Figure 1. A computed tomography scan showing a thickened small bowel.

An esophagogastroduodenoscopy (EGD) with push enteroscopy revealed 2 nodular, ulcerated lesions in the jejunum, and a biopsy was obtained (Figure 2). The pathology report showed small intestinal mucosa with focal glandular hyperplasia and no definitive evidence of the celiac disease process. A skin biopsy of the pretibial region showed leukocytoclastic vasculitis (Figure 3). The patient was treated with intravenous corticosteroids, intravenous hydration, and bowel rest. She improved and was discharged on pantoprazole, prednisone, and acetaminophen with codeine after a 1-week hospital stay. A repeat EGD with push enteroscopy after 1 month was normal.

Discussion

Henoch-Schonlein purpura (HSP) is a systemic small-vessel immunoglobulin (Ig) A–dominant vasculitis characterized by palpable purpura, hematuria, abdominal pain, and

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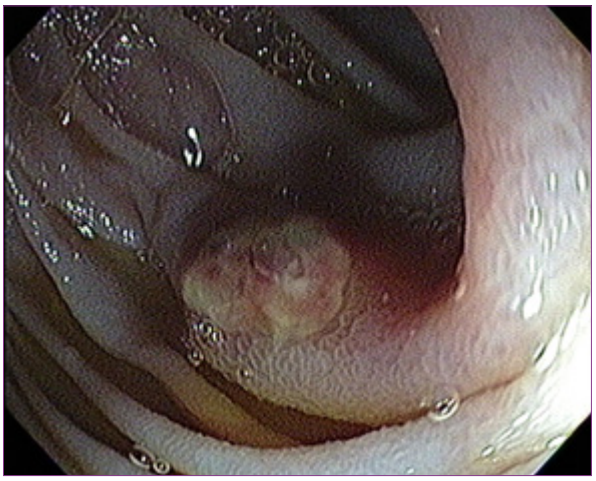

Figure 2. An endoscopic view of the jejunum.

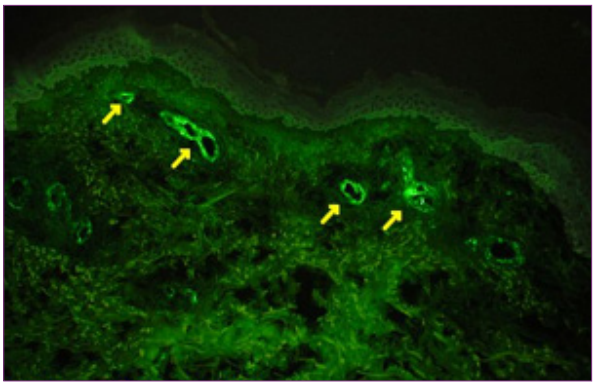

Figure 3. A skin biopsy showing positive immunofluorescence for immunoglobulin A.

arthritis.[1,2] HSP has been reported in patients as young as 6 months and as old as 86 years; however, 90% of patients are younger than 10 years.[1] The exact etiology and pathology of HSP remain unclear, but IgA immune complex deposition may play an important role. There are no specific tests available to establish the diagnosis. The postulated mechanism of HSP is abnormalities of immune response triggered by factors such as various drugs or infectious episodes, which is supported by the fact that HSP occurs more often in winter, autumn, and spring than in summer.[3]

In adults, HSP is less common and represents a more severe clinical syndrome with a higher frequency of renal involvement. In terms of GI involvement, 10% to 20% of patients in general present with abdominal complaints, and 85% have an abdominal complaint along with other symptoms.[2] Colicky abdominal pain, nausea, vomiting, and anorexia are the most common symptoms in patients with HSP; hematochezia can also occur, although less commonly.

The diagnostic criteria for HSP include the presence of at least 2 of the following: palpable purpura, age younger than 20 years at the onset of symptoms, bowel angina, and a biopsy with granulocytes in the vascular wall (leukocytoclastic vasculitis). The presence of 2 or more of these criteria identifies the illness with 87.1% sensitivity and 87.7% specificity.[4] Our patient met 3 of these criteria, confirming the diagnosis of HSP.

In HSP, the most common GI features are abdominal pain (86%), massive colorectal bleeding (20%), occult blood loss (66%), vomiting (40%), and diarrhea (20%).[5,6] GI symptoms result from submucosal and subserosal hemorrhages and the accumulation of fluid in the bowel wall caused by the underlying vasculitis.[6,7] In patients who present with only abdominal symptoms, upper and lower endoscopies are important for diagnosis.[8] In multiple case series, the small bowel and especially the duodenum (2nd portion) are most often affected. The duodenal bulb is rarely affected (which helps differentiate the condition from peptic ulcer disease). The stomach and colon are often involved as well.[8,9] The spectrum of endoscopic findings is based upon the severity of the vasculitis; usually, irregular, ulcerating, nodular lesions or hematoma-like protrusions are characteristic of HSP in the duodenum. The site preference and appearance also help differentiate HSP from peptic ulcer disease.[7-9] Our patient had small bowel involvement based upon CT and enteroscopy findings. The stomach, duodenum, and colon were not involved. The histology of jejunal lesions did not demonstrate leukocytoclastic vasculitis likely because of sampling error; however, the endoscopic appearance was very characteristic of HSP. Jejunal involvement likely caused GI bleeding in our patient.

HSP in adults, unlike in children, is associated with more frequent and severe kidney disease.[3,10,11] One study indicated that the outcome for adults is worse than for children, requiring aggressive treatment and a longer hospital stay and resulting in persistent kidney dysfunction years later.[11] Our patient did not have renal involvement, indicating a better prognosis.

The treatment of HSP with corticosteroids is controversial. Several case series have reported better outcomes in adult patients with GI and renal involvement who were treated with corticosteroids.[11,12] In a randomized trial, prednisone was effective in reducing abdominal pain; the incidence of severe abdominal pain requiring hospitalization was greater in the placebo group than in the prednisone group.[12] Analgesics, mainly acetaminophen, are useful for joint and muscle pain and for fever. Nonsteroidal anti-inflammatory drugs can be used to treat arthritis, but should be avoided in patients with GI and renal manifestations. Ranitidine, an H2 blocker, was found to be effective in patients with moderate GI involvement in a placebo-controlled trial.[2,13]

The authors have no relevant conflicts of interest to disclose.

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