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High-Grade Small Bowel Obstruction Secondary to Advanced Scleroderma

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Background	A 63-year-old woman presented with recurrent high-grade small bowel obstruction secondary to scleroderma.
Summary	This report describes a unique case of small bowel obstruction secondary to peritonealized sclerotic lesions surrounding the small bowel. A 63-year-old woman with a history of scleroderma and multiple prior small bowel obstructions presented with severe constipation and chronic small bowel obstruction requiring operative intervention. Intraoperatively she was found to have dense fibrotic circumferential sclerotic rinds surrounding the small bowel. The patient underwent extensive sharp and blunt lysis of adhesions and placement of a jejunal Baker tube intending to stent open the small bowel during the postoperative period. She regained bowel function on postoperative day (POD) 3, which continued after discharge. While gastrointestinal involvement in scleroderma is common, it is often related to impaired intestinal motility.
Conclusion	Scleroderma commonly results in gastrointestinal complications but rarely causes actual small bowel obstruction. This case highlights the unusual formation of circumferential peritonealized sclerotic obstructing scars that can be treated surgically without resection.
Key Words	scleroderma; small bowel obstruction; systemic sclerosis

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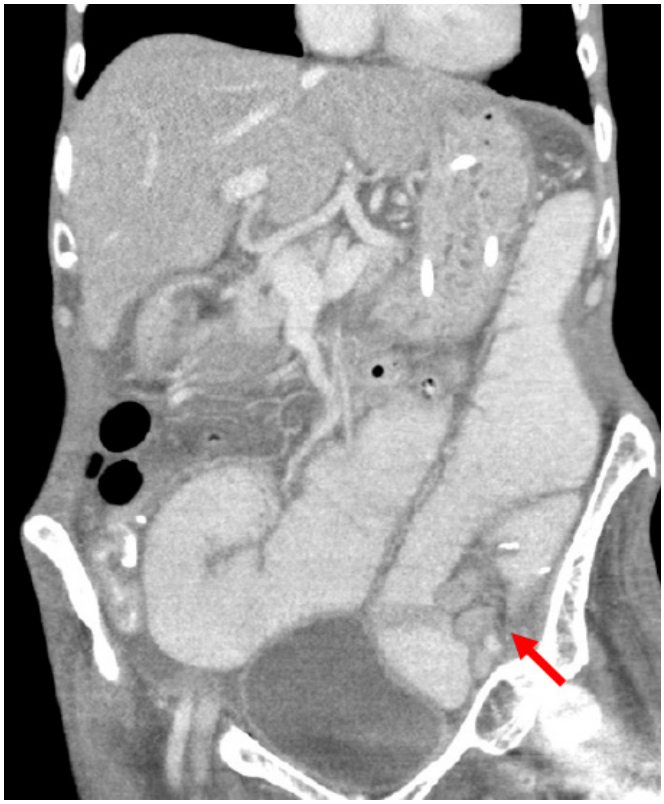
Case Description

The patient is a 63-year-old woman who was admitted for treatment of a high-grade small bowel obstruction secondary to scleroderma. Her medical history includes advanced scleroderma, Raynaud syndrome, cystocele, rectocele, small intestinal bacterial overgrowth, gastroesophageal reflux disease, pulmonary hypertension, interstitial lung disease, psoriatic arthritis, gastroparesis, and depression. Past surgical history is notable for total abdominal hysterectomy with cystocele/rectocele repair, inguinal hernia repair with mesh, ventral rectopexy with mesh, small bowel resection, sacral nerve stimulator placement and EGD with esophageal Botox injections.

Figure 1. Preoperative CT Scan. Published with Permission

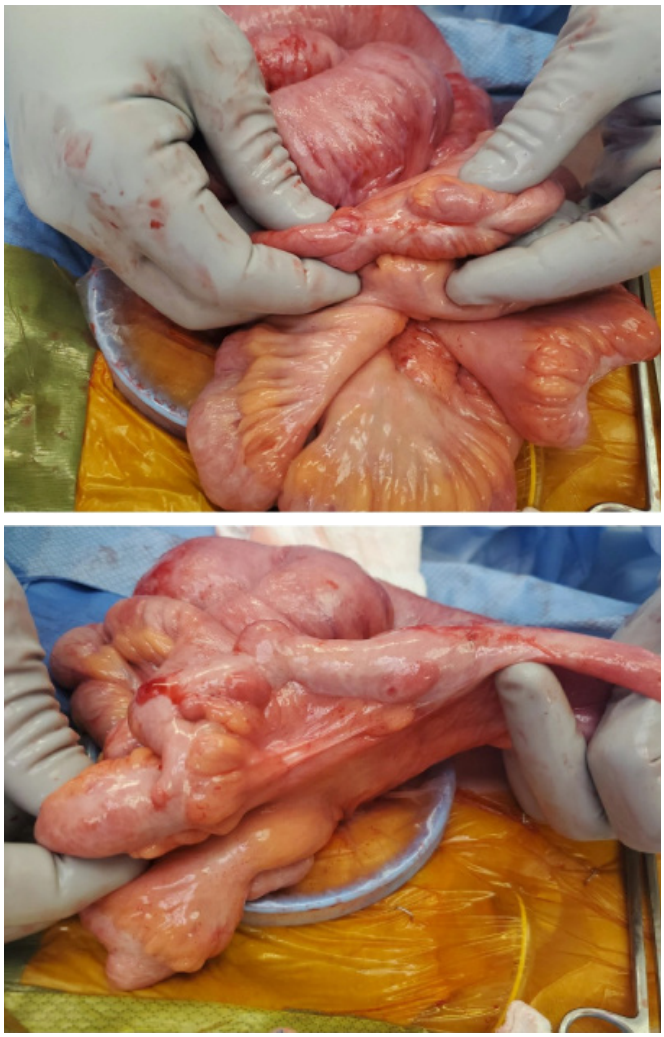


Scan appreciated as high-grade small bowel obstruction with transition point in distal jejunum/proximal ileum in left lower quadrant, at the site of the surgical anastomosis, marked by arrow.

She had multiple admissions for small bowel obstructions years prior that were successfully treated nonoperatively. Subsequently, she was found to have dense adhesions and dilated loops of small bowel during a rectal prolapse surgery. She underwent extensive lysis of adhesions (LOA), a small bowel resection, and rectal prolapse repair. Five months later, she was again admitted for high-grade small bowel obstruction (SBO). At that time, she had protein-deficient malnourishment. After failing medical management, she was discharged on total parenteral nutrition (TPN) with a decompressing gastric tube in place. She reported no bowel function for four consecutive months. During this time, she underwent a multidisciplinary conservative approach to treatment with gastroenterology, rheumatology, and surgery, which was unsuccessful in resolving her symptoms. Ultimately, while consideration was given to avoiding surgery in this patient with multiple medical comorbidities, the decision was made that operative intervention with lysis of adhesions and revision of her small bowel anastomosis was the best option. Given her multiple previous bowel surgeries and the expectation that she would have extensive dense adhesions, she was scheduled for an open procedure.

The findings in the OR were that the jejunum was markedly dilated (with no adhesions), and the ileum was found to have unusual sclerosis just distal to her previous intestinal anastomosis, consisting of a circumferential sclerotic rind that was peritonealized and extended through the distal third of the small intestine. The sclerotic scarring caused severe telescoping and kinking of the bowel leading to functional obstruction. There were no interloop adhesions. These circumferential adhesions were lysed mostly with blunt dissection (some sharp as well), revealing normal-appearing ileum beneath. After this dissection was complete, it was easy to milk the intestinal contents through the entire length of the small bowel. The previous anastomotic site was patent and not the site of her SBO. The circumferential adhesions were extensive, and without an identifiable lead point, small bowel resection was not an option without accepting a high risk of short bowel syndrome. While intestinal contents could be passed through the bowel at this point in the operation, believing that she was at very high risk of recurrence, the decision was made to place a long Baker tube (a 270 cm-long tube with a balloon at the end with a dual lumen) through the abdominal wall, into the proximal jejunum and passed all the way to the cecum. The goal was to stent open the bowel for some time, long enough to prevent these adhesions from forming again in the immediate postoperative period.

Figure 2. Intraoperative Images. Published with Permission



Note circumferential sclerotic rinds throughout distal small bowel that were amenable to blunt lysis that relieved her obstruction.

The patient did well postoperatively. She was placed on steroids (with taper), hoping she would be less likely to form more of these sclerotic scars. She regained bowel function on postoperative day (POD) 3. She was discharged home on POD 5 on a full liquid diet with supplementary TPN. She had persistent bowel function. The Baker tube was removed at the five-week visit without complication. She had continued bowel function at her most recent follow-up three months postoperatively.

Discussion

Scleroderma is clinically challenging and has the highest mortality of any rheumatologic disease.[1] Although the pathogenesis has not been fully elucidated, leading theories suggest that interactions between endothelial cells, lymphocytes, macrophages, and fibroblasts result in the activation of myofibroblasts, leading to extracellular matrix overproduction and tissue fibrosis.[2] This takes place in all body systems, and, as such, the disease can present in many different ways. Some more common manifestations include Raynaud's phenomenon, musculoskeletal abnormalities, fatigue, lung fibrosis, pulmonary artery hypertension, and renal failure.[1]

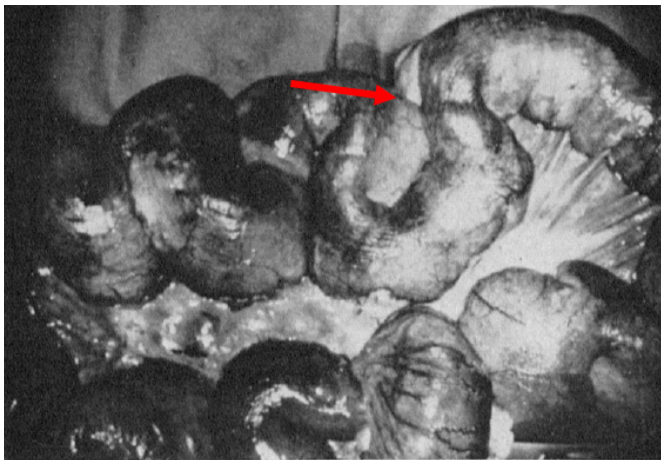
Gastrointestinal manifestations of scleroderma are common and frequently are one of the earliest manifestations of the disease.[3] In most cases, symptoms are related to impaired motility or contractility of the esophagus, stomach, small bowel, colon, or rectum.[4] This results in gastroesophageal reflux disease, delayed gastric emptying, diverticulosis, and fecal incontinence, among other complications. In the small bowel, scleroderma most commonly results in small intestinal bacterial overgrowth (SIBO) and intestinal pseudo-obstruction.[4] Patients often present similarly to an actual small bowel obstruction with abdominal pain, postprandial bloating, abdominal distention, and signs of malnutrition or malabsorption. When they develop SIBO, they also complain of diarrhea and develop electrolyte abnormalities.[5] Intestinal pseudo-obstruction occurs in 3.7%–5.4% of patients with scleroderma and is a high morbidity and mortality complication.[6,7]

While small bowel involvement and pseudo-obstruction are common, actual small bowel obstruction is rare and has only been documented once in the literature (in 1959[8]). Herrington et al. reported a case where a 55-year-old male with scleroderma presented with acute small bowel obstruction and recent weight loss of 20 pounds. Intraoperatively, they describe the jejunum as "grossly abnormal" with multiple "thickened, edematous, and grayish white" areas where the bowel was constricted, and the decision was made to resect 280 cm of small bowel (Figure 3). In our case, the decision was made not to resect any of the small bowel, as once the dense adhesions and rinds were lysed, the small bowel appeared patent, and it was easy to pass intestinal contents from the duodenum down to the ileocecal valve and into the colon. While there was no bowel resection and, as such, no specimen to compare to that reported by Herrington et al., the gross description and photo in their case were remarkably similar to our reported case.

The decision was made to place a Baker tube extending from the proximal jejunum into the cecum with the goal that this tube would act as a stent if these adhesions were to form again in the immediate postoperative period, and the

patient was placed on steroids postoperatively. Operative intervention for scleroderma is not unique to this case, and small bowel resection has been used in the past for intestinal pseudo-obstruction.[9] However, actual small bowel obstruction is a rare occurrence.

Figure 3. Intestine with Gross Findings Compatible with Scleroderma,8 with Notably Similar Appearance to This Case.



Reproduced from Herrington JL Jr. Scleroderma as a cause of small-bowel obstruction; successful treatment of a case by intestinal resection. AMA Arch Surg. 1959;78(1):17-24 with permission from JAMA Surgery

Here we describe a rare case of actual intestinal obstruction due to scleroderma that was treated successfully with mostly blunt lysis of the sclerotic circumferential scarring around the small bowel without needing resection. It is important to be aware of this potential complication so it can be identified and treated early. Small bowel obstruction has a high level of morbidity and mortality, with an incidence of 350,000 per year in the United States and a mortality of 10%. This mortality increases to 30% in cases with intestinal necrosis or perforation.[10] It is important to recognize SBO early. Patients with scleroderma symptomology consistent with bowel obstruction should not be assumed to have pseudo-obstruction based on their prior diagnosis of scleroderma alone. Early identification and intervention for actual obstruction could result in significantly better outcomes for these patients and recognize the possibility of appropriate successful surgical management with mechanical/blunt lysis of the circumferential adhesions (without the need for bowel resection).

Conclusion

Scleroderma commonly involves the gastrointestinal tract but only rarely causes actual small bowel obstruction. Our case reports a rare case of actual small bowel obstruction in the setting of scleroderma and a reasonable approach to treatment with mechanical lysis of circumferential bowel adhesions and Baker tube placement (without intestinal resection). This case highlights the need for awareness of the potential for true small bowel obstruction in patients with scleroderma.

Lessons Learned

Although intestinal involvement in scleroderma commonly results in pseudo-obstruction and small intestinal bacterial overgrowth, actual small bowel obstruction cannot be ruled out, and patients should be thoroughly worked up and evaluated. Operative intervention in these patients could result in significantly better outcomes and may be successful by bluntly breaking the circumferential scarring (without small bowel resection).

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