

Diagnostic challenge of intestinal obstruction secondary to Henoch–Schonlein purpura

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Henoch–Schonlein purpura is the most common form of vasculitis in pediatric age group. Gastrointestinal manifestations have been long recognized as part of the disease, but usually preceded by other manifestations. We present a case of a boy whose initial manifestation of the disease was bowel obstruction and ischemia requiring surgical intervention twice with diagnostic dilemma prior to diagnosis of Henoch–Schonlein purpura. Although common surgical diseases prove to be the culprit in most cases, a surgeon should always keep an open mind for possibility beyond surgical diseases. Multidisciplinary approach to situations where ambiguities in diagnosis exist proves to be a valuable tool aiding surgical

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Introduction

Henoch–Schonlein purpura (HSP) is the most common form of vasculitis in the pediatric age group. Gastrointestinal manifestations have been long recognized as part of the disease but usually preceded by other manifestations. We present a case of a boy whose initial manifestation of the disease was bowel obstruction and ischemia requiring surgical intervention twice with diagnostic dilemma before diagnosis of HSP.

Case report

A 6-year-old boy, previously healthy, presented to our Emergency Department with history of severe abdominal pain for 1 day. Pain was associated with bilious vomiting and obstipation. He had no previous history of similar illness. Physical examination showed dehydration, but otherwise vital signs were within normal limits. Abdominal examination was unremarkable apart from mild generalized tenderness on deep palpation. Abdominal radiograph did not show evidence of bowel obstruction, but because of persistence of symptoms an upper gastrointestinal contrast study was performed, which showed delayed gastric emptying and no midgut malrotation. Blood test showed mild leukocytosis ($14\,700/\text{mm}^3$) and platelet of $795 \times 10^9/\text{l}$. Chemistry panel was unremarkable. Patient underwent exploration laparoscopy and discovered enteroenteric intussusception in midbowel with no identified lead-point, which was reduced easily. Postoperatively, patient persisted to have no bowel motion, abdominal pain and distension, and bilious nasogastric tube aspirate. Abdominal computed tomography showed proximal bowel wall thickening suggestive of ischemia, with no recurrence of intussusception. On the fifth postoperative day, he was taken for exploratory laparotomy in which there was no mechanical bowel obstruction, but about 30 cm of proximal jejunum was ischemic. Resection and primary anastomosis was performed. Unfortunately, he had again no postoperative improvement and developed generalized abdominal

edema and started to have fever. Laboratory workup was unremarkable except low albumin and slightly increased immunoglobulin A (1.5 g/l). On the sixth day after the second exploration, he developed skin rash (hemorrhagic bullous lesions) mainly in the hands and feet (Fig. 1). When steroid therapy initiated (intravenous methylprednisolone), the abdominal pain, distension, and nasogastric aspirate started to improve gradually. Histopathology of the resected bowel came with evidence of small-vessel vasculitis with neutrophils extravasation (Fig. 2). Patient continued to improve on oral steroids and was discharged home on the 14th day postoperatively.

Discussion

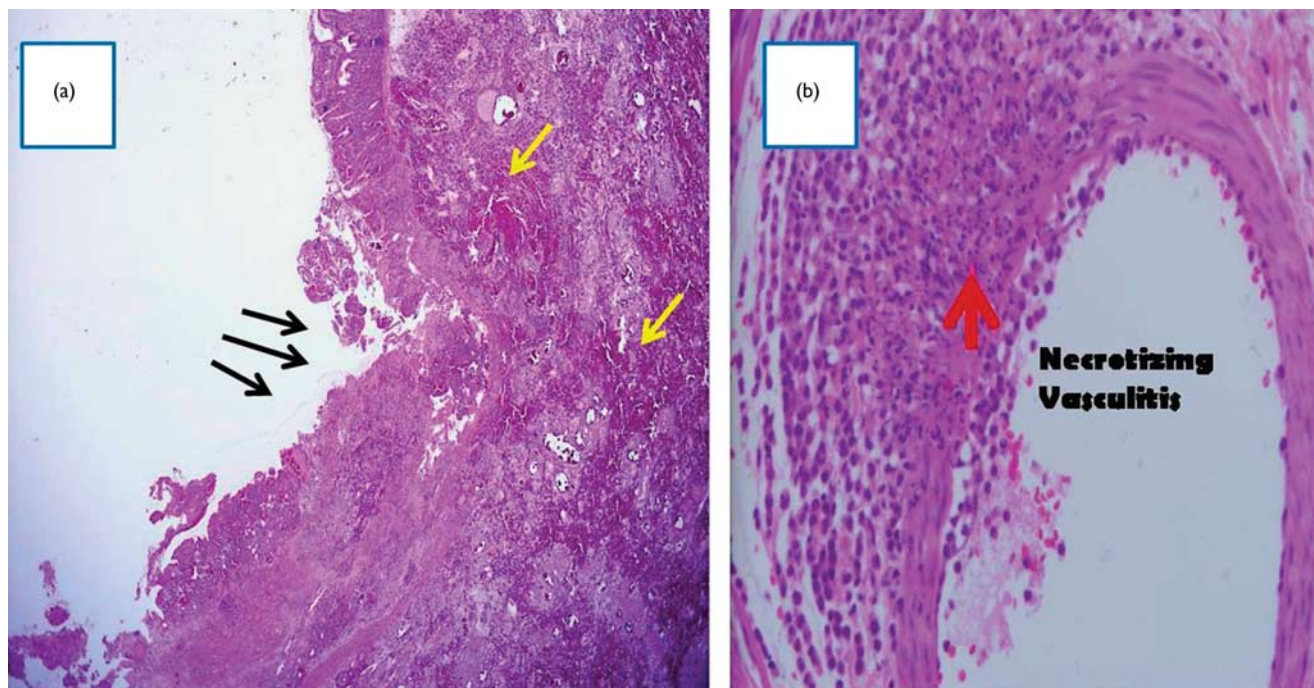
HSP is the most common form of vasculitis in children, with estimated annual incidence of 20.4 per 100 000 and the highest between the ages of 4 and 6 years (70.3/100 000) [1]. The exact cause is unclear, but it is recognized that HSP is an immune-mediated vasculitis associated with immunoglobulin

Fig. 1



Hyperpigmented hemorrhagic skin bullae at the feet.

Fig. 2



Pathology from the resected small bowel shows (a) congested small bowel wall (yellow arrows) and ischemic necrosis and ulceration of the mucosa, which was replaced by granulation tissue (black arrows) and (b) small vessels vasculitis with neutrophil extravasations.

A deposition. The clinical presentation of HSP is variable, with skin manifestations (purpura) being the most prominent feature and the initial manifestation in the majority of cases [2]. The American College of Rheumatology identified four criteria for the diagnosis of HSP: age of 20 years or below at disease onset, palpable purpura, acute abdominal pain, and biopsy showing granulocytes in the walls of small arterioles or venules. The presence of any two or more of these criteria distinguishes HSP from other forms of vasculitis with a sensitivity of 87.1% and a specificity of 87.7% [3].

Gastrointestinal manifestations have been reported to occur in about half of the patients with HSP [4] and can range from abdominal pain with nausea and vomiting to severe complications such as intussusception and bowel perforation. Gastrointestinal symptoms in previously undiagnosed HSP patients can be a challenge in diagnosis and management as in our case. Previous reports [4] showed that the most common symptom is abdominal pain (58%), followed by positive stool occult blood, and gross bloody stool. In this series, only one patient had intussusception, and one patient had bowel perforation. Intussusception is the most common complication requiring surgical attention with an estimated incidence of 3.5% [5]. It is postulated to be a result of bowel wall edema and hemorrhage acting as a lead-point, and it has been observed to be more in the small bowel (60%), in contrast to idiopathic intussusception that is mainly ileocolic. This fact is essential to the management of HSP-associated intussusception, which most likely will not respond to enema reduction in presence of a competent ileocecal valve, and ultimately will need surgical exploration. Other possible abdominal manifestations of HSP include acute appendicitis picture, bowel obstruction, paralytic ileus, bowel ischemia, sponta-

neous perforation, enteroenteric fistula, acute pancreatitis, pseudomembranous colitis, necrotizing cholecystitis, and ileal strictures [5].

Optimal management of HSP is not yet well established. Initial management aims at symptomatic relief with NSAIDs and supportive care. The use of steroids has been shown in a randomized, placebo-controlled trial to lower the risk for intussusception, although it was not statistically significant [6]. Concerns about masking symptoms of bowel perforation, along with insufficient data to support regular use, make steroid use unjustifiable in most cases.

Conclusion

Acute abdomen in the pediatric age group is always a challenge to the pediatric surgeon due to the variety of possible causes and the difficulty in most times to obtain informative history. In this case, the patient unusually presented with abdominal symptoms without any skin manifestation, making the diagnosis very difficult. Although common surgical diseases prove to be the culprit in most cases, a surgeon should always keep an open mind for possibility beyond surgical diseases. Multidisciplinary approach to situations where ambiguities in diagnosis exist proves to be a valuable tool aiding surgical decision.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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