

Terminal Ileitis Induced by Henoch–Schonlein Purpura That Presented as Acute Appendicitis

A Case Report

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Abstract: Henoch–Schonlein purpura (HSP) is a self-limited autoimmune disease, the cause of which is not clear. Gastrointestinal involvement is often the main symptom of HSP. We report an unusual and rare case in a patient who was diagnosed with HSP. This is the second report of terminal ileitis induced by HSP that presented as acute appendicitis.

We report a 21-year-old man who presented with right lower abdominal pain, and was diagnosed with acute appendicitis. Terminal ileitis was diagnosed intraoperatively, and when a rash occurred postoperatively, the final diagnosis was HSP.

When the rash occurred, HSP was diagnosed and methylprednisolone was administered for 5 days.

The diagnosis of HSP is difficult to establish, especially when the purpura occurs after gastrointestinal involvement; thus, abdominal pain should not be ignored and HSP should be considered.

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Abbreviations: CT = computed tomography, HSP = Henoch–Schonlein purpura, IgA = antigen–antibody.

INTRODUCTION

Henoch–Schonlein purpura (HSP) is a self-limited autoimmune disease, the etiology of which is not clear, but is known to be associated with infections, medications, tumors, vaccinations, α -1-antitrypsin deficiency, and familial Mediterranean fever.^{1,2} The clinical manifestations of HSP include palpable purpura, arthritis, and renal and gastrointestinal involvement.³ HSP is a systemic disease. The complement pathway is activated by antigen–antibody (IgA) complexes, which lead to small-vessel vasculitis and inflammation.¹ HSP is common in

children between 2 and 10 years of age, and 90% of affected individuals are <10 years of age.^{4,5} HSP occurs most often in the winter and spring. The incidence of HSP in adults is 3.4 to 14.3 per million; however, because HSP is self-limited, the actual incidence may be underreported.^{1,6}

Abdominal pain is the most common symptom of gastrointestinal involvement in patients with HSP. There are also other symptoms, such as nausea, vomiting, hematemesis, melena, and hematochezia. Abdominal pain precedes the rash in 12% to 19% of patients.⁵ We present an unusual and rare case, in which a patient was diagnosed with acute appendicitis preoperatively, terminal ileitis was diagnosed intraoperatively, and when the rash occurred postoperatively, the diagnosis was revised to HSP.

CASE PRESENTATION

A 21-year-old man presented to our emergency room with a complaint of right lower abdominal pain for 2 days. On physical examination, the vital signs were as follows: temperature, 37.3°C; heart rate, 94/min; respiratory rate, 20/min; blood pressure, 120/75 mm Hg. The abdominal examination was significant for tenderness and pain in the right lower quadrant upon palpation. There was no rebound tenderness or muscle rigidity. The remainder of the physical examination was normal. Laboratory testing revealed the following: white blood cell count, $9.54 \times 10^9/L$; neutrophilic granulocytes, 81.1%; hemoglobin, 136 g/L; platelet count, $232 \times 10^9/L$. Kidney function and other biochemical measures were normal. Computed tomography (CT, Figure 1A and B) showed pneumatosis in the appendix and fluid in the pelvic cavity. Based on the above findings, a diagnosis of acute appendicitis was made and a laparoscopic appendectomy was undertaken; however, the appendix was normal in appearance and an appendectomy was not performed. Intraoperatively, the terminal ileum was noted to be approximately 15 cm in diameter, hyperemic, and edematous (Figure 2); there was no apparent inflammation of the appendix. Thus, terminal ileitis was diagnosed. Three days postoperatively, the patient had rose-colored petechiae and palpable purpura on the lower extremities, and HSP was diagnosed. Subsequently, methylprednisolone (2 mg/kg/d iv) was administered for 5 days. Seven days postoperatively, the abdominal pain and purpura resolved. There were no recurrences in the subsequent 8 months, and long-term follow-up in the clinic was recommended.

DISCUSSION

HSP is a self-limited, systemic, nongranulomatous, autoimmune, small-vessel vasculitis with multiorgan involvement.¹ HSP is characterized by nonthrombocytopenic palpable purpura most often involving the lower extremities and buttocks,

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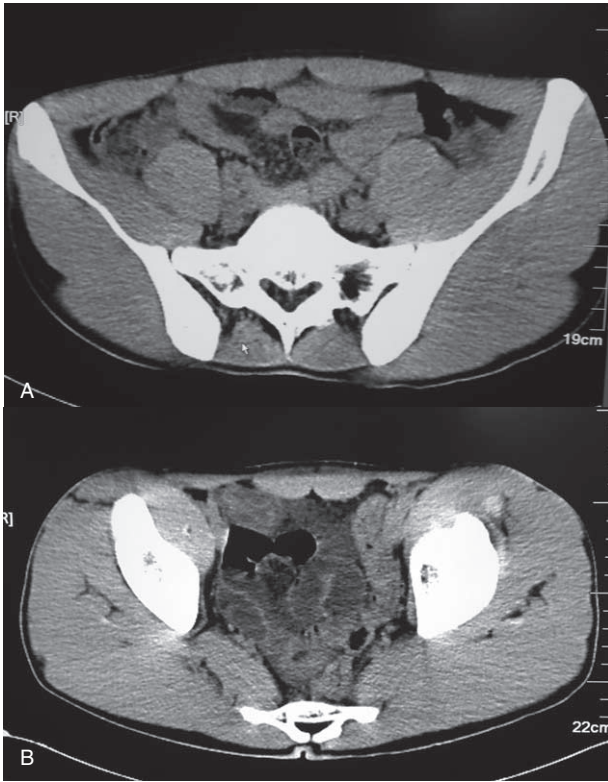


FIGURE 1. (A and B) CT scan shows pneumatosis in the appendix and fluid in the pelvic cavity. CT = computed tomography.

arthralgia/arthritis, bowel angina, and hematuria/proteinuria.⁷ HSP is a multisystem disorder, which can affect the gastrointestinal tract, skin, joints, and kidneys, but rarely the nervous system, lung, heart, and genitourinary tract are affected.⁸ Greater than 90% of patients with HSP are children, and the incidence in adults is low.⁹

The etiology of HSP is unclear but associated with infections (bacterial, viral, and parasitic), medications (adalimumab), vaccinations, tumors (non-small cell lung cancer, prostate cancer, and hematological malignancies), and α -1-antitrypsin deficiency.^{1,10}

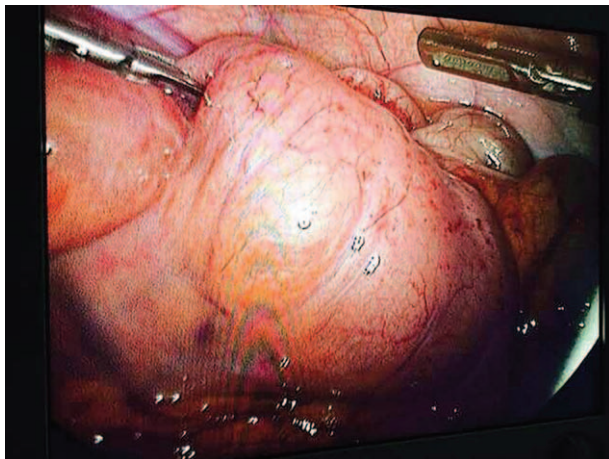


FIGURE 2. Hyperemia and edema involving the terminal ileum.

The highest incidence of HSP has been reported in Asians.¹ The incidence of HSP in North America is 13.5/100,000 children; whites have the highest incidence, and Afro-Americans have the lowest incidence.¹

HSP is usually diagnosed based on an atypical presentation; however, when atypical presentations are not evident, noninvasive diagnostic methods are needed to establish the diagnosis of HSP.⁵ β 2GPI facilitates the diagnosis of HSP,¹¹ and visfatin is abnormally elevated in patients with HSP.¹²

Abdominal pain is the most common symptom of the gastrointestinal tract in patients with HSP. In the present case, the patient came to the hospital for evaluation of right lower abdominal pain, and was thought to have acute appendicitis. HSP has been associated with other gastrointestinal symptoms, including nausea, vomiting, hematemesis, melena, and hematochezia. There are also other rare gastrointestinal symptoms in patients with HSP, such as intussusception, ischemic necrosis of the bowel wall, intestinal perforation, massive gastrointestinal bleeding, acute acalculous cholecystitis, hemorrhagic ascites with serositis, pancreatitis, and biliary cirrhosis.¹

Cutaneous symptoms often precede gastrointestinal symptoms in patients with HSP; nevertheless, skin lesions occur after gastrointestinal manifestations in 25% of patients, which make the diagnosis difficult and may even result in unnecessary surgery.^{13,14} Our case is a typical example that can pose a diagnostic dilemma for physicians. Specifically, right lower abdominal pain and CT findings led to laparoscopic surgery. Intraoperatively, the appendix was noted to be normal in appearance without inflammation, and the terminal ileum was congested and edematous, thus, terminal ileitis was diagnosed.

Terminal ileitis has rarely been reported in 9 patients with HSP.^{5,15,16} This is the second report of a patient with terminal ileitis who presented with acute appendicitis induced by HSP. The first case was reported by Yantis¹⁷ in 1973, which illustrated a patient presented with acute appendicitis and Crohn disease.

Three days after laparoscopic surgery, purpura developed on the lower extremities. At that time, the diagnosis was revised and with appropriate therapy, the patient recovered.

The differential diagnosis of HSP includes Crohn disease, Wegener granulomatosis, infective endocarditis, IgA nephropathy, and hemolytic uremic syndrome. The differential diagnosis of HSP accompanied by terminal ileitis should include Crohn disease, vasculitides, and infections.^{1,5} There are some diseases that should be considered in a patient with pain in the right lower quadrant is evaluated, including appendicitis, terminal ileitis, Crohn disease, HSP, and other infections.

CONCLUSION

Gastrointestinal involvement is frequent in patients with HSP. The diagnosis of HSP can be challenging, especially when abdominal symptoms precede cutaneous lesions (palpable purpura). HSP should be included in the differential diagnosis of an acute abdomen so that physicians avoid unnecessary surgical procedures.

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