Intestinal Pseudo-Obstruction and Pneumatosis Cystoides Intestinalis in a Scleroderma Patient

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A 34-year-old woman with a 3-year history of scleroderma treated with corticosteroids, presented with 2 weeks of abdominal pain and distension and, more recently, nausea. Two months earlier, she had a cesarean section at 29 weeks of gestation for fetal distress. Physical examination showed diffuse abdominal tenderness with signs of peritonitis, and no fever. She had normal white blood cell count, C reactive protein was 45 mg/L (normal range < 10 mg/L). X-ray (A) showed pneumoperitoneum with signs of diffuse gas infiltration. CT scans confirmed the pneumoperitoneum (B, arrow a) with free peritoneal exudates and dilated loops of the small bowel (B, arrow b). Retropneumoperitoneum (B, arrow c) with massive gas infiltration of the mesentery (B, arrow d) and two cysts of pneumatosis cystoides intestinalis (B, arrows e) were also seen on the CT scan.

At laparotomy, we discovered an incomplete strangulation of a small intestine loop on an incisional hernia of the Pfannenstiel section. The small bowel and the mesentery were massively infiltrated by multiple gas bubbles (C, arrows). Dilated loops of the small bowel were noted above the stricture. Other adhesions to the Pfannenstiel section were divided. Bacteriologic examination of diffuse peritoneal exudates revealed *Escherichia coli*. The patient’s postoperative course was uneventful, with bowel movement on day 4. Two years later, this young woman had no recurrence of the digestive symptoms.
About 50% of patients with scleroderma experience significant clinical involvement of the alimentary tract. Symptoms of gastroesophageal reflux and severe constipation are often reported. Manometric and electrophysiologic studies have shown evidence of a neuropathy of the enteric nervous system in the early stages of the disease, resulting in disturbances of digestive peristalsis and leading to gastroparesis, bacterial overgrowth of the small intestine, or constipation. Late collagen deposition and atrophy of the smooth muscle layer of the bowel wall cause loss of function of the lower esophageal sphincter or the anal sphincter and marked atony of parts of the intestine. Therapeutic options include H2-antagonists, proton-pump inhibitors, pro-kinetic drugs, octreotides, and antibiotics. Nutritional supplementation and surgical interventions are often of limited therapeutic value. In selected cases, operation for intestinal failure is an option with resection or bypass of affected segments or placement of enterostomy tubes for feeding or decompression. In some cases longterm total parenteral nutrition is warranted.

Pneumatosis cystoides intestinalis is a rare condition characterized by the presence of multiple gas-containing thin-walled cysts in the intestinal wall and mesentery. It is sometimes associated with chronic obstructive pulmonary disease, but has been described most often in patients with gastrointestinal disorders including duodenal and gastric ulceration, small bowel obstruction, regional enteritis, and gastrointestinal malignancy. It is a benign condition that often responds to conservative management, but it may be a harbinger of end-stage disease, particularly in progressive systemic sclerosis. Pneumatosis cystoides intestinalis may be caused by excessive hydrogen production by intestinal bacteria altering the partial pressure of nitrogen in the intestinal wall. Pneumatosis cystoides intestinalis should be considered in any patient with systemic sclerosis who experiences abdominal symptoms suggestive of acute or subacute intestinal obstruction, not readily explicable by other causes. These symptoms could be treated nonoperatively, but with this patient signs of small-bowel obstruction and peritoneal irritation laparotomy was mandatory. At laparotomy we discovered an incomplete strangulation of a small intestine loop on an incisional hernia of the Pfannenstiel section which explains the intestinal obstruction. We were impressed by the massive gas infiltration of the small bowel wall and the mesentery which is uncommon in classic forms of intestinal obstruction. Bacterial translocation from an intestinal obstruction may explain the presence of *E coli* in the peritoneal fluid.

REFERENCES