Henoch-Schönlein Purpura Presenting with Acute Abdomen

Oguz Catal, Bahri Ozer and Mustafa Sit

Department of General Surgery, Abant Izzet Baysal University Hospital, Bolu, Turkey

ABSTRACT

Henoch-Schönlein Purpura (HSP) is an autoimmune, systemic, non-granulomatous vasculitis characterised by self-limiting clinical course, and leukocytoclastic vasculitis of small vessels. Deposition of immune complexes that contain IgA is the hallmark of vascular involvement. Adults with HSP have a higher incidence of complications and death. The most common gastrointestinal (GI) symptom of HSP is abdominal pain. Vasculitis-related colitis, causing extensive lower GI hemorrhage, is uncommon in the course of HSP, which increases the risk of renal involvement.

Here, we present two cases of HSP with GI involvement. In the first case, surgery was performed. Whereas, the other patient was treated medically due to the experience gained from the first case.

The HSP has no specific treatment. Most of the cases are mild and need only supportive care due to its self-limiting nature. Although corticosteroids do not prevent recurrences, evidence in literature suggests that these are beneficial in resolution of the arthritis and abdominal pain. Aggressive therapy with corticosteroids or cyclophosphamide is not successful in reducing renal damage, except in patients with crescentic nephritis.

Key Words: *Henoch-Schönlein purpura, Gastrointestinal complications, Steroids, Surgery.*

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INTRODUCTION

Henoch-Schönlein Purpura (HSP) is an autoimmune, systemic, non-granulomatous vasculitis characterised by self-limiting clinical course, and leukocytoclastic vasculitis of small vessels.[1] Clinical signs are arthritis, palpable purpura without thrombocytopenia and coagulopathy, renal and gastrointestinal (GI) involvement. HSP is more common in childhood with a peak incidence in 4 - 6 years of age.[2] In adults, the incidence varies between 3.4 - 14.3 per million population. The prognosis of patients with childhood-onset HSP is good; however, in adults, HSP has a higher incidence of complications.[3] Abdominal pain is the most common symptom of GI involvement. Extensive lower GI hemorrhage, due to colitis associated with vasculitis, is an uncommon presentation of HSP; and it can be associated with an increased risk of renal involvement.[4]

Here, we present two cases of HSP. One of that case was an elderly man who had small vessel vasculitis of small intestine with ischemic changes presenting with abdominal pain and rectal bleeding. The other case also had small vessel vasculitis of small intestine with ischemic changes presenting with abdominal distension and abdominal pain.

Correspondence to: Dr. Oguz Catal, Department of General Surgery, Abant Izzet Baysal University Hospital, Bolu, Turkey
E-mail: otuzogur@gmail.com

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CASE REPORTS

Our first patient was a 60-year man who presented with purpura and petechia in both lower and upper extremities for 10 days. A skin biopsy revealed leukocytoclastic vasculitis. He also had abdominal pain for the last 3 days. On follow-up two days later, abdominal pain worsened and he developed vomiting, and hemorrhagic diarrhea. On physical examination, the body temperature was 37°C, a pulse of 110/min, and blood pressure of 130/80 mmHg. There were palpable purpuric rashes on both lower and upper extremities. On abdominal examination, muscle guarding and distension of the abdominal wall were detected. On auscultation, the bowel sounds were hypoactive. X-rays of the chest and abdomen were within normal limits. Diffuse thickening of the bowel wall and ascites were revealed by abdominal computerised tomography (CT). Laboratory tests were as follows: White blood count (WBC): 9,480/mm^3, Hemoglobin (Hb): 11.7 g/dl, and C-reactive protein (CRP): 235 mg/dl. Upper GI endoscopy revealed bulbitis, while lower GI endoscopic examination was normal. During surgery, small bowel looked edematous but not gangrenous, so we did not resect the bowel. The bowel was covered with Bogota bag; and after 48 hours, the bowel was reassessed. Edema subsided, bowel resection was not necessary, and the abdomen was closed. Meanwhile, intravenous treatment with prednisolone sodium succinate (60 mg first day, 40 mg after three days), and enoxaparin sodium, 6000 IU subcutaneous bid were administered. The skin lesions and abdominal pain were greatly reduced. After that, he was continued on oral prednisolone treatment for six months with no recurrence or progression of symptoms.

The second case was consulted eight months after the first case, when he was hospitalised in the Internal Medicine Clinic. This patient had a history of HSP. Abdominal pain, vomiting and abdominal distension developed, while he was being treated for diabetes mellitus in the Internal Medicine Clinic. This patient, 45-year male, had noticed purpura and petechia appearing on both lower and upper extremities. Since last two days, he had abdominal pain; and with increasing abdominal pain, he began developing vomiting and abdominal distention. On examination, the body temperature was 37.4°C, pulse was 105/min with sinus rhythm, and blood pressure was 110/80 mmHg. Physical examination revealed palpable purpuras on both lower and upper extremities. Per abdomen, abdominal distension, muscle tenderness and hypoactive bowel sounds were noted. X-ray of the chest and abdomen were normal. CT of the patient's abdomen demonstrated diffuse thickening of the bowel wall and ascites. Laboratory testing showed WBC of 13,200/mm^3, Hb: 13.4 g/dl, and CRP: 190 mg/dl. Surgical intervention was not considered in this patient, based on previous experience. The patient was started on intravenous prednisolone sodium succinate (1 mg/kg). Enoxaparin sodium, 6000 IU subcutaneous bid, was administered. The abdominal compartment pressure decreased to 15 mmHg 6-8 hours after the start of treatment. The skin lesions and abdominal pain greatly reduced. After that, the patient was continued on oral prednisolone treatment for six months with no recurrence or worsening of symptoms.

DISCUSSION

HSP has an unclear etiology. The possible causes of HSP are: infections (bacterial, viral, parasitic), certain medications (antibiotics, angiotensin converting enzyme (ACE) inhibitors and non-steroidal anti-inflammatory drugs (NSAIDs), cancer (non-small cell lung cancer, prostate cancer and hematological malignancies), vaccines, familial Mediterranean fever, and alpha-1-antitrypsin deficiency.

Antigen-antibody complexes accumulate in the wall of small vessels, thus alternate complement pathway is activated. This leads to neutrophil accumulation, resulting in non-granulomatous inflammation and vasculitis. Although any organ or system could be involved, skin, GI system, kidneys and joints are the most affected regions of the body. Edema and hemorrhage are caused by extravasation of blood and its components into the interstitial spaces as a consequence of vasculitis.[5] The most common GI symptom of HSP is abdominal pain. Nausea, vomiting, hematemesis, melena and hematochezia make the other GI manifestations of the disease. Vasculitis of the GI tract leads to edema and hemorrhage of the bowel wall; whereas, mesenteric vasculitis has a high risk of necrosis and massive GI bleeding.[6] Intussusception, infarction and perforation are important complications of GI involvement. Poor prognosis has been reported in HSP patients older than 60 years of age.[7] In our cases, small intestine was edematous and there was hemorrhage of the bowel wall. The patients had abdominal compartment syndrome due to edema of the bowel wall.

Although skin manifestations precede GI involvement, these developed after GI symptoms in four cases in literature. In our patients, skin manifestations preceded GI manifestations and this could be beneficial in early diagnosis and treatment. Differential diagnosis of acute abdomen should include 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%). GI symptoms result from submucosal and subserosal hemorrhages, thus the accumulation of fluid in the bowel wall caused by the underlying vasculitis.[8]

HSP has no specific treatment. Most of the patients with HSP have mild symptoms and require only supportive care. While corticosteroids do not prevent recurrences, evidence in literature suggests that these are beneficial in resolution of arthritis and abdominal pain.

First of our patient underwent symptomatic treatment and intravenous methylprednisolone, but the abdominal pressure increased upto 25 cmH2O; henceforth, he underwent surgery. Small bowel looked edematous but not gangrenous so we did not decide to perform resection; and we observed bowel under Bogota-bag. Edema resorbed subsequently and bowel resection was not necessary. Abdominal pain and gastrointestinal bleeding complaints were improved. Skin lesions faded, and GI involvement responded well to systemic steroid therapy. In second patient, we did not perform surgery; and treated the patient with intravenous methylprednisolone and there was good response to this treatment.

Intussusception is the most common surgical complication of HSP. Other complications include: acute appendicitis, bowel obstruction, ischemia and necrosis, spontaneous perforation of the bowel, entero-enteral fistula, hemorrhage, pseudomembraneous colitis, inflammatory bowel disease, steatorrhea, acute pancreatitis, gall bladder involvement, ileal strictures, and ventral abdominal involvement.

PATIENTS' CONSENT:

We obtained informed consents from both the patients to publish the data concerning this case.

CONFLICT OF INTEREST:

The authors declared no conflict of interest.

AUTHORS' CONTRIBUTION:

OC: Design of the work, interpretation and analysis of data, and writing manuscript.
BO: Collection of data.
MS: Drafting the work or revising it critically for important intellectual content.

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