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CASE REPORT | COLON

An Unusual Case of Obstruction Due to Colonic Intussusception in a Scleroderma Patient

Kristina N. Katz, MD[1], Kevin Skole, MD[2], and Jean M. Fede, DO[3]

[1]*Department of Internal Medicine, Rutgers Robert Wood Johnson Medical School, New Brunswick, NJ*
[2]*Department of Gastroenterology, University Medical Center of Princeton at Plainsboro, Plainsboro, NJ*
[3]*Department of Pathology, University Medical Center of Princeton at Plainsboro, Plainsboro, NJ*

Abstract

Scleroderma is an autoimmune disease that can affect multiple organ systems, including the gastrointestinal tract. Intussusception, the telescoping of the bowel that can lead to intestinal obstruction, is an uncommon phenomenon in adults. We report the first case of sigmoid intussusception in a patient with scleroderma.

Introduction

Scleroderma affects multiple organs, including the gastrointestinal (GI) tract.[1] GI manifestations include esophageal dysmotility, gastric antral vascular ectasia (GAVE) syndrome, small intestinal bacterial overgrowth (SIBO), malabsorption, and constipation.[2] Intussusception of the small intestine has also been associated with scleroderma.

Case Report

A 44-year-old female with systemic scleroderma presented with abdominal pain and constipation. Her last bowel movement was 11 days prior to admission despite trials of laxatives, suppositories, and enemas. Prior to this, she had been having diarrhea; she did not respond to treatment for recurrent SIBO and had used loperamide to reduce her bowel movements. Her other medications included enalapril, isradipine, esomeprazole, bupropion, aspirin, and as-needed etodolac and bisacodyl. She had not been on immunosuppression for several years.

The patient was tachycardic, afebrile, and normotensive. Her abdomen was distended and diffusely tender with absent bowel sounds. Labs were notable for a WBC of 13.1k/mm^3. Contrast-enhanced abdominal/pelvic computed tomography (CT) scan showed large bowel distention, sigmoid intussusception, mesenteric lymphadenopathy, and inflammatory changes (Figure 1). Sigmoidoscopy revealed a fecalith with surrounding necrotic-appearing bowel. The patient underwent a sigmoid resection and diverting colostomy. Surgical findings included an inflamed sigmoid colon with kinking and adherent omentum, and adhesions in the region of the sigmoid colon. Pathologic findings included ulcerated colonic mucosa with granulation tissue, fibropurulent exudate, marked acute inflammation, and serositis without any evidence of fibrosis, neoplasia, or diverticular disease (Figure 2). The patient had a complicated post-operative hospital course, and after a 1-month hospitalization, was discharged to a rehabilitation facility.

Discussion

Scleroderma often affects the gastrointestinal tract. This is usually due to collagen infiltration of the mucosa and submucosa with atrophy and thinning of the muscularis externa.[3] Small bowel involvement occurs in 17–57% of patients with scleroderma.[2] Small bowel dysmotility is the most common finding, likely due to the combina-

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Correspondence: Kristina N. Katz, 5 Indigo Drive, Old Bridge, NJ 08857 (katzkn@gmail.com).

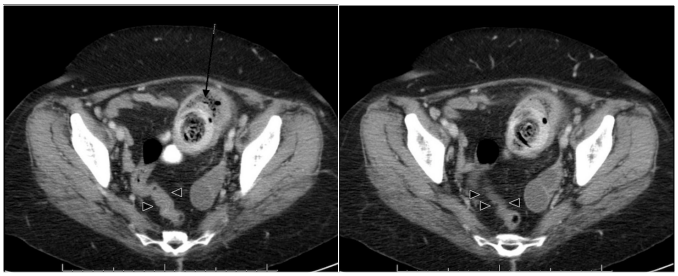

Figure 1. Abdominal/pelvic CT showing sigmoid intussusception (arrowheads) and proximally dilated colon with wall thickening and adjacent inflammatory changes (arrow).

tion of dysfunction of the cholinergic neural pathway and underlying myopathy.[2] The small bowel can have thickened folds or a "hide-bound" bowel appearance, with the latter finding due to small distances between the valvulae even in the presence of bowel dilation.[4,5] Bowel stasis can result in SIBO.[6] Pseudo-obstruction syndrome, which is a progressive dilation and paralysis of the bowel without mechanical obstruction, is also associated with scleroderma.[7]

The large bowel is involved in 10–50% of patients with scleroderma.[2] Constipation is the most common symptom, and can lead to ulceration and even colonic perforation.[6,8] Additionally, the gastrocolic reflex may be lost.[9] Thinning and weakening of the muscle wall can lead to formation of large diverticuli, and dysfunction of the anorectum can lead to fecal incontinence.[6] Typical radiographic findings of the large bowel include increased luminal fluid, dilation, and lack of haustrations.[2] Colonic atony can be seen on post-evacuation barium enema.[8] Pseudo-obstruction syndrome can be seen in the large bowel.[7]

Intussusception occurs when a bowel segment with its associated mesentery invaginates into the lumen of an adjacent bowel segment. In adults, intussusception is responsible for 1% of all bowel obstructions.[10] Intussusceptions may be caused by intraluminal, mural, and extraluminal processes.[11] In 70–90%, there is an identified organic lesion, with 20–50% of the lesions being malignancy.[10] Scleroderma was reported to be responsible in 1 of 58 adult intussusception cases during a 30-year review.[10] The etiology behind non-

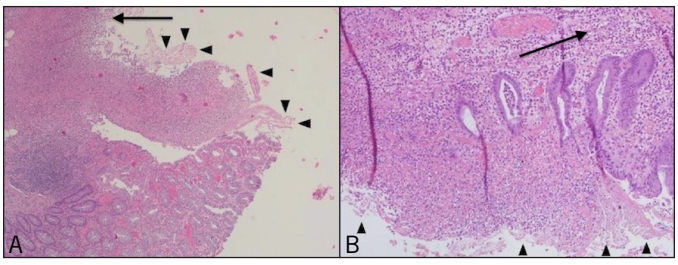

Figure 2. (A) Low-power view and (B) high-power view of colon tissue from sigmoid resection. Both show ulceration with polymorphonuclear leukocytes (arrow) and fibroproliferative exudative changes (arrowheads).

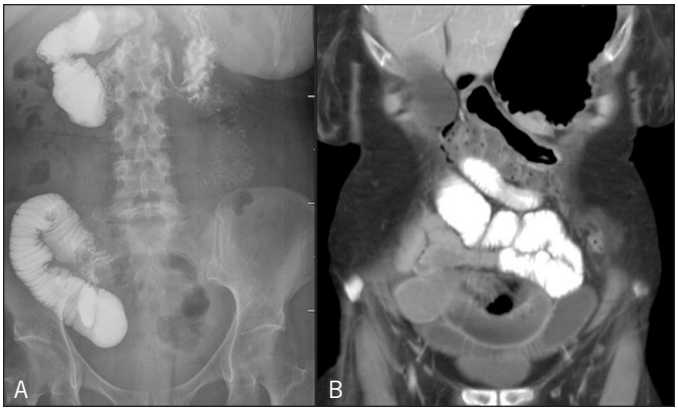

Figure 3. (A) Upper GI series showing decreased space between valvulae, thickening of small bowel folds and dilation of lumen, consistent with hide-bound bowel. (B) Coronal image from abdominal/pelvic CT showing similar findings of hide-bound bowel.

mechanical intussusception in scleroderma patients is unclear. It has been proposed that hide-bound bowel and disordered peristalsis causes a focal kink, which then acts as a lead point.[5,11] Our patient previously had findings consistent with hide-bound small bowel (Figure 3). Loss of haustration is commonly seen in sclerodermatous large bowel, but it is unknown if it could create a similar phenomenon. Furthermore, it has been reported that bowel edema and mesenteric lymphadenopathy may contribute to the mechanism of intussusception.[12] Our patient had enlarged mesenteric lymph nodes and bowel wall edema. She had sigmoid adhesion, without past abdominal surgeries to explain their formation. Non-surgical causes of adhesions include chronic intra-abdominal inflammation, and scleroderma is inherently an inflammatory disease associated with collagen deposition in the mucosa, and fibrosis of the luminal wall.[13] Any of these causes could create mural abnormalities, and might lead to the telescoping of bowel.[2,11] The histopathology in our patient did confirm acute inflammation, but did not reveal any collagen deposition or fibrosis.

We believe the intussusception in our patient was a result of multifactorial manifestations of her systemic disease. Scleroderma patients and their physicians should be aware of the possibility of colonic intussusception, as aggressive medical and surgical management may be needed to avoid significant morbidity and even mortality.

Disclosures

Author contributions: KN Katz and K. Skole wrote, edited, and reviewed the manuscript. JM Fede provided pathology images and reviewed the final manuscript. KN Katz is the author guarantor.

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