Surgical Aspects of Henoch-Schönlein Purpura in Adults

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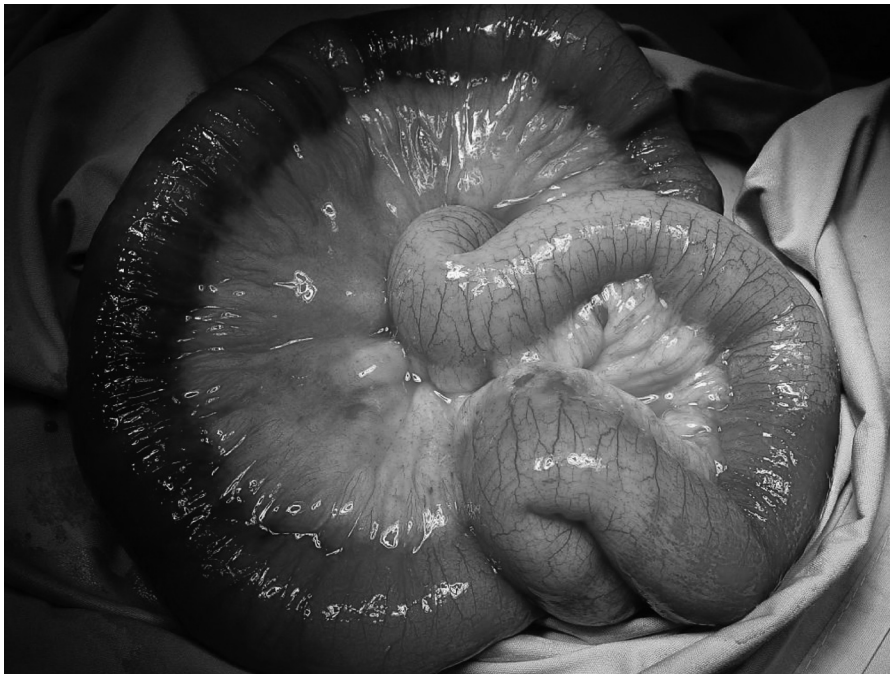
Henoch-Schönlein purpura is a leukocytoclastic vasculitis involving small vessels with deposition of immune complexes containing immunoglobulin A. It is characterized by the classic tetrad of palpable purpura, joint involvement, gastrointestinal symptoms and renal disease. Henoch-Schönlein purpura is mainly a childhood disease – 90% of cases occur between the age of 3 and 20 years, with peak incidence at age 5–7 [1]. In adults the disease is characterized by more severe renal involvement, less frequent gastrointestinal involvement, and poor general prognosis [2]. Henoch-Schönlein purpura is usually a self-limiting disease and long-term prognosis depends mainly on the initial renal involvement. Gastrointestinal manifestations can vary greatly and range from mild abdominal discomfort to frank acute abdomen. We present here two cases recently admitted to our department with substantial gastrointestinal manifestation and discuss possible surgical aspects of the disease.



Terminal ileum in case 1, note the patchy ecchymosis, intestinal and mesentery hyperemia and edema.

Patient Descriptions

Patient 1

A 54 year old man presented with 3 days of severe diffuse abdominal pain. He denied fever, diarrhea and vomiting and had no other complaints. The patient's medical history was significant for hypertension and mild diabetes. Physical examination revealed a diffusely tender abdomen without peritoneal signs, and palpable purpuric lesions on both legs. Laboratory findings showed elevated white blood cell count of 14×10^6/dl and impaired renal function. The stool tested positive for occult blood and there was microscopic hematuria. Computed tomography of the abdomen showed wall thickening of the distal ileum with increased density of the mesentery and a moderate amount of free fluid in the peritoneum.

On the basis of the clinical and radiological picture with the differential diagnosis of mesenteric ischemia versus small bowel vasculitis, we performed an exploratory laparotomy. Surgery revealed a 50 cm ileal segment with significant hyperemia and edema but without signs of necrosis or perforation [Figure]. Intraoperative Doppler showed good arterial flow without venous engorgement. There was no further intervention and a 10 mm trocar was left in the surgical wound for second-look laparoscopy. Treatment was started with heparin and hydrocortisone. Second-look laparoscopy 48 hours later revealed substantial improvement without need for further surgical intervention. A skin biopsy from the leg revealed leukocytoclastic vasculitis. Due to renal function deterioration the patient needed hemodyalisis for several days. Under continuous corticosteroid therapy, gradual recovery was noted and the patient was transferred to the medical ward. Four weeks later he was discharged with no gastrointestinal disturbances and improved renal function.

Patient 2

An 86 year old patient presented with a 3 day history of rectal bleeding with progressive weakness and dizziness. He had been diagnosed with Henoch-Schönlein purpura on a previous admission 1 week earlier, based on the findings of palpable

purpura, acute renal failure and leukocytoclastic vasculitis on skin biopsy. Treatment with oral corticosteroids was started.

On admission the patient was tachycardic and had blood pressure of 90/60 mmHg. Hemoglobin was 6.9 mg/dl. He was treated with four units of blood and stabilization of blood counts was noted. Upper gastrointestinal endoscopy was normal. Colonoscopy revealed patches of hyperemic and ecchymotic lesions of the cecum and terminal ileum. The patient was stable, the bleeding stopped, and he was discharged after observation. Two weeks later he returned to the hospital with the same complaints of rectal bleeding. A second colonoscopy was performed and revealed no apparent source of bleeding. Under treatment with blood transfusions his condition stabilized, his abdominal symptoms gradually subsided, hemoglobin was steady and bleeding stopped. Unfortunately, a few weeks later the patient developed a massive pneumonia followed by respiratory failure and arrest.

Comment

Although 50–85% of patients with Henoch-Schönlein purpura have some type of abdominal symptoms, it is rare that such patients are treated in the surgical ward or need surgical intervention. The two patients presented here were recently admitted to our service; one of them needed exploratory laparotomy for tenderness and CT findings that were suspicious for bowel ischemia. Both patients demonstrate well the spectrum of gastrointestinal symptoms in this disorder: small bowel ischemia due to vasculitis in the first patient and lower gastrointestinal bleeding in the second. Gastrointestinal symptoms in Henoch-Schönlein purpura patients usually develop within 8 days of the appearance of the typical rash but may precede the rash in as many as 15% of cases. The gastrointestinal manifestations range from mild vomiting, diarrhea, abdominal pain and transient paralytic ileus to more severe presentations as gastrointestinal hemorrhage, bowel ischemia and necrosis, intussusception and bowel perforation. Guaiac-positive stool is found in about 50% of patients, but life-threatening massive gastrointestinal hemorrhage is rare [3]. Abdominal symptoms associated with this disease are mainly caused by hemorrhage and edema within the bowel wall and mesentery. Henoch-Schönlein purpura in adults is generally not a surgical disease. Full-thickness involvement of the bowel wall and as a consequence bowel ischemia, necrosis or obstruction is extremely rare [4]. Therefore, surgical intervention is seldom necessary. Nevertheless, the clinical picture and imaging can be misleading, especially when the abdominal symptoms develop before or without the appearance of typical rash. Up to 10% of Henoch-Schönlein purpura patients undergo exploratory laparotomy because of either acute abdomen at presentation or a delay in diagnosis [5]. We believe that frequent clinical assessments with early involvement of the surgical team in order to evaluate the need or timing of surgical intervention, together with the surgeon's awareness of this rare disease and its pathophysiology will ensure the best outcome for Henoch-Schönlein purpura patients who suffer from major abdominal symptoms.

References

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