Pneumatosis Intestinalis in Limited Scleroderma

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A 43-year-old Native American woman presented to the hospital with a 1-year history of diarrhea and 100-pound weight loss. A diagnosis of limited scleroderma was made based on a history of Raynaud's phenomenon, calcinosis cutis of forearms and legs, and cutaneous telangiectasias. Autoimmune serologies included a positive antinuclear antibody and positive anticentromere antibody. She had prolonged gastric emptying and small-bowel transit studies indicative of moderate

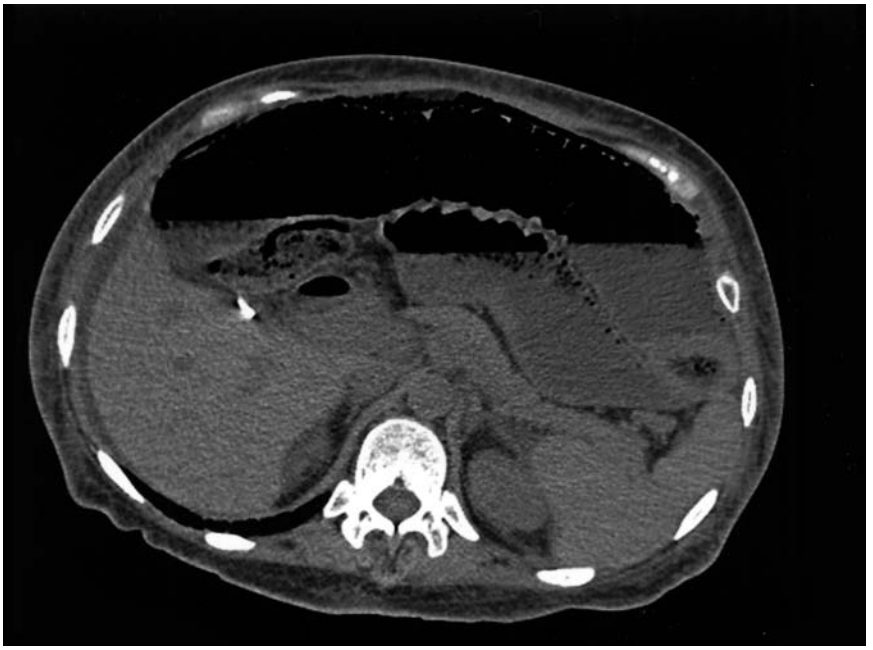

Figure 1. CT of abdomen and pelvis with intravenous contrast shows pneumatosis in the jejunum of the left flank.

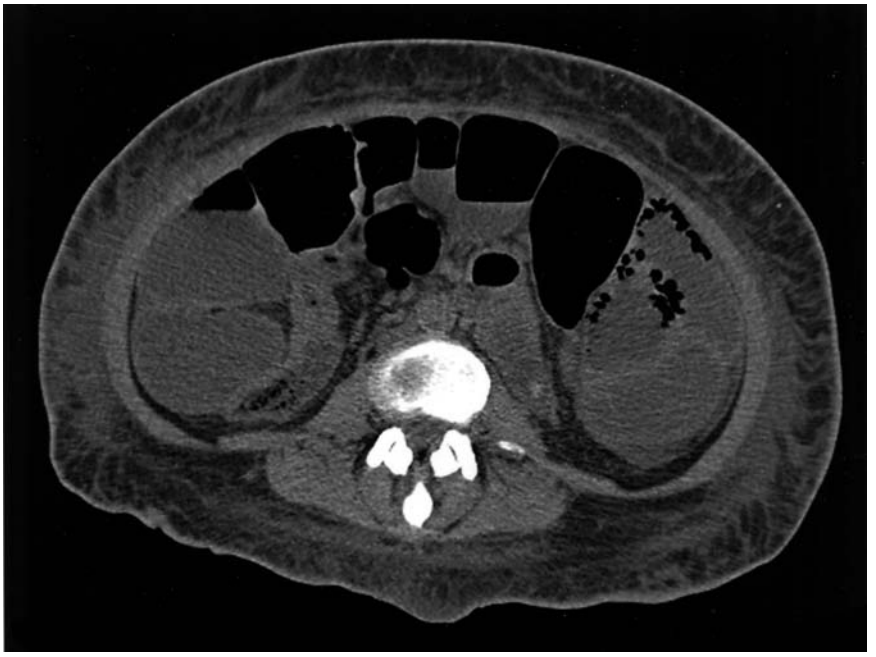

Figure 2. CT of abdomen and pelvis with intravenous contrast shows pneumoperitoneum and pneumatosis in the jejunum.

gastroparesis and small-bowel dysmotility. Upper endoscopy aspirates of the small bowel were remarkable for bacterial overgrowth. Computed tomography (CT) scan of the abdomen and pelvis revealed pneumoperitoneum and pneumatosis in the jejunum (Figures 1 and 2). There was no evidence of bowel perforation. She had no abdominal pain, and examination of the abdomen was negative for peritoneal signs. She was managed conservatively with oral antibiotics and bowel rest, with improvement of diarrhea upon discharge.

Although traditionally considered a surgical emergency, pneumatosis intestinalis, characterized by gas in the bowel wall, is a rare manifestation of connective tissue disease. The intraabdominal process generally follows a benign course with conservative management in patients with scleroderma[1]. Stasis coupled with bacterial overgrowth and small-vessel vasculopathic changes of the gut mucosa may be factors associated with increased risk[2,3]. In scleroderma, it is a marker of poor prognosis. Immunosuppressive therapy is not indicated[1,3,4].

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