BRIEF REPORT

Henoch-Schönlein Purpura: A Diagnosis Not To Be Forgotten

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Henoch-Schönlein purpura (HSP) is a systemic, generalized vasculitis of unknown etiology thought to be related to an IgA-mediated autoimmune phenomenon. Diagnosis is based on a constellation of physical findings that include the characteristic nonthrombocytopenic petechial or purpuric rash, migratory polyarthralgias, abdominal pain, and renal complications. We report the case of a 19-year-old man with a diagnosis of HSP who had severe abdominal pain and endoscopic documentation of duodenal involvement. Though not clear at presentation, the diagnosis became obvious when the characteristic rash emerged.

KEY WORDS. Schönlein-Henoch purpura; abdominal pain; gastrointestinal bleeding; rash.
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Henoch-Schönlein purpura (HSP) is a systemic, generalized vasculitis of unknown origin that primarily affects children but can also occur in adult patients. Diagnosis is based on a constellation of physical findings, including: (1) a characteristic nonthrombocytopenic petechial or purpuric rash, (2) migratory polyarthralgia, (3) renal involvement, and (4) gastrointestinal involvement. Failure to recognize this disease may lead to unnecessary tests and/or surgery being performed on these patients.

Case Report

A 19-year-old man presented with a 24-hour history of severe joint and muscle pain. He had intermittent abdominal pain for the previous 2 weeks and a sore throat for 1 week. The increasing colicky abdominal pain resulted in hospitalization. Physical examination revealed a temperature of 98°F, weight of 67.6 kg (75th to 90th percentile) and height of 172 cm (75th to 90th percentile). Examination of the joints showed tenderness and swelling of the right elbow and metatarsal phalangeal joints and tenderness over the left elbow. All other joints were normal.

On the first hospital day, several 1-cm red macules that blanched with pressure were noted over the ankles. On the third day, these lesions did not blanch, had increased in number, and were palpable. Additional lesions were noted on the back of the legs and the buttocks. The abdomen was scaphoid with mild guarding, but there was no rebound tenderness. Rectal examination revealed a small amount of stool, which was positive for occult blood, but no tenderness or perianal lesions.

Laboratory values were: white blood count (WBC) 11,500/mm^3, with polymorphonuclear leukocytes 83%, lymphocytes 8%, monocytes 8%, and eosinophils 2%, hemoglobin (Hgb) 15.9 g/dL, platelets 252,000/mm^3, erythrocyte sedimentation rate (ESR) 24 mm/h, aspartate aminotransferase 12 IU/L, alanine aminotransferase 13 IU/L, alkaline phosphatase 65 U/L, total protein 6.4 g/dL, albumin 3.5 g/dL, and uric acid 4.6 mg/dL. Rheumatoid factor and anti-nuclear antibody tests were negative. Urinalysis was within normal limits. Throat culture was negative. Plain radiograph of the abdomen was normal. Based on the physical findings and the palpable purpura, the patient's condition was diagnosed as HSP.

To evaluate the gastrointestinal (GI) bleeding and epigastric tenderness, an upper gastrointestinal endoscopy was performed. This examination revealed extensive ulceration with exudate of the entire descending duodenum and patchy areas of redness in the duodenal bulb. The rest of the exam-

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ination was normal. Biopsies of the descending duodenum showed hemorrhagic and ischemic changes of the surface epithelium and rare fibrin thrombi in the mucosal capillaries. Antral biopsies were normal and silver stain for *Helicobacter pylori* were negative. To evaluate the extent of small bowel involvement, a barium study of the stomach and small bowel was done. This revealed marked irregularity of the descending duodenum with partial narrowing of the third part of the duodenum. No other abnormal findings were noted.

The diagnosis of HSP was established based on the skin lesions and joint and gastrointestinal involvement. The patient was started on 40 mg of intravenous methylprednisolone. Within 24 hours, there was a marked decrease in abdominal pain and an increase in appetite. He was treated for 48 hours with intravenous methylprednisolone and discharged on oral prednisone at 40 mg/day. Within the next 2 weeks, the joint swelling and rash improved. The abdominal pain did not recur. The prednisone was tapered and stopped over 8 weeks. Follow-up examination at 2 months showed a weight gain of 3 kg, normal physical examination, and stools negative for occult blood. Subsequent urinalyses, up to 12 months, were also within normal limits. The patient refused a repeat upper GI endoscopy.

Discussion

This case demonstrates several typical features of HSP but also brings to light several atypical features. HSP is usually considered a disease of children: three fourths of patients with this condition are under the age of 7.[1] Our patient, at age 19, is atypical in that respect. HSP occurs primarily in males (2:1 ratio), with an incidence of 14 cases per 100,000 population.[1] It occurs most frequently in the spring and fall.[2,3] The disease is preceded by an upper respiratory or pharyngeal infection in up to two thirds of the patients.[3,4] Various precipitating factors, such as streptococcal infection, viral agents, insect bites, and drug and food allergies, have been implicated as causes of HSP.[2]

Our patient had symptoms that are considered classic of HSP: rash, joint involvement, and abdominal pain. On initial presentation, however, the rash was atypical in that it blanched with pressure and was macular in appearance, which delayed the diagnosis. In addition, this patient had gastrointestinal involvement in the form of severe abdominal pain, hypoalbuminemia, and endoscopically demonstrated severe duodenal lesions.

The reported frequency of gastrointestinal involvement in HSP varies between 29% and 69%.[5] Gastrointestinal symptoms most frequently include abdominal pain and guaiac-positive stools.[5] Other gastrointestinal manifestations of HSP are protein-losing enteropathy,[6] functional intestinal obstruction,[7] steatorrhea,[5] hydrops of gallbladder,[8] and pseudomembranous colitis.[8] Potentially life-threatening complications that might need surgery, such as intussusception, intestinal obstruction, bowel infarction, necrosis and perforations, massive GI bleeding, pancreatitis, hydrops of gallbladder, and scrotal swelling mimicking testicular torsion. These complications are rare and usually occur less frequently than the 8% reported in one series.[8] Increased awareness of these manifestations of HSP, particularly abdominal pain or scrotal swelling or both in the absence of the typical rash, is needed to prevent unnecessary surgical exploration.[8]

Determination of gastrointestinal involvement generally occurs through radiological studies,[9] operative observation of intestinal lesions visible from the serosal side,[10] or endoscopy.[11,12] Changes noted from the serosal side of the small bowel during surgery are similar to those described in inflammatory bowel disease.[8] The indications for upper GI endoscopy in patients with HSP are epigastric pain, hematemesis, and melena.[12] In both children and adults, there have been few reports of endoscopic changes of the gastrointestinal tract associated with HSP.[1,5,10-13] Changes described in these reports included gastritis, duodenitis, and multiple colonic "red" lesions. As in our case, many of the previously reported cases have had severe mucosal involvement of the descending duodenum, indicating that this site might be a characteristic area of involvement in HSP.[12] Similar changes in the duodenum may also be seen in other diseases, such as chronic inflammatory bowel disease. Evidence of vasculitis from the biopsied samples combined with the clinical picture, however, confirms the diagnosis of HSP. *H pylori* has been incriminated as the cause of recurrent HSP in one patient.[14] In this case, the skin rash, GI complaints, and proteinuria improved with eradication of the *H*

pylori. In our patient, *H pylori* was absent in the antral biopsies.

Changes in the GI tract in patients with HSP as detected by barium study are nonspecific.[14] Changes in the small bowel include separation of the loops and mucosal fold thickening. In the colon, these changes may appear as mucosal scalloping and "thumbprinting." The findings in the small bowel may be indistinguishable from Crohn's disease, lymphoma, or other reversible conditions in which submucosal bleeding occurs.

The rash of HSP occurs in some form in all cases, but is the presenting feature in 50% of patients.[2] The rash may consist of erythematous maculopapules, urticarial wheals, large palpable purpura, petechiae, erythema multiforme, and, in severe cases, necrotic lesions. The distribution of the rash is symmetrical and tends to be on the dependent part of the body. Thus, the lower extremities, particularly the extensor surfaces and buttocks, are classically involved, with palpable purpura, petechiae, and ecchymoses. In a typical presentation, these lesions occur in crops that fade over several days. Should there be recrudescence, new lesions may recur at the sites of previous lesions. The upper extremities, trunk, and head tend to be involved more often in children under 2 years of age, as the entire body is dependent much of the time. Angioedema may be present, especially in infants and young children in whom it may be a prominent feature, involving the scalp, periorbital area, hands, and feet. The rash tends to fade over 1 to 4 weeks, although, in rare cases, it may persist for months or even years.[1,2,5] Other than rash, the most common manifestations of HSP are arthralgia and arthritis of the large joints: the knees, wrists, shoulder, and elbows. Joint involvement, which is transient, occurs in up to 90% of cases.[1]

Of the multiple organ systems involved in HSP, the kidney has the greatest potential for serious morbidity or mortality. In a recent review of reports from several unselected populations, the proportion of patients with "renal involvement" varied widely, from 20% to 100%.[15] Our patient is somewhat atypical in not having evidence of renal involvement since, according to Meadow,[15] microscopic hematuria or albuminuria (or both) are found in 30% to 70%. Additionally, there is an impression that the kidney is more likely to be involved when intestinal manifestations are severe. The prognosis is most favorable for those with minimal or no renal manifestations at the onset of illness. The mortality cited by Meadow of 1% to 3% includes patients with severe acute illness at onset (rapidly progressive crescentic glomerulonephritis) and those with significant residual renal disease and slowly progressive decline in renal function, culminating in end-stage renal disease. At 1 year after onset, our patient had no evidence of renal disease and is expected to have a good outcome.

Apart from the usual features, the following unusual features of HSP have also been described: epistaxis, focal neurological deficits, seizures, intracranial hemorrhage, peripheral mononeuropathies, pulmonary hemorrhage, pulmonary interstitial disease, myocardial infarction, and cardiac tamponade.[2] Delayed effects of HSP, which are rare, include esophageal stenosis,[16] ileal stricture, and stenosing ureteritis.[8]

The common inflammatory process affecting multiple organ systems in HSP is that of leukocytoclastic vasculitis triggered by subendothelial deposition of circulating IgA immune complexes in small arteries and capillaries. The consequent activation of the complement system causes release of chemotactic factors, attracting polymorphonuclear leukocytes into the wall of arteries that, in turn, release proteolytic enzymes, resulting in vascular necrosis.[17] Inflammatory changes and deposition of IgA immunoglobulins have been noted in biopsies of gastrointestinal mucosa, kidneys, and skin.[10]

The differential diagnosis for a patient presenting with rash, abdominal pain, and joint symptoms includes inflammatory bowel disease, *Yersinia* enterocolitis, meningococcal disease, rheumatic fever, hepatitis B prodrome, infectious mononucleosis, as well as Rocky Mountain spotted fever (*Rickettsia rickettsii*). Lupus and other systemic vasculitides might also be considered in the differential diagnosis. The combination of the characteristic purpuric skin rash, inflammatory joint pain, and GI symptoms should suggest a diagnosis of HSP; renal involvement confirms the diagnosis of HSP. Surgical exploration should be considered cautiously in a patient with severe, colicky abdominal pain in the absence of other more definitive symptoms of HSP, such as intestinal obstruction or

peritonitis. This precaution may avoid unnecessary surgery, as the rash, confirming HSP, may follow the abdominal pain in some patients with this disease.

Laboratory evaluation generally demonstrates a normal to mild leukocytosis, occasionally accompanied by eosinophilia. The platelet count is invariably normal. ESR and C-reactive protein may be moderately elevated, although they are just as likely to be within normal limits. Urinalysis will generally demonstrate hematuria, proteinuria, and occasional red cell casts. Immunoglobulin electrophoresis will generally—at least 50% of the time—demonstrate elevation of IgA. Renal dysfunction, as manifested by elevated blood urea nitrogen or serum creatinine is considerably less frequent. Serologic testing for viral hepatitis, rheumatoid arthritis, systemic lupus, and streptococcal infection will help eliminate other considerations.

In general, treatment is supportive, including acetaminophen or nonsteroidal anti-inflammatory drugs for arthralgia and discontinuation of any drugs suspected as an allergen. Because of our patient's severe abdominal pain and endoscopic findings, a short course of steroid therapy was extremely helpful in ameliorating his symptoms. The use of steroid therapy for abdominal pain in HSP is controversial. A retrospective review of 43 children with HSP and abdominal pain who were treated with prednisone 1 to 2 mg/kg showed that the treatment may have a role in hastening resolution of the self-limited pain.[4] These patients, however, were not separated into guaiac-positive and guaiac-negative groups; doing so might have explained the severity of the gastrointestinal lesions. In the experience of one of the authors (T.S.G.), patients with HSP and endoscopically documented gastrointestinal involvement have shown a significant improvement in abdominal pain within 48 hours of treatment with steroids, although the eventual outcome may not have been different. Other successful interventions include the use of cytotoxic drugs as well as the use of plasmapheresis. These more aggressive therapies might be considered in patients with evidence of severe renal disease.[15] HSP rarely requires such aggressive intervention, however, and most patients fully recover within 1 or 2 months of disease onset.

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