A rare complication of Henoch-Schönlein Syndrome : gastrointestinal infarction and perforation

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To the editor,

We want to report an intestinal perforation with intestinal ischemia as a result of fatal complication of Henoch-Schönlein purpura (HSP).

A 39 year old male patient was admitted with 5 days history of abdominal pain, melena and vomiting. Abdominal pain was severe and colicky with vomiting. *The patient had respiratory tract infection 2 weeks before admission. He had also edema and pain in both ankles a week before admission. On the next day of admission petechial rashes emerged on posterior of both legs and in front of each tibias. Biopsy samples from rashes were taken.* Meanwhile, abdominal, knee and ankle pain worsened. *The first laboratory tests revealed the following results : white blood cells 20.4 × 1000/dl, haemoglobin 10.2 gr/dl, thrombocytes count 83 × 1000/L.* Abdominal examination disclosed diffuse tenderness and rebound tenderness. There was severe distension in the abdomen with reduced bowel sounds.

Vital findings were : body temperature : 38.2 C, blood pressure 94/48 mm/Hg, heart rate was 124/min. and respiratory rate was 28/min. Plain abdominal x ray showed free abdominal gas and bowel lumen extension. Abdominal CT scan demonstrated diffuse free liquid and thickness on the bowel wall (Fig. 1).

Explorative laparotomy was performed with the diagnosis of acute abdomen. During laparotomy diffuse free fluid was found in the abdomen. *There were wall thickness, ischemia and bowel edema over a length of 155 cm length in the terminal ileum. A procedure of small bowel resection with proximal end ileostomy was performed.* The patient was transferred to intensive care unit (ICU) for postoperative surveillance. *In ICU inotropic support and antibiotics with anaerobic effect were initiated because of sepsis. On the second postoperative day the patient was extubated.*

Skin biopsy revealed a leukocytic vasculitis with necrotic vessels containing neutrophils surrounded by fibrin strands. The histopathological analysis of resected ileum demonstrated ischemia, extensive mucosal ulceration with mild to moderate patchy infiltrate of acute and chronic inflammatory cells in lamina propria (Fig. 2).

The clinical and pathological findings of the patient were consistent with mesenteric ischemia secondary to HSP. Methyl prednisolone 1 mg/kg was started. *Clinical and laboratory values improved progressively and methylprednisolone was continued for 2 weeks. The patient recovered completely within 2 weeks and then referred to the ward.*

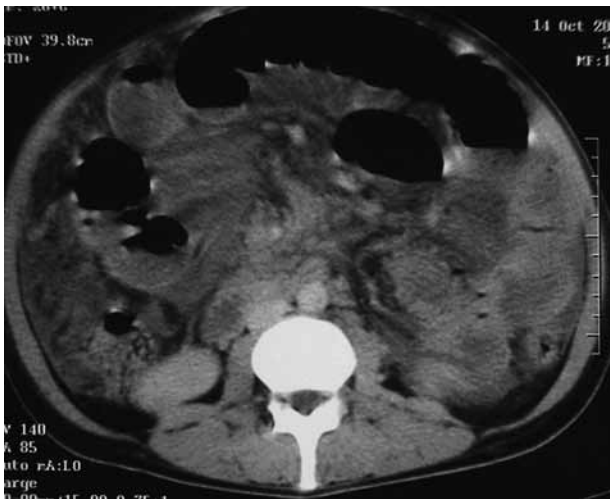

Fig. 1. — Contrast enhanced CT scan shows diffuse free liquid and thickness on the bowel wall.

HSP is a vasculitis characterised by palpable purpura, abdominal pain, arthralgia and hematuria. HSP is the most common acute vasculitic illness affecting children, but is relatively uncommon in adults, with reports occurring in patients up to 86 years age (1). The disease occurs most commonly in children between 3 and 10 years old : however in some series 30% of affected patients have been reported to be more than 20 years old. Gastrointestinal manifestations occur in up to 85% of HSP patients and are usually limited to abdominal pain and sometimes rectorhagia. Although gastrointestinal (GI) manifestations of HSP are common (50-75%), a few endoscopically described cases of terminal ileitis in adults exist (2). Small bowel perforation is an extremely rare complication of HSP ; it has been described in chil-

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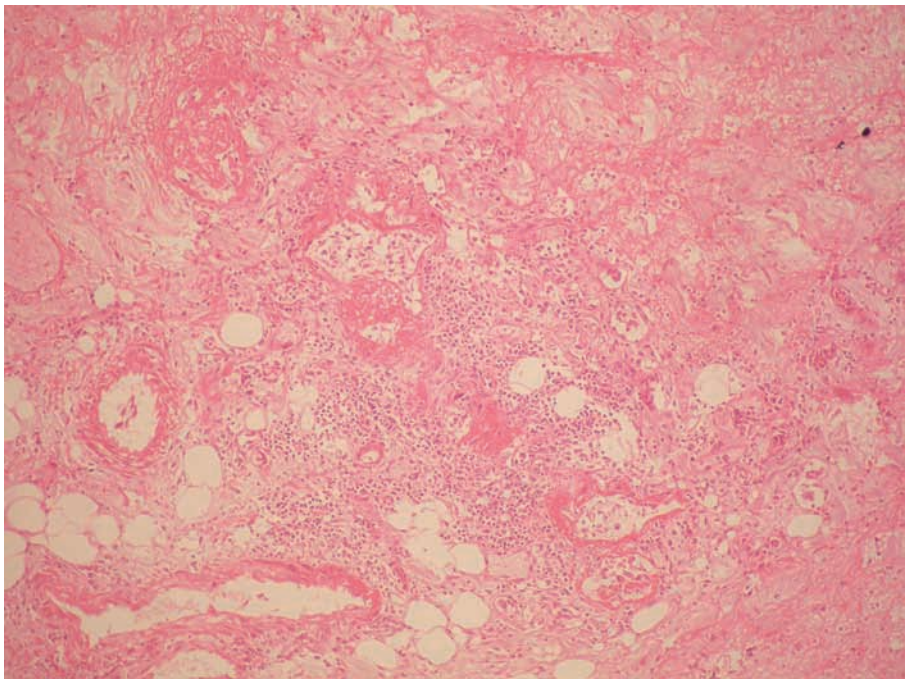

Fig. 2. — In the histopathologic assessment ischemia was present due to vasculitis in the small and middle vessels

dren, sometimes associated with intussusception and sometimes not (3). *A minority (2-6%) of patients with HSP requiring surgery, such as intussusception, perforation and infarction were reported* (4). Our patient experienced typical manifestations of HSP with palpable purpura, polyarthralgia and gastrointestinal manifestations. *In this report only the gastrointestinal manifestation with perforation is discussed*. This complication has been seen only very rarely. Gastrointestinal bleeding occurred more frequently in adults (59.1%) compared to children (28.3%, p : 0.01) (5). Gastrointestinal manifestations occur in 35-70% of cases. In most patients, the abdominal symptoms usually subside within a week, even without the use of corticosteroids. About 10% of patients may undergo laparotomy for bleeding, perforation, infarction, intussusception and obstruction (6). Many clinical experiences support the value of corticosteroid therapy in GI symptoms of HSP. There have been case reports suggesting that corticosteroid pulses may be helpful in patients with massive GI hemorrhage and widespread mesenteric vasculitis (7).

To the best of our knowledge this case describes one of the rare complications intestinal perforation due to HSP in an adult. So in conclusion, HSP is uncommonly seen in adults and can have serious GI involvement, such as small bowel infarction which may be fatal.

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