Leucocytoclastic Vasculitis (Hypersensitivity Angiitis) of the Small Bowel Presenting with Severe Gastrointestinal Hemorrhage

Farooq P. Agha, M.D., F.A.C.G., Timothy T. Nostrand, M.D., and David F. Keren, M.D.

Department of Radiology, Internal Medicine-Division of Gastroenterology, and Pathology, University of Michigan Hospitals and Medical Center, Ann Arbor, Michigan

A case of leucocytoclastic vasculitis involving the entire small bowel is reported. A high index of suspicion and recognition of the early palpable purpuric skin lesions in patients with acute abdominal pain and gastrointestinal hemorrhage might avert unnecessary surgical exploration in some patients.

INTRODUCTION

Leucocytoclastic vasculitis (LCV) and Henoch-Schonlein purpura (HSP) are syndromes characterized by vasculitis of the postcapillary venules and "palpable purpura" as the clinical hallmark (1–5). Gastrointestinal involvement associated with vasculitis has been well recognized in children with HSP (7–11) and in patients with polyarteritis nodosa (12–14), but information regarding diagnosis and management of leucocytoclastic vasculitis with gastrointestinal involvement is limited to few reports (1, 15–17). This report illustrates a case of LCV with severe gastrointestinal hemorrhage and emphasizes the systemic nature of this disease and urges restraint before performing exploratory surgery on such patients.

CASE REPORT

A 35-yr-old white man sustained 10% burns of his right upper extremity after a firework accident on July 4, 1984. He underwent partial and full thickness skin grafts at a local hospital. He was recovering well until July 24th, when he developed crampy abdominal pain associated with nausea, vomiting, and passage of melena stools per rectum. He was transferred to the University of Michigan Hospitals for management.

At the time of admission to the University Hospital his vital signs were within normal limits except for a resting tachycardia of 116/min without orthostatic changes. The physical examination including auscultation of the chest was unremarkable. The abdominal examination revealed mild periumbilical tenderness without rebound or rigidity. The rectal examination revealed melena and no other abnormalities.

The admission laboratory findings revealed Hb of 16.6 g, hematocrit 50.4%, white blood cell count of 24,300/mm³ with a slight left shift, and normal platelet count. The blood urea nitrogen was 11, creatinine 0.9, and the serum electrolytes were normal. The protein electrophoresis showed mild hypogammaglobulinemia and antinuclear antibody screening was negative. His urinalysis showed no evidence of proteinuria.

Plain abdominal radiographs revealed an abnormal loop of small bowel in the left side of the abdomen showing thickened mucosa and thumbprinting (Fig. 1A), scattered gas in the small bowel, and minimal distension of the transverse colon. Upper endoscopy showed no significant abnormality in the esophagus, stomach, and duodenum. Flexible sigmoidoscopy was normal up to 40 cm. A small bowel follow through examination revealed marked submucosal edema of the jejunum and proximal small bowel (Fig. 1B). The abdominal pain persisted with intermittent melena stools. Visceral angiography was performed on the 3rd day after admission which showed normal mesenteric vasculature. Twenty-four hours after angiography his abdominal pain became constant and increased in intensity. Repeat stool examination revealed bright red blood. The skin examination at this time revealed two petechiae and purpuric spots on his lower extremities. His Hb dropped to 10.5 g and the hematocrit to 48%. The white blood count increased from 24,300/mm³ to 38,700/mm³. Bowel ischemia with possible infarction was suspected and the patient underwent exploratory laparotomy on the 4th day after admission. At surgery the entire small bowel was inflammed with numerous scattered patches of petechial hemorrhages on the serosal surface. These changes were more severe in the jejunum and proximal small bowel. No mechanical obstruction or mass lesion was found. The colon was normal and the remainder of the abdominal exploration was unremarkable. The bowel was viable and no
FIG. 1. A, plain abdominal radiograph shows a thickened edematous loop of small bowel in the left upper quadrant with "thumbprinting" indicating submucosal infiltration (arrows). B, a small bowel examination shows mucosal thickening and edematous loops of jejunum and proximal small bowel with no evidence of obstruction or mass lesion.

resection was done. Biopsies of the small bowel and purpuric lesions from his lower extremity were obtained. He was treated with intravenous hydrocortisone intraoperatively and postoperatively. The histopathological examination of the biopsied material revealed LCV affecting the small bowel and the skin lesions (Fig. 2). Direct immunofluorescence examination of the purpuric skin lesions and the bowel using monospecific-fluorescein isothiocyanate-conjugated antihuman IgG, IgA, IgM, C3, and fibrin showed weak granular IgA deposition in the small vessels along with intense fibrin deposition. There was no deposition of IgG, IgM, or C3. The findings were consistent with immune-complex vasculitis (leucocytoclastic). He was treated with steroids which were gradually tapered and he was discharged on the 12th day of hospitalization. At 1-yr follow-up there is no recurrence of vasculitis.

DISCUSSION

LCV is a pathological designation characterizing those conditions in which the blood vessel walls are infiltrated by polymorphonuclear leucocytes and nuclear fragments in varying stages of necrosis (1). The postcapillary venule is the usual site for this lesion (2). There is reasonable evidence that this inflammatory process is initiated by the subendothelial deposition of antigen-antibody complexes which are derived from the circulation. These immune complexes activate the complement system, releasing chemotactic factors which attract polymorphonuclear leucocytes. Proteolytic enzymes are then released and mediate the vascular necrosis (3, 4).

LCV or necrotizing vasculitis consists of several distinct clinical syndromes affecting small vessels less than 0.1 mm in diameter, which are characterized by immune-complex deposition of IgA in the walls of the affected vessels and some form of complement activation. Hypersensitivity angiitis, allergic vasculitis, cutaneous systemic vasculitis, and hemorrhagic capillary toxicosis are other terms used for this syndrome complex. Identical pathological changes are seen in anaphylactoid or Henoch-Schönlein purpura, hypocomplementemic vasculitis, and essential mixed cryoglobulinemia (4). Skin involvement is very common and is often the only site affected by characteristic leucocytoclastic vasculitis. The skin lesions first appear on the lower extremities as flat erythematous or urticarial patches progressing to purpuric papules and are mostly concentrated on dependent areas. The palpable quality of these lesions is an important feature since this distinguishes them from other forms of purpura (4). In addition to the characteristic purpuric papules, hemorrhagic bullae and irregular superficial erosions covered by an eschar may be seen. The disease may be limited to the skin or there may be systemic involvement of
joints, kidneys, lungs, and the gastrointestinal tract. This type of vasculitis may be precipitated by a drug or a microorganism, but often the specific antigen which triggers the process remains unidentified.

Lopez et al. (5) in a review of 93 cases with LCV reported that 32 (34%) had significant gastrointestinal manifestations. They further noted that gastrointestinal manifestations were much more common in patients younger than 16 yr of age (66%) than in older patients (26%). Gastrointestinal tract involvement by LCV may manifest as abdominal pain that is usually colicky in nature. Nausea, vomiting, and diarrhea are often present. Mild to moderate gastrointestinal bleeding has been reported in 52% of the cases and 21% of patients may present with acute abdomen (5).

HSP is established as a distinct subtype of LCV. It may involve any age group, but the majority of patients are children with a peak incidence from 4 to 8 yr of age (8-11). Although the small bowel is more frequently involved, cases of esophageal, gastroduodenal, and colorectal involvement has been rarely reported in HSP. Abdominal complications such as intussusception, bowel obstruction, perforation and ischemic necrosis have been reported in patients with HSP. Although gastrointestinal tract involvement is found in over 50% of children with HSP, only few reports describe the gastrointestinal manifestations associated with LCV or HSP in adults. Information regarding diagnosis and medical management of gastrointestinal vasculitis syndromes in adults is thus limited (1, 7, 15, 16).

The demonstration of weak granular IgA and intense fibrin deposition in the small vessels in our patient is consistent with an immunocomplex vasculitis (4-6). The source of this antigenic stimulation is unknown. These patients usually present difficult diagnostic dilemma. The clinical history is not helpful at all. Barium studies of the gastrointestinal tract may show a spectrum of nonspecific features ranging from mucosal edema to typical thumbprinting. Occasionally radiographic changes may simulate Crohn’s disease or acute

**Fig. 2.** A, this photomicrograph of the small bowel biopsy from the same patient demonstrates passing through the muscularis mucosa (M) a vessel (V) which in the submucosa becomes distorted (arrows) by acute inflammation with prominent fibrin deposition (F) (hematoxylin and eosin, original magnification ×470). B, this photomicrograph of the skin biopsy from the present case discloses a classic leukocytoclastic vasculitis with a vessel (arrow) being infiltrated by acute inflammatory cells and showing prominent endothelial proliferation that obscures the lumen (hematoxylin and eosin original, magnification ×470).
ileitis. Endoscopy of the upper or lower intestinal tract may reveal submucosal hemorrhage with or without focal vasculitis. Recently computed tomographic appearance of segmental mural thickening and luminal narrowing in LCV correlating well with the barium small bowel studies and endoscopic findings have been reported (18). Endoscopic biopsies may reveal mucosal necrosis or small vessel involvement in the lamina propria. Although the temporal association of palpable purpura to gastrointestinal manifestations in LCV is variable, it is advisable that adults with gastrointestinal bleeding without obvious source be examined for palpable purpura of LCV. This may detect potential cases of LCV and would allow steroid therapy to produce resolution of symptoms and thereby avoiding unnecessary surgery.

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Reprint requests: Dr. Farooq P. Agha, Department of Radiology—Box 013, University Hospital, 1405 East Ann Street, Ann Arbor, MI 48109.

REFERENCES

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