Intestinal obstruction and pseudo-obstruction in patients with systemic sclerosis

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Summary
Chronic intestinal pseudo-obstruction is a known complication of patients with systemic sclerosis, manifested as nausea, vomiting, constipation, abdominal distension and pain. We report a series of cases with systemic sclerosis that presented with signs of intestinal obstruction. In all cases, the differentiation between a pseudo-obstruction and true mechanical obstruction remained a formidable challenge. Our goal was to present different scenarios of patients with systemic sclerosis and features of intestinal obstruction, with a review on its clinical approach.

Key words. Pseudo-obstruction, intestinal obstruction, systemic sclerosis.

Obstrucción intestinal y pseudo-obstrucción en pacientes con esclerosis sistémica

Resumen
La pseudo-obstrucción intestinal crónica es una conocida complicación en los pacientes con esclerosis sistémica que se manifiesta con náuseas, vómitos, estreñimiento, distensión y dolor abdominal. Mostramos una serie de casos con esclerosis sistémica que presentaron con signos de obstrucción intestinal. En todos los casos la diferenciación entre pseudo-obstrucción y obstrucción mecánica representó un desafío. Nuestro objetivo fue presentar diferentes escenarios de pacientes con esclerosis sistémica y un cuadro de obstrucción intestinal junto a una revisión del abordaje clínico.

Palabras claves. Pseudo-obstrucción, obstrucción intestinal, esclerosis sistémica.

Gastrointestinal manifestations occur in near 90% of patients with systemic sclerosis (SSc) as a result of small vessel vasculitis and proliferation of connective tissue. One of the common gastrointestinal complications is recurrent bowel pseudo-obstruction. However, differentiation between pseudo-obstruction and mechanical intestinal obstruction may be challenging and lead to either delay of surgery or unnecessary operation in cases that benefit from conservative management. We describe a series of patients with SSc presenting with signs of intestinal obstruction, each one with different etiology, and all representing a formidable challenge to the diagnostician.

Case reports
Case 1
A 57-year-old woman was admitted after presenting with constipation, nausea, vomiting, abdominal distension and pain that began one week prior to her admission. Her past history included SSc diagnosed two years ago, complicated with intermittent bouts of vomiting, abdominal distension, weight loss, and diarrhea alternating with constipation, that had worsened over the past six months. She had two previous admissions for episodes of acute bowel pseudo-obstruction that resolved with conservative treatment. Multiple tomographic imaging studies over the past eight months showed diffuse small bowel and colon dilation without a definite transition point, consistent with pseudo-obstruction. She was again admitted with diagnosis of pseudo-obstruction, and treated as the previous episodes with nothing by mouth and nasogastric tube (NGT) suction. Despite four days of conservative treatment, the patient had worsening increase in abdominal distension and discomfort. Further imaging was obtained with CT enterography, which showed massively dilated loops of small bowel with a sharp transition zone and an internal hernia at the root of the mesentry in the distal small bowel (Figure 1). During laparotomy a small bowel volvulus was found and straightened with...
immediate filling of the colon with fluid confirming the diagnosis of mechanical obstruction. The patient improved at day 7 after surgery and was later discharged.

**Case 2**

Another 57-year-old woman with history of systemic scleroderma diagnosed one year prior to admission, and complicated with esophageal dismotility, gastroparesis, and recurrent episodes of intestinal pseudo-obstruction, was admitted with acute onset abdominal pain associated to intractable nausea and vomiting. She had had similar but less intense episodes in the past and used to take metoclopramide on a regular basis. Gastrografin upper GI X-ray study done upon admission showed dilatation of the stomach and proximal loops with evidence of a partial distal small bowel obstruction. These findings were confirmed by subsequent abdominal CT scan (Figure 2). A NGT was inserted for decompression and intravenous fluids and total parenteral nutrition were started. One week after conservative treatment was started; there was no resolution of the symptoms. Surgical evaluation was obtained and the patient was taken to the operating room. A dense adhesion was visualized at the level of distal jejunum with proximal distention and distal collapse of bowel loops. The area of intestine compressed by the adhesion showed a palpable stricture that did not expand after lysis of the adhesion, and required resection with side to side anastomosis. The patient had significant improvement in her symptoms and was discharged with prokinetics, diet modifications and parenteral nutritional support.

**Figure 1.** CT enterography showing diffuse dilatation of small bowel loops. Arrow indicates a transition point at the root of the mesentery.

**Figure 2.** Computed tomography with oral contrast showing dilated stomach and small intestine loops. Arrow indicates a transition point in which the lumen is reduced.

**Case 3**

A 56-year-old woman with history of scleroderma and polymyositis was admitted for sudden onset abdominal pain, nausea and vomiting. She had been having several months of alternating episodes of constipation and diarrhea, and had been hospitalized several times for conservative management. A proctocolectomy with ileostomy for rectal prolapse was performed two months prior to admission after she had recurrent episodes of diarrhea and fecal incontinence that had not improved with medical management. The physical exam on admission was pertinent for a distended abdomen without peritoneal signs. Abdominal computed tomography revealed diffuse small bowel dilatation with a pneumoperitoneum. The upper gastrointestinal gastrographin series revealed diffuse dilatation of the small bowel with obstruction probable located at the proximal ileus. Management was started with NGT decompression and total parenteral nutrition. However, there was immediate recurrence of symptoms every time oral intake was attempted. A diagnostic laparoscopy was performed, and it revealed diffusely dilated small bowel down to the end of the ileostomy. No collapsed loops or evidence of mechanical obstruction were visualized. Her postoperative period was prolonged by respiratory, infectious, and gastrointestinal complications that required a lengthy stay in the intensive care unit.
Discussion

SSc is an autoimmune disease of unknown origin characterized by involvement of several organs with neural and endothelial-cell damage. There is proliferation of connective tissue that leads to skin and visceral thickening.1 The progression of the disease is characterized by early vascular and neural plexus lesions that may induce changes in intestinal permeability, absorption and transport, followed by muscle atrophy and fibrosis.2 The small and large bowel can be affected in up to 90% of the cases manifesting as constipation, malabsorption syndrome and pseudo-obstruction. Esophagus is the digestive tract part most commonly affected.3 Previous studies revealed that patients with SSc had prolongation of phase III of the migrating motor complex with an interrupted propagation, manifested as a delayed transit time.4 Chronic intestinal pseudo-obstruction syndrome is characterized by progressive bowel dilatation and paralysis with no structural evidence of mechanical obstruction, usually without response to conservative medical or surgical management.5 In rare cases, intestinal pseudo-obstruction may become lethal, leading to extreme constipation and intestinal rupture.6 Plain abdominal x-ray shows severely dilated bowel loops with air fluid levels; and with barium enema the “hide-bound” bowel sign (narrow separation between valvulae conniventes of normal thickness seen on distended bowel loops) have been described in patients with scleroderma.7 Medical management can be very challenging and is mainly focused on symptomatic relief. The use of prokinetics, subcutaneous octreotide, antibiotics (if bacterial overgrowth is suspected) and endoscopic or surgical procedures has been described as treatment for patients with gastrointestinal involvement secondary to SSc.7,9 However, prokinetics have no effect at end stages of the disease.2 In pseudo-obstruction, the roles of endoscopy and surgery are mostly confined to facilitating nutrition and providing decompression rather than restoring bowel motility.10

Rake reported in 1932 one of the first cases of SSc that required surgical exploration after an apparent intestinal obstruction.11 The identification of patients that may benefit from surgical approach in the setting of slow gastrointestinal transit and recurrent pseudo-obstruction can be especially challenging.12 The possible post-operative complications, which may include respiratory failure due to pulmonary fibrosis, poor healing due to chronic use of steroids, short bowel syndrome if significant resection is required, or superimposed ileus, makes a surgical approach an option of last resort. There is also the suggestion that risk for perforations is increased if anastomotic reconstructions are required.13 If surgery is required, it is advisable to direct it toward the segmental distribution of the disease, trying to resect as minimal bowel as possible. Since the available information in this matter is scarce and mainly based on surgical experience instead of evidence, the decision of surgery and the type of surgery relies on broad surgical principles.14

We present three cases of intestinal obstruction in the setting of SSc. Each one represented a different cause of obstruction: one due to small bowel volvulus (a rare complication in patients with advanced SSc), another with a mechanical obstruction secondary to adhesions (a common event in general inpatients, and not necessarily secondary to SSc), and the third with true intestinal pseudo-obstruction that was taken to the operating room after failed medical management. Case number 1 represents a rare event, and except for the ones described by Hendy and Meuwissen, we were unable to find in the literature other cases of small bowel volvulus in patients with scleroderma.12,15 In the United States, the incidence of small bowel volvulus is considerable low, corresponding to 6.9% of all cases of small bowel obstruction in adults.16,17 Small bowel volvulus possesses a high risk for necrosis early on the course of the illness. Our patient had a non-necrotic volvulus with a slow progression of her condition, in the setting of systemic sclerosis and for which CT enterography, a different imaging technique, was required for proper diagnosis. CT enterography is a modification of conventional abdominal CT in which a neutral contrast like water or mild is used to cause small bowel distension followed by a rapid infusion of intravenous contrast. The enterography protocol is also for magnetic resonance imaging with similar efficacy. The goal is to achieve enough contrast between the enhanced small bowel wall and the lumen helping in the visualization of the entire small bowel anatomy and mesenteric fat.18 After reviewing the available literature and from our own experience, we believe that the decision to take these patients to surgery presents a formidable challenge. As suggested by our case series, the clinical manifestations and radiological assessment among mechanical obstruction and pseudo-obstruction are often indistinguishable, adding uncertainty to physicians aware of the adverse events if surgery is delayed in mechanical obstruction or promoting surgical intervention in those with pseudo-obstruction given risk of severe complications in those with SSc.
Intestinal obstruction and systemic sclerosis

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References