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Matt Baichi

*Thomas Jefferson University*

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## A Case Of Chronic Intestinal Pseudo-Obstruction

Matt Baichi, MD, Resident, Internal Medicine 2000-2003

### Introduction

Scleroderma is a systemic disease characterized by the deposition of excessive collagen and other matrix elements in the skin as well as in multiple internal organs. Scleroderma can be classified into diffuse cutaneous disease and limited cutaneous disease<sup>1</sup>. Limited cutaneous disease is characterized by skin involvement limited to the hands, face, feet, and forearms; it includes the CREST variant (calcinosis, raynauds, esophageal dysmotility, sclerodactyly, and telangiectasia). Diffuse cutaneous disease is characterized by skin involvement as well as early and diffuse visceral involvement. Clinically significant gastrointestinal involvement occurs in approximately 50% of all patients with scleroderma<sup>2</sup>. The esophagus is the most common site of involvement followed by the ano-rectum, small bowel, colon, and stomach. Several recent reviews of the gastrointestinal manifestations of scleroderma have been written<sup>3-5</sup>. Here, a case of diffuse gastrointestinal scleroderma presenting as chronic intestinal pseudo-obstruction is described.

### Case report

The patient is a 52-year-old female with a 30-year history of GERD, 10-year history of chronic constipation, and a questionable 4-year history of Crohn's disease. The patient was in her usual state of health until three months prior to this admission. At that time, she was hospitalized for nausea and vomiting. Work-up revealed a non-mechanical small bowel obstruction that resolved with conservative management. She was re-hospitalized six times for similar complaints. Each admission revealed non-mechanical small bowel obstruction. Her last admission, however, revealed an ileal stricture, which was subsequently removed. The patient now presents to our hospital with complaints of nausea, bilious emesis, bloating, abdominal distention, increased belching, and decreased appetite. She denies dysphagia, abdominal pain, melena, hematochezia, or diarrhea. She denies history of hepatitis, pancreatitis, or gallbladder disease. Since her first hospitalization three months ago, she has lost 25 pounds. Social history is remarkable for 80-pack years of tobacco. Family history reveals no gastrointestinal disease. Review of systems is remarkable for persistent paresthesias and Raynaud's.

The patient is a moderately cachectic white female. Vital signs are unremarkable except for mild tachycardia. Abdominal exam reveals normal bowel sounds, marked distention, tympanic percussion, soft and non-tender to palpation, without guarding or rebound, and without hernia. Rectal exam reveals external hemorrhoids with brown stool positive for occult blood. Rheumatologic exam reveals mildly thickened and cool fingers, no inflamed joints, normal skin. Neurologic exam reveals normal muscle tone and strength. Blood count is remarkable for white count 14600/mL with normal differential. Chemistry profile is remarkable for potassium 2.5 Meq/L. Liver profile, amylase, and lipase were within normal limits. ESR 17 seconds, ANA 1:40, speckled. Anticentromere Ab and Anti Scl 70 Ab were negative. Obstruction series showed multiple dilated small bowel loops with relative collapse of the large bowel, air fluid levels within the small bowel, and no intra-peritoneal free air. Pathology of the resected terminal ileum (from previous hospitalization) revealed patchy marked atrophy and fibrosis of the muscularis propria with preferential involvement of the circular layer, vacuolar degeneration of muscle fibers were present, without inflammatory changes of Crohn's Disease. Esophageal manometry revealed lower esophageal sphincter resting pressure in the lower range of normal with aperistalsis of the esophageal body. Small bowel radiographs revealed moderate mega-duodenum with "hide-bound" configuration of mucosal folds throughout the small intestine, but no stricture. Upper endoscopy revealed erosive esophagitis and moderate hiatal hernia. Colonoscopy revealed neoileo-colonic anastomosis that was friable and ulcerated. Biopsy of the anastomosis showed granulation tissue and lamina propria compatible with the anastomotic site.

### Diagnosis

Our patient presented with chronic intestinal pseudo-obstruction<sup>6-9</sup>. This is a rare condition characterized by recurrent episodes of intestinal obstruction in the absence of any mechanical defect. Symptoms include nausea, vomiting, abdominal distention, bloating, abdominal pain, and constipation. The differential diagnosis can be divided into myopathic vs neuropathic disorders. Myopathic conditions include infiltrative processes such

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as scleroderma and amyloidosis, familial conditions such as visceral myopathies, and neurologic conditions such as myotonic and other dystrophies. In this case, the history is significant for esophageal dysmotility (longstanding GERD) and Raynaud's phenomenon. Recent ileal biopsy shows patchy atrophy and fibrosis of the muscularis with preferential involvement of the circular layer. These findings are consistent with scleroderma. Histologically, the most significant changes occur in the muscularis layer of bowel. Atrophy and fragmentation of smooth muscle occurs. Initially the atrophy is patchy and later becomes more extensive with associated fibrosis<sup>10,11</sup>. These changes are more pronounced in the circular smooth muscle layer<sup>12</sup>, and atrophy usually exceeds fibrosis<sup>13</sup>. Subsequent small bowel imaging and esophageal manometry confirmed the diagnosis (see discussion).

### Discussion

The esophagus is the most commonly affected organ in the gastrointestinal tract. Normal esophageal manometry shows high amplitude, ordered peristaltic waves and a high resting pressure of the lower esophageal sphincter (LES). On swallowing, the LES relaxes to baseline, and the LES relaxation precedes the arrival of the peristaltic wave. Manometry studies in early scleroderma show increased velocity of the peristaltic wave, discordance of the peristaltic wave with LES relaxation, and failure of the LES to relax to baseline. Later findings include decreased amplitude of peristalsis and low LES resting pressure progressing to aperistalsis in the smooth muscle portion of the esophagus with absent LES resting pressure<sup>14,15</sup>. Gastroesophageal reflux disease (GERD) is the most significant clinical condition. GERD occurs as a result of both the decreased LES resting pressure, which allows an increased number of reflux events, and poor peristalsis, which delays acid clearance<sup>16,17</sup>. The complications of longstanding GERD include candida esophagitis, erosive esophagitis, esophageal stricture, Barrett's esophagus, and aspiration. Diagnostic evaluation should therefore include upper endoscopy to rule out complications.

The small intestine is also commonly involved. In the normal small bowel, a cyclical pattern of contractile activity occurs at regular intervals. This is called the migrating myoelectric complex (MMC). The MMC acts as an "intestinal housekeeper" by clearing remnants of digestion and preventing bacterial overgrowth<sup>18</sup>. Patients with scleroderma often lack this MMC. Clinically, chronic intestinal pseudoobstruction and malabsorption are seen. Malabsorption is the result of bacterial overgrowth<sup>19</sup>. The H<sub>2</sub> breath test is a noninvasive and reliable diagnostic procedure. A nonabsorbable carbohydrate such as lactulose is ingested and a premature rise in exhaled H<sub>2</sub> suggests the presence of intestinal bacteria. The diagnosis of chronic intestinal pseudoobstruction from scleroderma can be made with small bowel barium imaging or biopsy<sup>20,21</sup>. A "hide-bound" configuration of the small bowel is a characteristic mucosal fold pattern in which there is a relative decrease in the distance separating the valvulae conniventes. This creates an accordion-like appearance. This finding is uniquely associated with scleroderma. Less common intestinal presentations include pneumatosis cystoides intestinalis, small intestine telangiectasia, and small bowel diverticula. Prokinetic drugs such as metaclopramide, erythromycin, and octreotide are the treatments of choice.

In the colon, the ano-rectum is commonly affected. Manometry reveals an absent or diminished rectoanal inhibitory reflex<sup>22</sup>. Also, colonic transit is prolonged<sup>23</sup> and the normal postprandial increase in rectosigmoid motility is lost<sup>24</sup>. Clinically, fecal incontinence and constipation are the most common presentations. Less common presentations include diarrhea, rectal prolapse, spontaneous perforation, and colonic infarction. Radiographic evaluation of colonic involvement includes barium enema, which may show characteristic "wide mouth", or "fish mouth" diverticula, which are broad based true diverticula caused by the uneven distribution of atrophic muscularis<sup>20</sup>. Treatment includes conservative measures such as increased fluid and fiber intake. Prokinetic agents can be used for improved colonic inertia.

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