Spontaneous Perforation of the Small Intestine Due to Scleroderma

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Several comprehensive studies have discussed the various intestinal manifestations of scleroderma (1-4). Although 50% of patients with scleroderma may have small bowel involvement (3, 4), spontaneous perforation of the small intestine has been reported only recently (5). We report a second documented case of spontaneous small-bowel perforation due to sclerodermatous involvement of the bowel wall to further emphasize this rare but potential complication which can be initially mistaken for pseudo-obstruction and cause a delay in definitive treatment.

CASE REPORT

A 42-year-old black woman presented to the Hospital of the University of Pennsylvania with hypopigmentation of face and chest. She had noted progressive skin tightness over the fingers and perioral areas, arthralgias, heartburn, and Raynaud’s phenomenon for the past two years. Skin biopsy was consistent with scleroderma. Esophageal manometrics showed a hypotensive lower esophageal sphincter and normal peristalsis. Pulmonary function tests revealed mild restrictive disease. LE prep, ANA, and rheumatoid factor were negative.

Subsequently, she was admitted with a 3-day history of crampy abdominal pain, nausea, vomiting, chills, and temperature of 101°F. She had noted also a 30-pound weight loss over the last year. Physical examination was remarkable for dehydration, scattered areas of hypopigmentation, sclerodactyly, and taut facial skin with decreased mouth aperture. The abdomen was distended, mildly tender with hypoactive bowel sounds. Stool was positive for occult blood.

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Pertinent laboratory data were: hemoglobin, 12.5 gm/dl; hematocrit, 40%; WBC, 13,500/mm³ with 53% polymorphonuclears, 22% bands, and 25% lymphocytes; BUN, 36; and creatinine, 1.4 mg/dl; amylase, 243 Somogyi U/dl; LDH, 498 IU/liter; and normal bilirubin, aspartate aminotransferase (AST), alanine aminotransferase (ALT), and alkaline phosphatase. Obstruction series showed dilated loops of small bowel and air fluid levels. Chest x-ray was normal.

Treatment was initiated with intravenous fluids and nasogastric suction for presumed pseudoobstruction. Daily temperature spikes occurred and cephalothin and gentamicin were begun. Clindamycin was later added because Bacteroides fragilis was isolated from blood cultures obtained on admission. On the fifth hospital day, a Miller-Abbott tube was passed because of persistent dilated small-bowel loops on x-ray. The tube remained in the proximal small bowel over several days due to poor peristalsis and finally on the ninth day of hospitalization appeared to be in the terminal ileal region. Clinically she improved but continued with nocturnal temperature elevation to 101°F. Gallium scan showed a persistent area of increased uptake in the pelvic region. Proctoscopy was normal to 14 cm. Barium enema revealed a normal colon without wide-mouthed diverticula. Reflux of barium into the terminal ileum demonstrated the tip of the Miller-Abbott tube to be lying in an extraluminal space which was thought to represent an abscess in free communication with the terminal ileum. This was verified by injecting barium through the Miller-Abbott tube (Figure 1).

On the thirteenth hospital day, laparotomy was performed. A perforation of the terminal ileum and a necrotic pelvic abscess cavity was found. The surrounding bowel was viable. A segmental ileal resection with entero-enterostomy was performed as well as drainage of the abscess. Pathologic findings of the resected bowel were consistent with scleroderma and included diffuse collagenous replacement of the muscularis mucosa, submucosa and serosa with perivascular fibrosis and diffuse chronic lymphocytic infiltration (Figure 2). Scattered mucosal ulcerations were noted. The perforation was identified in an area of total fibrous replacement of the muscle layer (Figure 3). The serosa was thickened with fibrous adhesions and new vessel formation along with chronic inflammatory reaction. No evidence of vasculitis was found.
PERFORATION DUE TO SCLERODERMA

Fig 1. Barium and air injected through a Miller-Abbott tube fill an abscess cavity in the pelvis (arrows). Some of the barium has escaped into the terminal ileum via the connection with the abscess and is seen filling the cecum.

Her postoperative course was uneventful. One year after discharge, she has remained well and has gained 30 pounds. Outpatient studies to assess malabsorption were normal, including radiographic studies of the stomach and small bowel.

Serologic data included a negative LE prep, positive ANA to 1:40 dilution, and normal serum complement. Antibody to extractable nuclear antigen (ENA) was positive in low titer (1:80). Nuclear ribonucleoprotein (RNP) and Sm antibodies were negative.

DISCUSSION

Small-intestinal involvement is commonly seen in scleroderma (3, 4). The classic pathologic findings include muscular atrophy, increased connective tissue with fibrosis of the muscularis, submucosa, and serosa along with lymphocytic infiltration (6). Dilatation of the duodenum and jejunum are the most common radiographic abnormalities and malabsorption and pseudoobstruction are commonly seen clinical complications (4). Myoelectric studies have suggested that small-bowel motor dysfunction in patients with scleroderma may be the result of diminished excitability of the bowel to mechanical and hormonal stimuli (7). Small-intestinal involvement may occasionally be the outstanding feature of scleroderma; however, there has been little correlation between its presence and the overall severity of the disease (3).

Infarction and perforation of the small bowel in association with vasculitis occurs in patients with systemic lupus erythematosus (SLE) and polyarteritis (8). Peritonitis associated with infarcted terminal ileum and ascending colon due to thrombosis of the celiac and superior mesenteric arteries as well as massive intestinal infarction on a nonocclusive basis have been reported in scleroderma (8, 9). The ileal perforation and intra-