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Single Case

Chronic Intestinal Pseudo-Obstruction in Systemic Sclerosis: An Uncommon Presentation

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Chronic intestinal pseudo-obstruction · Pseudo-obstruction · Systemic sclerosis · Scleroderma

Abstract
Chronic intestinal pseudo-obstruction (CIPO) is an uncommon disorder characterized by the presence of bowel dilatation and abnormal motility. It is an important cause of chronic intestinal failure in patients with systemic sclerosis (SSc). Although intestinal pseudo-obstruction is an infrequent reason for hospitalization in these patients, it has been correlated with high in-hospital mortality compared to SSc patients hospitalized for other reasons as well as patients with intestinal pseudo-obstruction arising from other causes. Patients present with signs and symptoms of mechanical bowel obstruction, such as nausea, vomiting, constipation, abdominal pain, and abdominal distension, in the absence of an anatomic lesion blocking the movement of intestinal contents. Despite breakthroughs in our understanding of this disorder, these patients continue to be treated largely with organ-based symptomatic therapy. Unfortunately, despite treatment, they often experience decreased quality of life and impairment in their everyday lives. Here, we present an interesting case of an individual with SSc for many years who presented with signs and symptoms of CIPO.

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Introduction

Approximately 90% of individuals with either subtype of systemic sclerosis (SSc) – diffuse cutaneous SSc and limited cutaneous SSc – have some degree of gastrointestinal involvement [1, 2]. In fact, the digestive system is the organ system most commonly implicated in SSc. One-third to one-half of these individuals may not even experience symptoms [1–3]. However, 8% have gastrointestinal involvement that is severe, and mortality is elevated in these patients [3]. Any part of the gastrointestinal tract, from the mouth to the anus, can be affected, but the most common site of involvement is the esophagus, followed by the anorectum and the small intestine [3].

A dreaded complication in individuals with SSc is chronic intestinal pseudo-obstruction (CIPO), which is characterized by bowel dilatation and abnormal motility. It is a key cause of chronic intestinal failure in these individuals. In many cases, to differentiate between a pseudo-obstruction and a true mechanical obstruction is challenging [4]. Frequently, CIPO is an unclear diagnosis with limited therapeutic options. Therefore, our objective is to report a relevant case regarding an individual with SSc who presented with features of CIPO, as well as to provide a discussion on its clinical approach.

Case Presentation

A 52-year-old African American woman, with a past medical history of SSc diagnosed 5 years earlier and progressively worsening constipation refractory to treatment with multiple bowel regimens, presented to the Emergency Department with abdominal pain and distention, regurgitation, and intermittent vomiting for the last few weeks. She reported weight loss of roughly 10 pounds, from 115 to 105 pounds, over the last 1–2 months. She denied any recent symptoms of fevers, chills, diarrhea, blood in the stool, or black stools. She denied any recent change in her diet or any recent history of travel outside the United States. She reported that none of her family members had experienced similar symptoms. Her social history was remarkable for a former cigarette smoking history of 10 pack-years. She denied any alcohol use or illicit drug use. And, she denied any drug allergies. Two months ago, the patient had undergone esophagogastroduodenoscopy (afterward reported as normal) and colonoscopy (afterward reported a dilated lumen with no mucosal lesions).

On physical examination, the patient appeared emaciated and exhausted. She was afebrile (temperature 97.6°F) and hemodynamically stable (pulse rate 68 BPM, blood pressure 142/85 mm Hg). The abdomen was moderately distended with no tenderness. There was no guarding nor rigidity. Bowel sounds were decreased. Laboratory testing was performed and the comprehensive metabolic panel was remarkable for hypokalemia with potassium of 3.4 mEq/L (3.5–5.0 mEq/L) and hypochloremia with chloride of 94 mEq/L (96–110 mEq/L). The complete blood count was remarkable for hemoglobin of 9.6 g/dL (12.0–15.5 g/dL) with an unknown baseline, hematocrit of 29.4% (34–48%), and mean corpuscular volume of 102 fL (80–96 fL) – findings consistent with a macrocytic anemia. Thyroid function tests were within normal limits. She was later found to have several vitamin deficiencies.

Plain abdominal X-rays were performed and showed dilatation of the small and large bowel (Fig. 1). Computed tomography scan of the abdomen was performed and showed marked dilatation of the small and large bowel with air-fluid levels (Fig. 2). The gastric emptying study could not be performed due to intolerance and vomiting of the preparation.

The patient improved with medical treatment consisting of intravenous hydration, correction of electrolyte imbalances, and, later, total parenteral nutrition for a short time. Since then, with conservative treatment with prokinetic agents, laxatives, and occasional enemas, she has been able to tolerate small quantities of oral feeds several times each day but continues to have periods of severe constipation.

Discussion

SSc is a complex multisystem autoimmune disease that is described pathologically by variable fibrosis of the skin and internal organs, and extensive vasculopathy [5]. The disease is frequently complicated by small and large bowel involvement, characterized by reduced peristalsis and hypomotility with resulting stasis, intestinal dilatation, and, occasionally, CIPO.

In a large case-control study, Valenzuela et al. [6] attempted to measure the burden of intestinal pseudo-obstruction among patients with SSc by investigating its prevalence and outcomes, with the use of the Healthcare Cost and Utilization Project Nationwide Inpatient Sample for the period 2002–2011. They discovered that, even though intestinal pseudo-obstruction is an uncommon reason for hospitalization in these patients, it has been correlated with high in-hospital mortality compared to SSc patients hospitalized for other reasons as well as patients with intestinal pseudo-obstruction arising from other causes. Furthermore, from this large United States-based hospitalization database, they discovered that (1) the lifetime prevalence of intestinal pseudo-obstruction among SSc patients was 5.4%, (2) SSc patients with intestinal pseudo-obstruction had a greater tendency to receive parenteral nutrition as well as to die, and (3) intestinal pseudo-obstruction in SSc was correlated with death in more than 7% of hospitalizations for this indication.

The pathogenesis of SSc is complex and involves the activation of the immune system, vascular injury, and excessive synthesis and accumulation of collagen in the connective tissue [7, 8]. These processes are all recognized to be essential in the development of the disease. A fundamental hypothesis regarding the pathophysiology of gastrointestinal involvement in SSc consists of four stages: vasculopathy, neural dysfunction, smooth muscle atrophy, and tissue and muscle fibrosis [9]. The earliest pathology is frequently characterized by vasculopathy with minor changes in intestinal permeability, transport, and absorption. This stage is followed by neural dysfunction, in which the individual begins to develop clinical symptoms, such as dysphagia, heartburn, hoarseness, and bloating. At this point, prokinetic agents can largely reverse the functional abnormalities. The vasculopathy and neural dysfunction then lead to the third stage of smooth muscle atrophy, which is characterized by only partial response to medications. Lastly, the fourth stage is tissue and muscle fibrosis, when medications are no longer effective. In severe cases, disruption of small and large bowel motility can progress to CIPO over time.

In the diagnostic evaluation of a patient who presents with chronic symptoms of vomiting, constipation, abdominal pain, and abdominal distension – especially, in patients with SSc – physicians should consider CIPO as a diagnosis. A plain abdominal X-ray should be performed, and electrolyte imbalances and hypothyroidism should be excluded. A computed tomography scan of the abdomen should be performed to rule out a mechanical cause [3].

Treatment for CIPO includes prokinetic agents, such as erythromycin and metoclopramide, as well as laxatives and occasional enemas. In severe cases, conservative management with bowel rest, nasogastric decompression, and intravenous hydration is used. Octreotide, a long-acting somatostatin analogue, has been demonstrated to have some benefit in

refractory cases [10]. However, when octreotide is administered during the daytime with meals, it significantly delays gastric emptying and small bowel transit of solids, which may, in fact, be harmful to individuals with CIPO [11, 12]. Therefore, it is advised that, if octreotide is used to treat CIPO, it should be administered at bedtime. Surgical intervention in CIPO is frequently complicated by prolonged ileus and, so, is avoided when possible [3].

In short, CIPO is an uncommon but severe complication of SSc. Conversely, gastrointestinal involvement of any form is relatively common and appears early in the natural history of the disease. Physicians should not depend on patients' symptoms to make an early diagnosis, as one-third to one-half of individuals with some degree of gastrointestinal involvement are asymptomatic. Rather, physicians should properly question their patients regarding potential gastrointestinal involvement and begin treatment early before irreversible impairment occurs. Unfortunately, despite treatment, patients often experience decreased quality of life and impairment in their everyday lives. Further investigation is needed to develop therapies targeted at improving intestinal motility in SSc.

Statement of Ethics

The authors have no ethical conflicts to disclose.

Disclosure Statement

The authors of this case report declare that they have no competing interests. The authors declare that they have no affiliations with or involvement in any organization or entity with any financial interest or nonfinancial interest (such as personal or professional relationships, affiliations, knowledge, or beliefs) in the subject matter discussed in the manuscript. The authors of this case report declare that no financial support nor grant support has been received for the preparation of the manuscript.

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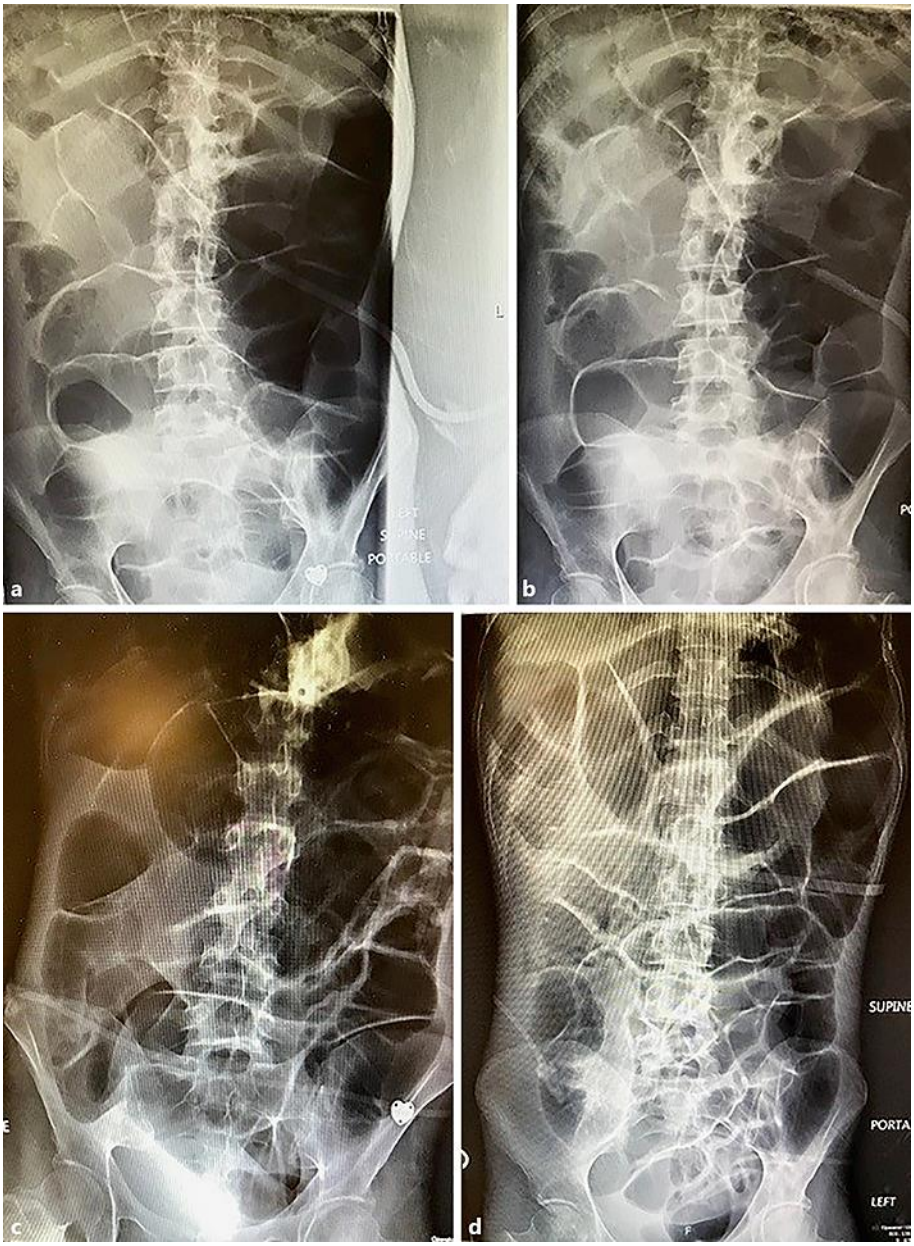

Fig. 1. a–d Chronic intestinal pseudo-obstruction. There is dilatation of the small and large bowel, consistent with pseudo-obstruction.

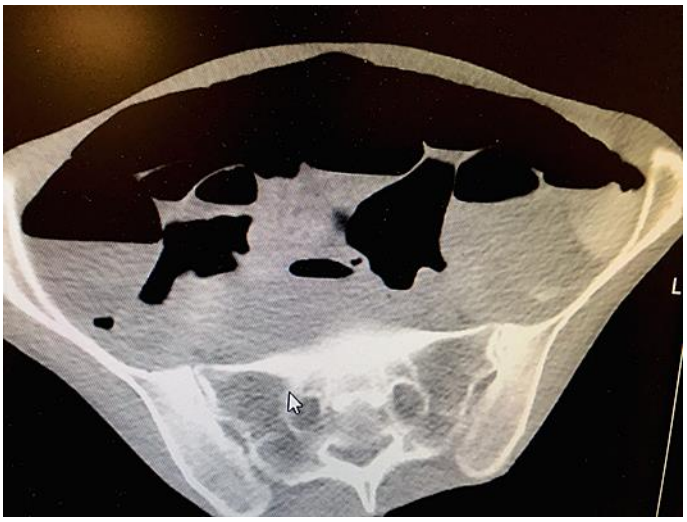

Fig. 2. Chronic intestinal pseudo-obstruction. There is marked dilatation of the small and large bowel, as well as multiple air-fluid levels, consistent with pseudo-obstruction.