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Gastrointestinal Manifestations of Henoch-Schönlein Purpura

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We report a case of the intestinal lesion in Henoch-Schönlein purpura, presented with an acute abdomen in a 4 year old boy. Five days after sudden colicky abdominal pain, skin purpura and painful joint swelling developed. These manifestations were associated with abdominal distension, hematemesis, hematochezia and hematuria. Exploratory laparotomy revealed a marked bowel distension with edema and patchy dark reddish discoloration of the jejunum and ileum. These patchy areas showed transmural hemorrhage and necrosis associated with characteristic leukocytoclastic vasculitis in and around the hemorrhagic lesions. These vasculitis was thought to be related to Henoch-Schönlein purpura.

Key Words: *Henoch-Schönlein purpura, Leukocytoclastic vasculitis, Intestine, Hemorrhage*

INTRODUCTION

In the course of Henoch-Schönlein purpura, diverse gastrointestinal manifestations are seen up to 60% of patients. (Rodriquez-Erdmann & Levitan, 1968; Byrn et al, 1976; Agha et al, 1986). Abdominal pain is the well-known symptom and is usually colicky in nature. Diarrhea, nausea, and vomiting and actual gross or occult blood loss are often present. These symptoms vary in intensity and at times, emergency laparotomy has been performed for a suspected acute abdominal condition (Althansen et al, 1937; Allen et al, 1960).

Recently we have experienced such a case that the intestinal symptoms preceded the skin and renal lesions and emergency laparotomy was necessitated.

CASE REPORT

A 4-year-old boy presented with intermittent periumbilical and lower abdominal pain on May 5, 1989. Five days later, swelling of ankle joint with pain developed. Over the next several days, skin purpura appeared from the left fifth toe and progressed to lower and upper extremities, ear and buttock (Fig 1). He was managed with prednisolone and supportive care under the impression of Henoch-Schönlein purpura. However, above symptoms did not improve. Additionally, abdominal distension, hematemesis, hematochezia and hematuria were associated. On admission, blood pressure was 110/80 mmHg, heart rate 108/min, respiratory rate 24/min and body temperature was 36.7C. Laboratory studies revealed the following values; Leukocyte count, 18,400/cmm, hemoglobin, 12.8 gm/dl, hematocrit 35.8% and platelet count was 708,000/cmm; Prothrombin time was 12 second and 100%. Activated prothrombin time was 28 second. Initial urinalysis was normal. However, one month after the onset of the abdominal pain, urine albumin was +++ and blood ++++. Serum creatinine, 0.6mg/dl; and blood urea nitrogen was 7 mg/dl; Sodium 129 mmol/l, Potassium 3.7 mmol/l, Chloride 97 mmol/l. Ultrasonogram showed thickened folds of the small intestine as well as the mural thickening (Fig. 2). Endoscopy showed a diffuse hyperemia with hemorrhage and

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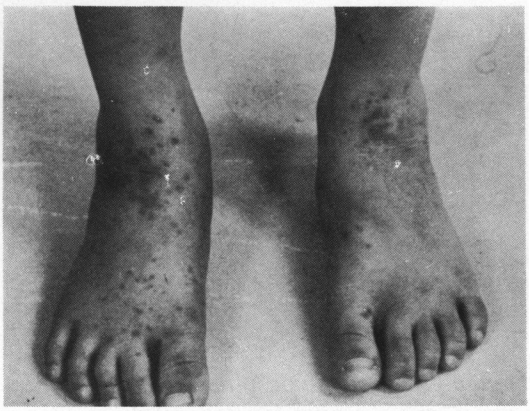

Fig. 1. Gross picture of the patient shows purpura of both lower legs, which represents extravasation of red blood cells into the dermis.

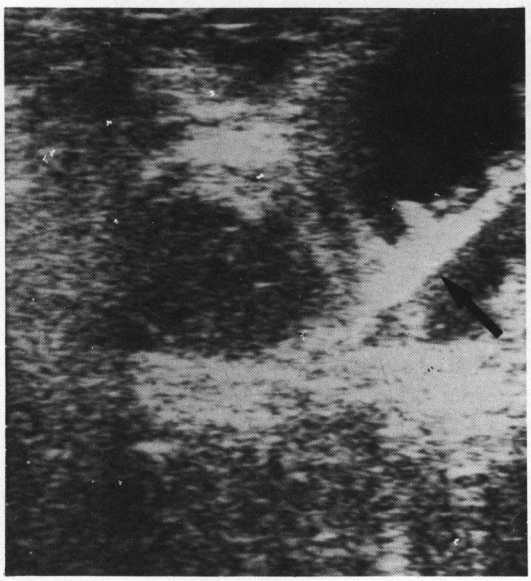

Fig. 2. Ultrasonogram reveals hyperechoic thickened intestinal wall and mucosal fold (arrow).

patchy erosions in the antrum of the stomach, duodenal bulb and most part of the rectum. During small bowel series, the barium did not pass through. At this point an emergency laparotomy was carried out under the impression of mechanical ileus. On operation, the jejunum and ileum showed marked distension and swelling with dark reddish discoloration and multiple hemorrhagic spots in the serosa. Involved segment measured 90cm in length, starting from a point 80cm distal to the Treitz ligament. Segmental resection of the lesion and anastomosis were performed. Grossly, the specimen showed hemorrhage and edema without

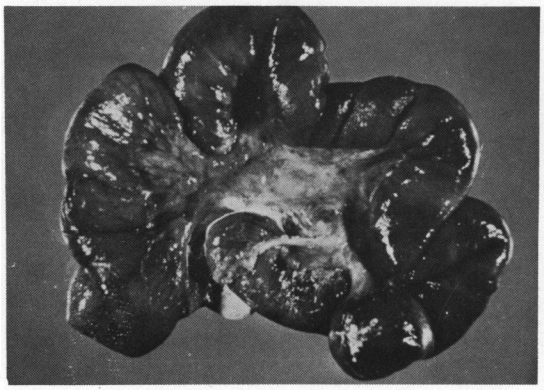

Fig. 3. Gross picture shows segmentaliy resected jejunum and ileum showing marked edema with dark red discoloration

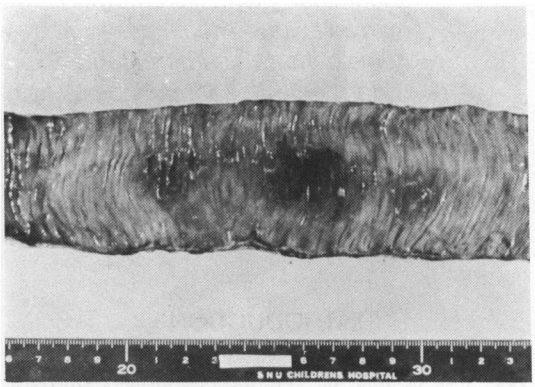

Fig. 4. The mucosa shows edema and focal erosion with patchy red discoloration.

skip lesion or obstruction (Fig. 3). The mucosa showed generalized edema with patchy hemorrhage and erosion (Fig. 4). Microscopically, the intestinal wall revealed transmural hemorrhage and segmental hemorrhagic necrosis (Fig. 5) with characteristic leukocytoclastic vasculitis (Fig. 6). The involved vessels were venules and arterioles. The affected vessels were located mainly in submucosa. Polymorphonuclear leukocytes with nuclear and cytoplasmic fragments were leaking from the vessel wall with radiating fashion (Fig. 7). On PTAH stain, the fibrin could be demonstrated from the involved vessel wall. This vasculitis was seen prominently in hemorrhagic areas. But this was also noted in fairly normal-looking viable intestinal wall around the lesions.

Postoperatively, gastrointestinal symptoms had much improved, but mild intermittent pain and tenderness persisted. Two weeks after operation, abdominal distension, severe swelling of the scrotum and facial edema developed. One month after the onset of skin

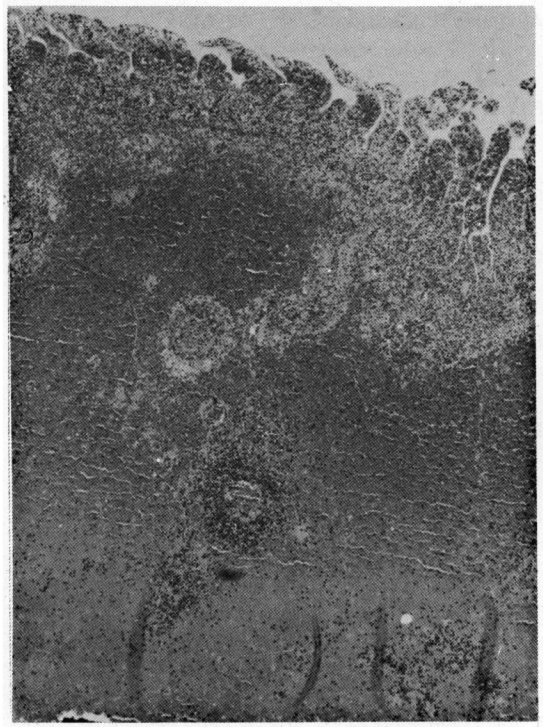

Fig. 5. Low power view of the small intestine shows transmural hemorrhage with necrotizing inflammation of the mucosa. (H&E. X40)

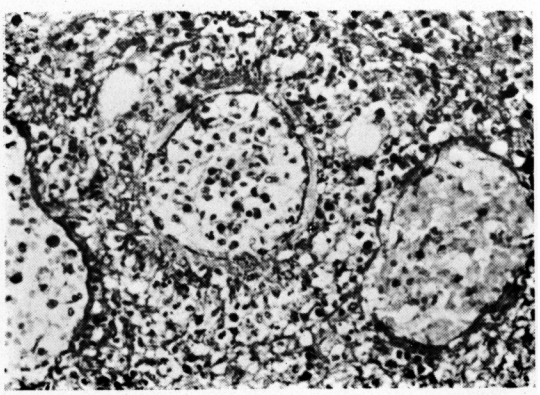

Fig. 6. Submucosal arterioles show leukocytoclastic vasculitis and fragmentation of internal elastic lamina. (Elastic stain, ×200)

purpura, the skin lesion subsided. However, microscopic hematuria (RBC 5-7/HPF) has persisted for the subsequent 6 months.

DISCUSSION

Characteristic leukocytoclastic vasculitis (LCV) found in bowel specimen in this case made us consider

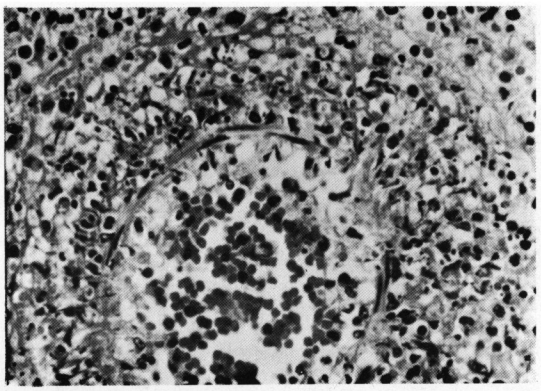

Fig. 7. High power view of a arterioles shows polymorphonuclear leukocyte infiltration in the vessel wall with nuclear and cytoplasmic debris with radiating fashion. (H&E, X400)

Henoch-Schönlein purpura (HSP) as the first differential diagnosis. Although the chance to see gastrointestinal lesion of the HSP microscopically is fairly limited, this phenomenon is not rare. In the course of HSP, diverse gastrointestinal manifestations are common and mentioned to occur in from 35% to over 60% of the patients (Allen et al) 1960; Feldt & Stickler, 1961). Lopez et al (1980) in a review of 93 cases with LCV reported that gastrointestinal manifestations were much more common in patients younger than 16 year of age than in older patients. Although small bowel is more frequently involved, cases of esophageal, gastroduodenal, and colorectal involvement have been rarely reported in HSP (Handel & Schwartz, 1957; Allen et al, 1960; Winkleman & Ditto, 1964; Rodriguez-Erdmann & Levitan, 1968; Glasier et al, 1981). Clinically gastrointestinal tract involvement may manifest as colicky abdominal pain, vomiting, melena, and hematemesis. According to Allen et al. (1960) abdominal pain was the commonest symptom, being present in over one half of all patients and in a almost 80% of those with gastrointestinal symptoms. The next commonest GI symptom was gross melena, which occurred in over one-half of patients. Hematemesis occurred in about 1/10 of 131 patients of their review.

It is not clear why in some cases like our case the intestine is involved before the classic skin or renal lesion. Since HSP is a systemic disease and characterized by vasculitis it is understandable that the initial site of involvement can vary by different case. This seems to be the main reason why unnecessary laparotomy could be carried out. The indication of exploratory laparotomy in HSP could be intussusception, perforation or gastrointestinal bleeding. In our case it was the suspicion of intestinal obstruction because no barium could be passed through. However, there was

no true obstruction found at operation. It could have been due to massive edema and hemorrhage of the intestinal wall. Actually intussusception, bowel obstruction, perforation and ischemic necrosis have all been reported in patients with HSP. The exploratory laparotomy in our case can be well justified by finding segmental hemorrhagic necrosis with vasculitis found at pathological examination. These involved segment would most probably have been complicated by perforation if not managed surgically.

Allthough Agha et al (1986) described weak granular IgA deposition in the vessel wall of the involved segment, we could not study the immunologic abnormalities in this case because of unavailability of the fresh specimen. However, our case could reasonably be diagnosed as intestinal HSP by characteristic leukocytoclastic vasculitis, segmental involvement, and transmural hemorrhage along with clinical presentation. The hemorrhage into the intestinal wall appears to be due to diapedesis of the red blood cells from the arterioles and venules that are extremely dilated and often necrotic. Although characteristic radiological findings of thumb print or picket fence pattern (Whitmore & Peterson, 1946; Esposto, 1950; Handel & Schwartz, 1957; Schwartz et el, 1966; Glasier et al 1981) was not found in our case, thickening of the wall and thickened mucosal folds could be seen on ultrasonography.

Postoperative scrotal swelling in this case could have been due to scrotal involvement which occurs 2-38% of the cases (Sahn & Schwartz, 1972; Clack & Kramer, 1986). Other than gastrointestinal tract, kidney, scrotum and lung could also be the site of vascular involvement in HSP (Agha et al, 1986; Clark & Kramer, 1986).

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