Pneumatosis Intestinalis Associated With Henoch–Schönlein Purpura

abstract

Henoch–Schönlein purpura (HSP) is the most common vasculitis in children. It is a disorder of the inflammatory cascade leading to immunoglobulin A deposition and leukocytoclastic vasculitis of small vessels of skin, kidneys, joints, and gastrointestinal (GI) tract. A wide variety of GI manifestations are seen in ~50% to 75% of patients with HSP. Diffuse colicky abdominal pain is the most common GI symptom. The small bowel is the most frequently involved GI site. Intussusception is rare but is the most common surgical complication. We report the case of a 2-year-old girl with a 5-day history of abdominal pain followed by a palpable purpuric rash. Her urinalysis, complete blood cell count, and tests of renal function were normal. An acute abdominal series was unremarkable initially, and abdominal ultrasound imaging showed ascites and thickened small bowel loops. She was diagnosed with HSP. The abdominal pain worsened, and an abdominal computed tomography scan demonstrated distal small bowel wall thickening and pneumatosis intestinalis in the descending colon. She was started on total parenteral nutrition and antibiotics and placed on bowel rest. She was given 2 mg/kg of intravenous immunoglobulin. Her abdominal pain gradually improved over the next week, and a repeat computed tomography scan showed significant improvement of the small bowel wall thickening and pneumatosis. The purpuric rash improved, and her abdominal pain resolved. We report a case of HSP and pneumatosis intestinalis, an association that has not been reported previously. *Pediatrics* 2014;134:e880–e883

AUTHORS: Ayesha Fatima, MD,* and Donald Paul Gibson, MD†

*Pediatric Gastroenterology, Department of Pediatrics, and †Department of Diagnostic Radiology, Beaumont Children’s Hospital and Oakland University William Beaumont School of Medicine, Rochester, Michigan

KEY WORDS

Henoch–Schönlein purpura, pneumatosis intestinalis, pneumatosis coli, abdominal pain

ABBREVIATIONS

CT—computed tomography

GI—gastrointestinal

HSP—Henoch–Schönlein purpura

PI—pneumatosis intestinalis

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Address correspondence to Ayesha Fatima, MD, 3601 W Thirteen Mile Rd, Royal Oak, MI 48073. E-mail: afatima@beaumont.edu

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Henoch–Schönlein purpura (HSP) is associated with a variety of gastrointestinal (GI) manifestations, and these are seen in 50% to 75% of patients. We report the first case of HSP with associated pneumatosis intestinalis (PI).

CASE REPORT

A 2-year-old girl with an uneventful past medical history presented with an acute onset of periumbilical abdominal pain followed by a rash over her lower extremities. One week before this episode, she was diagnosed with a left otitis media and started on amoxicillin. Examination in the emergency department was unremarkable except for a pinpoint erythematous rash over her lower extremities, and she was discharged with a diagnosis of viral syndrome.

She returned to the emergency department 4 days later with sharp, intermittent abdominal pain and a palpable purpuric rash that now involved the lower extremities, buttocks, upper arms, and lower back. Her urinalysis, complete blood cell count, and tests of renal function were normal. An abdominal ultrasound examination revealed moderate ascites and both proximal and distal small bowel wall thickening. She was diagnosed with HSP, and her abdominal pain was controlled by acetaminophen.

She returned 5 days later with a history of passing loose stools and bright red blood. An acute abdominal series was unremarkable, and repeat abdominal ultrasound imaging showed ascites and persistently thickened small bowel loops. The previous laboratory studies were repeated and were normal. She was admitted and received 1 dose of prednisone, 1 mg/kg. Overnight, she had worsening of abdominal pain, and an abdominal computed tomography (CT) scan demonstrated PI in the descending colon and thickening of the distal small bowel wall (Figs 1, 2, and 3). She was started on total parenteral nutrition, bowel rest, and vancomycin and piperacillin/tazobactam. Because of the PI, prednisone was discontinued, and she was given 2 mg/kg of intravenous immunoglobulin. She developed right upper extremity venous thrombosis at the venous catheter line site. Detailed hematologic investigations including complete blood cell count with platelets, prothrombin time, partial thromboplastin time, fibrinogen, anti-thrombin III, protein C activity, protein S activity, anticardiolipin antibody, factor V Leiden genotyped, prothrombin mutation genotyped, inhibitor screen, and factor VIII level were all normal. The hyperhomocystinemia genotype was heterozygous for the MTHFR C677T allele, but the serum homocysteine level of 2 μmol/L was low normal. Her abdominal pain gradually improved over the next week, and a repeat CT scan showed significant improvement of the pneumatosis and small bowel wall thickening. The rash improved, her abdominal pain resolved, and she was discharged from the hospital on a regular diet.

Two weeks after her discharge, she again developed abdominal pain and a worsening rash, and she was started on prednisone 1 mg/kg per day. This was tapered when her symptoms resolved.

DISCUSSION

HSP is the most common vasculitis in children. It is a disorder of the inflammatory cascade leading to immunoglobulin A deposition and leukocytoclastic vasculitis of small vessels of the skin, kidneys, joints, and GI tract. Palpable
purpura is the hallmark of the disease,\(^1\) and abdominal symptoms precede the purpuric rash in 14% to 36% of patients.\(^2\) Colicky abdominal pain is the most common abdominal symptom and is attributed to bowel ischemia.\(^2,3\) Small bowel is the most frequently involved GI site, with the proximal small bowel and distal ileum usually more involved.\(^3\) Other GI manifestations include bleeding, gastric ulcers, intussusception,\(^1,3\) pancreatitis, gallbladder hydrops, and protein-losing enteropathy. Less common surgical complications include intestinal obstruction, stricture formation, acute appendicitis, enteric fistulas, spontaneous bowel perforation, esophageal strictures, GI hemorrhage, and infarction.\(^2,5,6\) There are no laboratory features diagnostic for HSP, and imaging with abdominal ultrasound usually identifies the surgical complications.\(^2,3,5\) In our patient, worsening abdominal pain with a normal acute abdominal series suggested the need for a CT scan that demonstrated PI. Although the natural course of the disease is usually benign, given the known surgical complications, recurrent abdominal pain or other variation from the expected course warrants additional investigation. Her worsening abdominal pain prompted us to perform CT imaging, and this was done with low-dose protocols, keeping radiation exposure to the minimum.

PI is defined as the presence of intramural gas in the small or large bowel.\(^7\) It has been associated with a variety of diseases beyond the neonatal period\(^8\) and has been observed as an incidental finding in healthy children.\(^7,9,10\) It has also been described in patients with nonintestinal conditions such as organ and bone marrow transplants, congenital heart disease, bronchopulmonary dysplasia, rheumatoid arthritis, and hemolytic anemia.\(^8\) It occurs more frequently in immunocompromised patients who have received steroids, chemotherapy, or radiotherapy.\(^7\) The clinical manifestations include diarrhea, bloody stools, abdominal pain, abdominal distension, tenesmus, vomiting, constipation, and weight loss, and the management is symptomatic (bowel rest, nasogastric suction, antibiotics) and depends on the underlying disease. Surgery is indicated in fulminant cases and is associated with a high mortality.\(^7,9\)

Mechanical and bacterial causes have been proposed to explain gas entry into the bowel wall.\(^7,9\) Mechanical causes include the introduction of intramural air after forced bowel distension or a mucosal break. PI has been described in patients with elevated intraabdominal pressure due to violent coughing or vomiting.\(^7,11\) Gas within the bowel wall could be the result of the entry of gas-forming bacteria through mucosal breaks. In our patient, bowel wall ischemia might have led to mucosal disruption, allowing bacteria and intraluminal gas to enter the bowel wall. Mucosal ulceration and localized

**FIGURE 2**
An axial image from a contrast-enhanced CT scan demonstrates the circumferential air collections within the descending colonic wall (arrows).

**FIGURE 3**
An axial CT image through the pelvis shows evidence of several thick-walled distal small bowel loops (arrows) consistent with the diagnosis of HSP.
vasculitis associated with HSP can also predispose the mucosa to injury. Although steroids can alter mucosal integrity and increase permeability,

they are unlikely to have been the cause of the pneumatosis in our patient because she had received only one dose.

More than 80% of patients with HSP recover within 2 weeks, but in the first year it recurs in ~30% to 40% of patients, often within 4 months. Recurrence of symptoms within 12 weeks of onset is considered part of the initial disease, whereas later recurrence is considered true recurrence. Abdominal pain is a common recurrent symptom. HSP without renal involvement is usually a self-limiting disease with complete resolution. PI is uncommon, has a heterogeneous presentation, and is associated with a variety of diseases, but because it can have serious consequences, PI must be considered as a possible complication of HSP.

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