Ischemic Colitis Associated with Systemic Lupus Erythematosus

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A case of colonic ischemia, infarction, and perforation secondary to systemic lupus erythematosus (SLE) is described in a 37-year-old woman. The incidence and significance of gastrointestinal complications in SLE are discussed. [Key words: Systemic lupus erythematosus, complications; Colitis, ischemia; Vasculitis, colon; Blood vessels, inflammation]

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THE GASTROINTESTINAL TRACT is a frequent, yet often unrecognized, site of complications in patients with systemic lupus erythematosus (SLE).[1,2] Greater than one-half of patients will present with abdominal pain, nausea, and vomiting during the course of the disease and these symptoms are usually attributed to uremia, central nervous system involvement, gastroenteritis, secondary infection, or the side effects of medication.[3] Mesenteric ischemia and infarction are often not considered because the abdominal findings are generally mild or nonexistent even in the face of an abdominal catastrophe. We describe such a case in a 37-year-old woman with longstanding SLE and immunosuppression, who remained remarkably asymptomatic despite ischemic colitis which eventually resulted in stricture, infarction, and perforation. The incidence, clinical spectrum, and radiographic manifestations of mesenteric vasculitis in SLE are reviewed.

Report of a Case

A 37-year-old white woman with a 12-year history of SLE presented with a three-day history of nausea, vomiting, midabdominal pain, and diarrhea. She denied hematemesis, melena, hematochezia, fever, or chills. Physical examination results revealed a macular rash on the left thigh, telangiectasis on the neck, a small skin ulcer on the left leg, and a soft, nondistended abdomen with diminished bowel sounds. The patient complained of diffuse tenderness but no rebound tenderness was elicited and stools were guaiac negative. Admitting laboratory values revealed a positive ANA at a dilution of 1:1280, depressed complement values, white blood cell count of 4,100 with normal differential, and hemoglobin level of 12.3 g/dl.

Pertinent past medical history included Raynaud's phenomenon, arthralgia, telangiectasis, pleurisy, and multiple cutaneous ulcers of the lower extremities. The disease, however, was relatively quiescent at the time of admission except for the cutaneous manifestations. There was no history of renal or gastrointestinal involvement.

Plain abdominal films were normal and an air-contrast barium-enema examination (Fig. 1) demonstrated a 12-cm segment of mucosal ulceration and irregularity of the midtransverse colon. Colonoscopy confirmed these findings; however, the mucosa was judged too friable to safely permit biopsy. The diagnosis of lupus vasculitis producing ischemic colitis of the colon was made and the patient was discharged on 60 mg of prednisone per day.

The patient did well after discharge and the corticosteroids administration were gradually tapered. The patient presented six weeks later with crampy abdominal pain, nausea, vomiting, and guaiac-positive loose stools. Bowel sounds were decreased and there was moderate tenderness to palpation across the lower abdomen. Laboratory values and vital signs were normal except for a white blood cell count of 5,700 with 73% polymorphonuclear cells. Plain abdominal films showed a dilated right colon and small bowel with no free air. Film of a water-soluble enema (Fig. 2) demonstrated a 3-cm narrowed segment of midtransverse colon, measuring 1 cm in greatest diameter, with proximal dilation. The proximal colon demonstrated "thumbprinting" and there was thickening of the valvulae conniventes of the distal ileum. At laporotomy, a 1.2-cm perforation of the cecum was found and focal necrosis and ulceration without perforation were noted in the transverse colon. The distal ileum, cecum, ascending and transverse colon were resected and an ileostomy and mucous fistula created. Pathologically, a fibrinous peritonitis, mesenteric arteritis with organizing thrombus, and acute and chronic vasculitis of the subserosal vessels were noted involving the right and transverse colon.

The patient was discharged three weeks after surgery with future plans to take down the ileostomy.

Discussion

Systemic lupus erythematosus is characterized by the deposition of circulating immune complexes which produce a necrotizing vasculitis of small arteries, arterioles, and venules throughout the body.[4,5] While the renal and central nervous system manifestations of SLE are well known, mesenteric involvement is also quite common but less well appreciated. Inflammation of the small blood vessels of the gut produces a spectrum of complications including intestinal ischemia, hemorrhage, ileus, ulceration, infarction, and perforation.[6] Serous peritonitis, pancreatitis, malabsorption, protein-losing enteropathy, and lower esophageal aperistalsis associated with Raynaud's phenomenon are other com-

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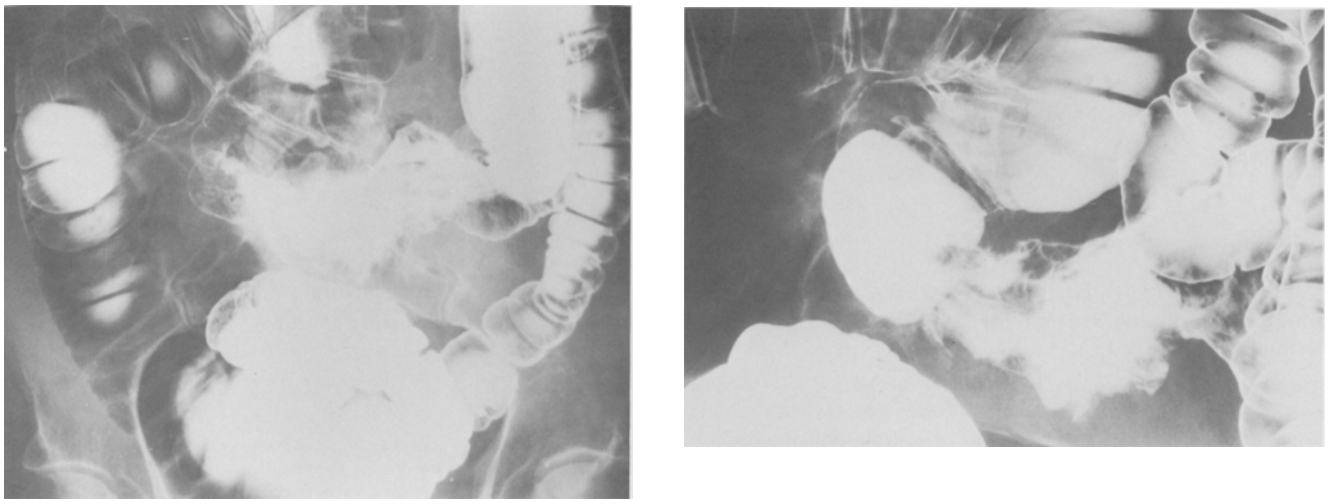

FIG. 1. Air-contrast examination film (left) of the colon demonstrates diffuse ulceration of the midtransverse colon with the remainder of the colon appearing normal. Spot film (right) of midtransverse colon showing deep ulcerations.

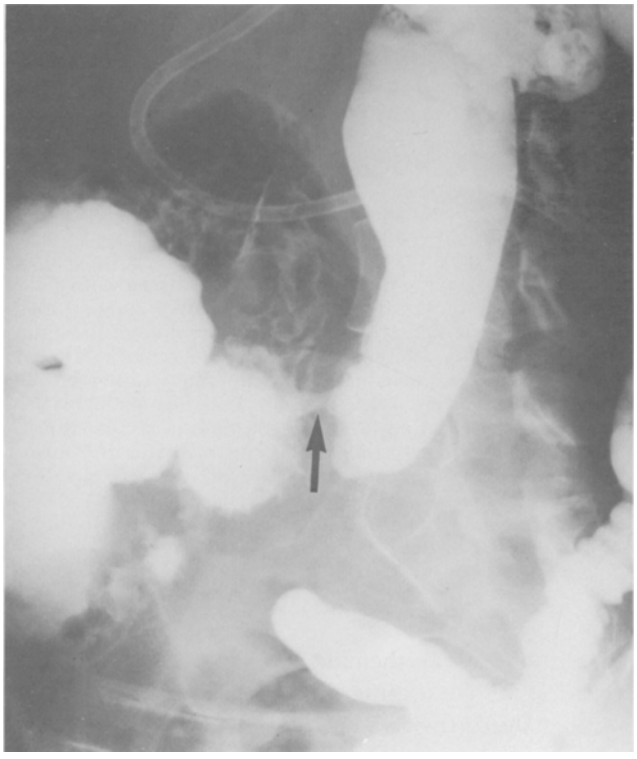

FIG. 2. Left posterior oblique film of hypaque enema obtained 6 weeks later shows stricture of midtransverse colon (arrow) and "thumbprinting" of the colon proximally, indicating ischemia.

plications related in varying degree to ischemia.[3] Several of these complications are potentially lethal because patients with SLE are often on high doses of corticosteroids and azathioprine therapy which mask abdominal symptomatology. Peptic ulcer disease and hepatic dysfunction may also occur as a result of the toxic effects of these medications.[7]

Clinically, patients with the intestinal manifestations of SLE present with a variety of signs and symptoms which depend upon the degree of ischemia and immunosuppression. This is further complicated by the fact that abdominal symptoms may also be produced by uremia, secondary infections, and central nervous system involvement. Prompt and intensive radiographic evaluation is essential to differentiate the potential causes of abdominal distress.

Plain abdominal radiographs may show only an ileus or pseudo-obstruction pattern which may be due to ischemia, sepsis, or uremia. Edematous haustra and valvulae conniventes are seen with progression of ischemia.[8,9] Pneumatosis intestinalis may be seen as a result of necrotic bowel[10] or may be secondary to air working through defects in the mucosa and altered connective tissue support of the gut, in the absence of intestinal ischemia.[11] Free intraperitoneal air may loculate in patients with perforation because of the peritoneal serositis and adhesions so common in this disease. In the absence of specific plain film findings, contrast studies are necessary to further elucidate difficult clinical problems.

Double-contrast upper gastrointestinal examination results may show superficial erosions in the stomach and duodenum secondary to steroids. Immobility of the distal esophagus may also be seen in patients with concomitant Raynaud's phenomenon as in scleroderma. Small-bowel series may demonstrate spasm, thickening of the valvulae conniventes, or a malabsorption pattern due to ischemia. These small-bowel changes can be reversed in some situations with intensive antibiotic and corticosteroid therapy.[8,11,12]

In the presence of ischemia, film of barium-enema examinations often demonstrates "thumbprinting" or

indentation of the barium column by submucosal hemorrhage and edema. This may progress to ulceration which is at first superficial but can deepen, producing craters which penetrate the submucosa and may eventually perforate.[9] The bowel may heal completely or result in a stricture. Often a stricture leads to infarction and perforation because the mucosal blood supply is attenuated by dilatation and mural stretching proximal to the narrowed segment of colon.[9] This may have been an important factor in our patient. Most patients with SLE demonstrate ischemic changes in the splenic flexure, a region sensitive to decreased blood supply because it is the watershed area between the superior and inferior mesenteric artery circulations.

Laboratory findings in patients with SLE and mesenteric ischemia are often nonspecific. Elevation of the white blood cell count and changes in vital signs are unreliable indicators of disease. The only consistent laboratory findings are related to active lupus such as elevated sedimentation rate, decreased serum complement, and positive ANA.[3]

Management of the abdominal manifestations of SLE is difficult. In the absence of compelling radiographic or clinical findings suggestive of infarction or perforation, intensive corticosteroid, antibiotic, and fluid therapy is indicated since these patients do not tolerate surgery well and have a propensity to form adhesions. As stated before, some lesions may resolve or regress or, as in our patient, progress to stricture and infarction. Predicting which course a certain individual patient may take is impossible. Unfortunately, there is no simple gauge to assess the extent and degree of intestinal ischemia comparable to BUN and creatinine levels in evaluating renal lupus. Close surveillance and a high degree of clinical suspicion are necessary in these patients.

References

1. Stoddard CJ, Kay PH, Simms JM, Kennedy A, Hughes P. Acute abdominal complications of systemic lupus erythematosus. Br J Surg 1978;65:625–8.
2. Jacobs AJ, Luthra HS. Intra-abdominal complications of systemic lupus erythematosus. Minn Med 1982;65:183–4.
3. Hoffman BI, Katz WA. The gastrointestinal manifestation of systemic lupus erythematosus: a review of the literature. Semin Arthritis Rheum 1980;9:237–47.
4. Kleinman P, Meyers MA, Abbott G. Necrotizing enterocolitis with pneumatosis intestinalis in systemic lupus erythematosus and polyarteritis. Radiology 1976;121:595–8.
5. Weiser MM, Andres GA, Brentjens JR, Evans JT, Reichlin M. Systemic lupus erythematosus and intestinal venulitis. Gastroenterology 1981;81:570–9.
6. Kistin MG, Kaplan MM, Harrington JT. Diffuse ischemic colitis associated with systemic lupus erythematosus - response to subtotal colectomy. Gastroenterology 1978;75:1147–51.
7. Runyun BA, LaBrecque DR, Anuras S. The spectrum of liver disease in systemic lupus erythematosus. Am J Med 1980;69: 187–94.
8. Shapeero LG, Myers A, Oberkircher PE, Miller WT. Acute reversible lupus vasculitis of the gastrointestinal tract. Radiology 1974; 112:569–74.
9. Gore RM, Calenoff L, Rogers LF. Roentgenographic manifestations of ischemic colitis. JAMA 1979;241:1171–3.
10. Meyers MA, Ghahremani GG, Clements JL, Goodman K. Pneumatosis intestinalis. Gastrointest Radiol 1977;2:91–105.
11. Freiman D, Chon H, Bilaniuk L. Pneumatosis intestinalis in systemic lupus erythematosus. Radiology 1975;116:563–4.
12. Train JS, Hertz I, Cohen BA, Samach M. Lupus vasculitis: Reversal of radiographic findings after steroid therapy. Am J Gastroenterol 1981;76:460–3.