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Pneumatosis cystoides intestinalis: Is surgery always indicated?

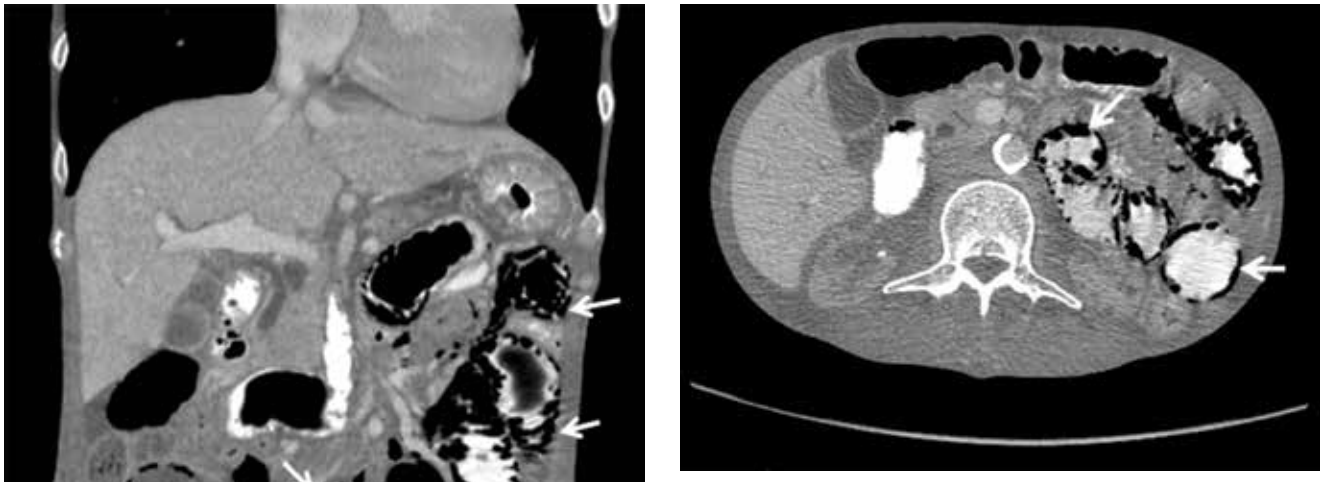

FIGURE 1. On abdominal computed tomography, the coronal view (left) and the sagittal view (right) showed pockets of intramural gas within the small intestine (arrows).

A 57-year-old man with long-standing systemic sclerosis presented with worsening diffuse abdominal pain associated with several episodes of nonbloody emesis for 5 days. He had been hospitalized numerous times over the past 2 years for similar symptoms. In those instances, abdominal radiography and computed tomography (CT) had revealed nonspecific intestinal pseudo-obstruction that had resolved within a few days with bowel rest, antibiotics for small-intestinal bacterial overgrowth, and supportive care.

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At the time of this presentation, physical examination showed stable vital signs, a tympanic, distended abdomen with diffuse tenderness, and diminished bowel sounds with no sign of peritonitis. Complete blood cell counts, renal function testing, and serum lactate levels were unremarkable.

Abdominal radiography showed mildly dilated loops of small bowel with multiple fluid levels, raising concern for intestinal obstruction. Interestingly, abdominal CT revealed extensive pneumatosis cystoides intestinalis of the entire small bowel with sparing of the colon, which raised concern for acute bowel ischemia (FIGURE 1). However, given the patient's underlying systemic sclerosis and current stable condition, the general surgeon recommended conservative management with bowel rest, rifaximin to treat the small-intestinal bacterial overgrowth, and intravenous fluids, which resulted in significant clinical improvement. A liquid diet was initiated and advanced as tolerated to a soft diet before he was discharged home after 8 days of hospitalization.

A RARE, USUALLY BENIGN COMPLICATION OF SYSTEMIC SCLEROSIS

Pneumatosis cystoides intestinalis is a rare gastrointestinal complication of systemic sclerosis characterized by intramural accumulation of gas within thin-walled cysts. It is postulated to result either from excess hydrogen gas produced by intraluminal bacterial fermentation and altered partial pressure of nitrogen within the intestinal wall (the bacterial theory),[1] or from the transgression of gas cysts through the layers of bowel wall as a result of high luminal pressure from intestinal obstruction (the mechanical theory).[2]

The more widespread use of diagnostic CT in recent years has led to increased recognition of this condition, a finding that also often raises concern for intestinal necrosis or perforation.[3] Meticulous correlation of the clinical presentation with corroborative laboratory testing should determine whether a conservative medical approach or emergency surgical exploration is appropriate.[4]

Pneumatosis cystoides intestinalis in patients with systemic sclerosis is a benign condition that generally resolves with bowel rest, antibiotics, inhalational oxygen therapy, and supportive care.[5] An elevated venous oxygen concentration from high-flow oxygen therapy is believed to attenuate the gaseous cysts by decreasing the partial pressure of the nitrogenous gases and by being toxic to the anaerobic gut bacteria.

About 3% of patients with pneumatosis cystoides intestinalis develop complications such as pneumoperitoneum, intestinal volvulus, obstruction, or hemorrhage. Evidence of pneumoperitoneum or bowel infarction—such as the presence of portomesenteric venous gas, a decreased arterial pH, or an elevated lactic acid or amylase level—warrants immediate surgical intervention. Overall, early recognition and watchful monitoring for bowel necrosis or perforation are preferred over reflexive surgical exploration.

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