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CASE REPORT

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Systemic sclerosis with chronic megacolon complicated by bowel obstruction: a case report

Zhe-Jia Liu[1], Jung-Chun Lin[2] and Yi-Chiao Cheng[3*]

Abstract

Background Systemic sclerosis (SS) is a systemic connective tissue disease characterized by changes in the collagen structure that lead to fibrosis of the skin and internal organs. The gastrointestinal (GI) tract is most commonly affected, and it significantly impacts the daily lives of patients. A fatal complication of GI involvement is chronic megacolon, which has been detected in only a few patients in recent years.

Case presentation A 51-year-old female with a 10-year history of poorly controlled SS had suffered constipation, abdominal bloating, vomiting, and fever for two weeks and eventually developed chronic megacolon. Conservative treatments and medications failed to relieve the symptoms and resolve the condition. The patient ultimately required surgical intervention. This case highlights the importance of timely evaluation and management of SS-related GI complications.

Conclusions Procedures such as colonoscopy are instrumental in monitoring disease progression and providing therapeutic relief. Frequent physical examinations may help predict the failure of conservative approaches and guide clinicians toward surgical interventions to prevent life-threatening complications.

Keywords Systemic sclerosis, Megacolon, Constipation, Chronic intestinal pseudo-obstruction

Background

Systemic sclerosis (SS) is an autoimmune disorder that affects the structure of normal collagen and causes fibrosis of the skin and other organs. There are two main classifications of SS. Cases involving only the skin are classified as limited SS, and those involving the internal organs are classified as diffuse SS. The gastrointestinal (GI) tract is the most commonly affected organ. Approximately 90% of patients experience symptoms that can impact any part of the GI tract, and the esophagus is the most frequently affected region [1]. GI involvement is the third most common cause of mortality after cardiopulmonary and renal involvement in patients with SS [2]. GI tract involvement can impair oral intake, cause fecal continence or functional obstruction, and severely affect the quality of life of patients. Therefore, disease control is an essential component of treatment. We present the case of a 51-year-old woman with poorly controlled SS complicated by interstitial lung disease and GI tract involvement that eventually progressed to chronic megacolon. Despite

*Correspondence:
Yi-Chiao Cheng
ndmcjoe@gmail.com
[1]School of Medicine, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan
[2]Division of Gastroenterology, Department of Internal Medicine, Tri-Service General Hospital, National Defense Medical Center, Taipei, Taiwan
[3]Division of Colon and Rectal Surgery, Department of Surgery, Tri-Service General Hospital, National Defense Medical Center, Number 325, Section 2, Chenggong Rd., Neihu Dist., Taipei City 114202, Taiwan

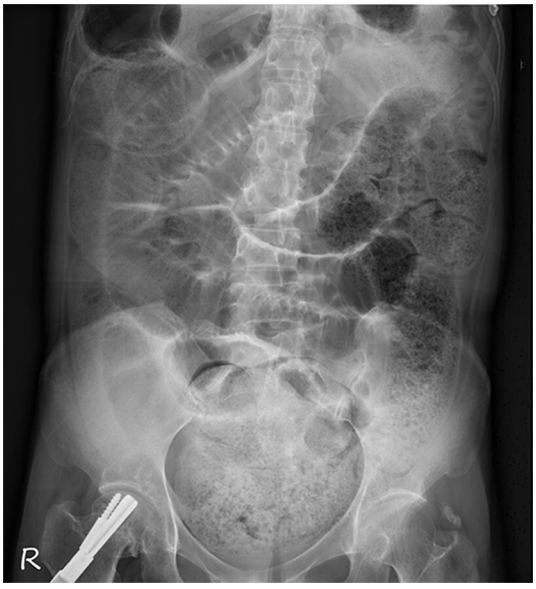

Fig. 1 Abdominal radiograph obtained at presentation. Severe bowel obstruction and internal fixation of the right femur are observed

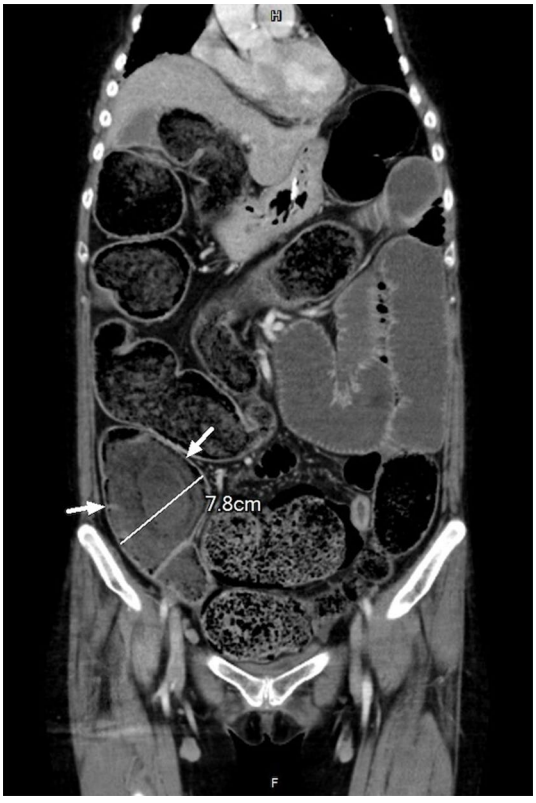

Fig. 2 Computed tomography image obtained at presentation. Stool impaction with bowel loop distention and dilation (7.8 cm) of the cecum region (arrow) are observed

conservative treatment, including decompression and prokinetic drugs, surgical intervention was necessary.

Case presentation

A 51-year-old woman with a history of poorly controlled SS for more than 10 years that was complicated by interstitial lung disease presented to the emergency department with constipation, abdominal bloating, vomiting, and fever that had persisted for 2 weeks. A physical examination revealed sinus tachycardia and abdominal tenderness; however, rebounding pain was not observed. Blood tests revealed increased lactate (2.8 mmol/L) and C-reactive protein (25.88 mg/dL) concentrations. Abdominal radiography revealed severe bowel obstruction (Fig. 1), and subsequent computed tomography findings suggested stool impaction with bowel loop distention (Fig. 2). The diagnosis of megacolon was confirmed after ruling out malignancy using imaging and colonoscopy, which did not reveal luminal narrowing. The patient preferred to avoid surgery. Hence, conservative treatment comprising nasogastric tube placement and anal decompression was performed, resulting in the drainage of a stool-like substance. The patient was admitted to our hospital because megacolon was suspected.

During admission, the patient received empirical antibiotics, peripheral parenteral nutrition, gastric decompression, and anal decompression. A rheumatologist was consulted for the poor control of SS; however, no treatment was suitable for acute illness. Sigmoidoscopy performed on day 5 after admission revealed normal mucosa with fecal bezoar impaction in the region comprising sigmoid and uterine prolapse. On day 10 of admission, the bowel obstruction and stool impaction gradually worsened, and fecal matter began draining from the mouth of the patient and nasogastric tube. Additionally, episodes of suspected fecal aspiration and intermittent fever were observed, and indigestion and electrocyte imbalance became more severe. Laboratory data revealed sodium, potassium, chloride, magnesium, free calcium, and hemoglobin concentrations of 133 mmol/L, 3.4 mmol/L, 95 mmol/L, 1.7 mg/dL, 4.63 mg/dL, and 6.9 g/dL, respectively. Total colectomy was performed on day 15 after admission following a discussion of the case with the family of the patient (Fig. 3). Due to impaired gastric motility, we also inserted a nasal-duodenal tube, which facilitates bypassing the stomach and reduces the risk of gastroesophageal reflux and aspiration. Pathological examination of the resected colon revealed flattened colonic mucosa with atrophy, lymphoepithelial and granulocytic epithelial lesions, *Melanosis coli*, and mild subserosal fibrosis, which were consistent with chronic megacolon. The patient was admitted to the intensive care unit for weaning and rehabilitation after surgery. Her digestive ability improved, and her electrocyte

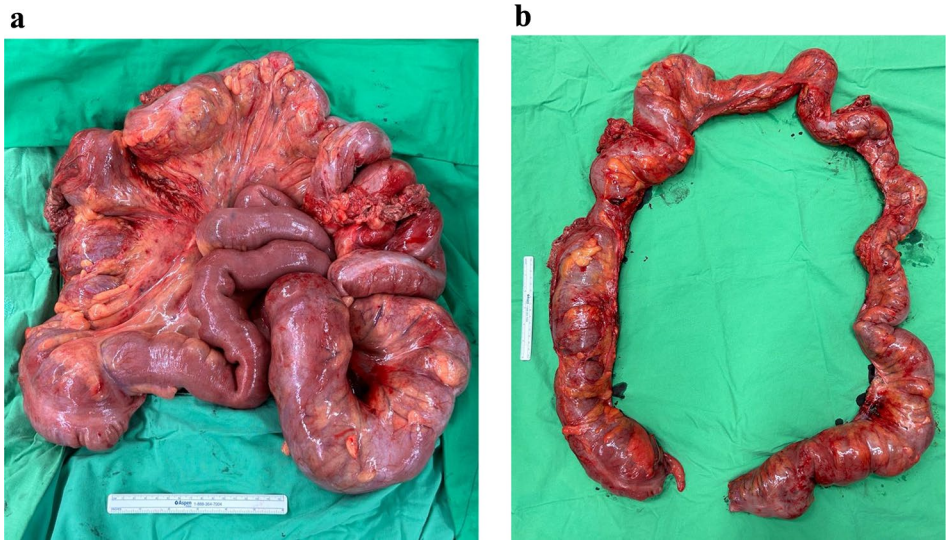

Fig. 3 Surgical findings. **(a)** Severe dilation of the entire colon is observed. **(b)** The resected colon has a length of 225 cm, circumference of 9.2 cm, and wall thickness of 0.2 cm. A 15-cm ruler is shown for scale

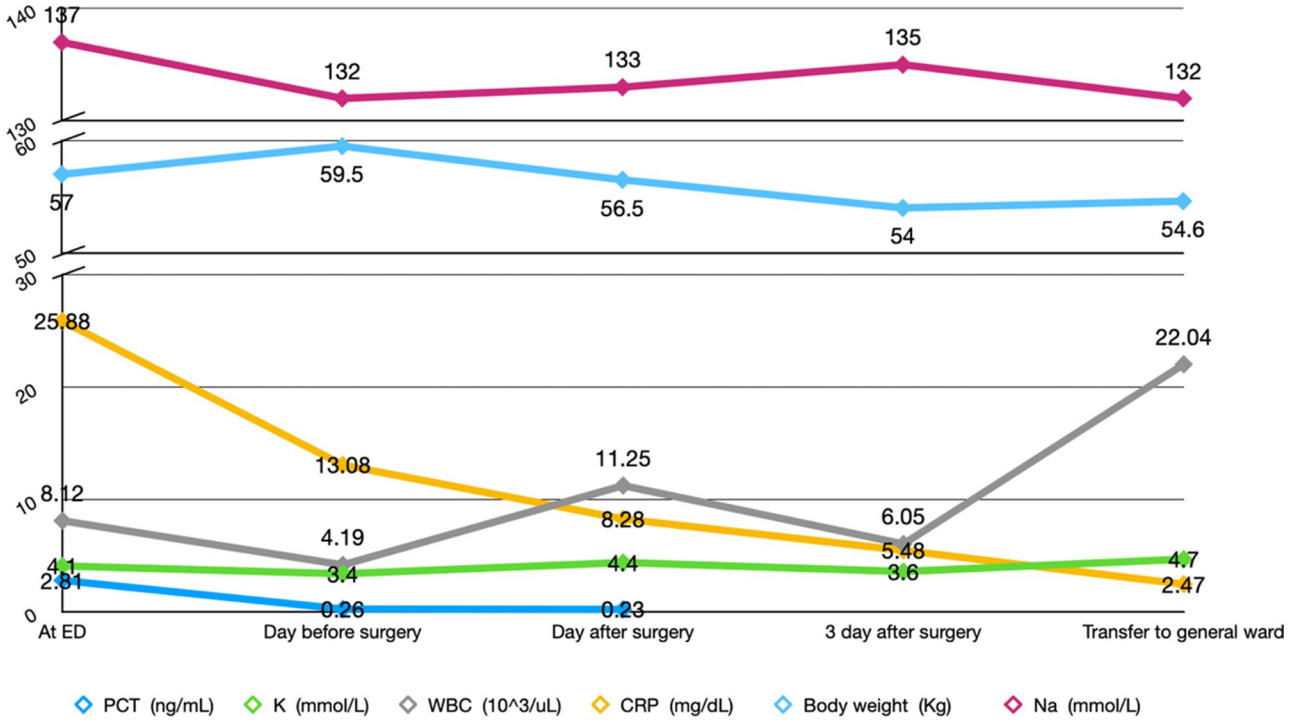

Fig. 4 Changes in blood chemistry and body weight during the perioperative period. The perioperative changes in blood chemistry are shown, including sodium (Na), potassium (K), white blood count (WBC), C-reactive protein (CRP), and procalcitonin (PCT). ED, Emergency department

imbalance was corrected after surgery. Therefore, she was transferred to the general ward 1 month later. Figure 4 shows the changes in blood chemistry and body weight during the perioperative period. However, she experienced pneumonia in the general ward, which was potentially linked to the underlying interstitial lung disease. Empirical antibiotics were administered for infection control. The patient was discharged 2 months later with a relatively stable condition.

Discussion and conclusions

The patient's treatment for SS before this admission included nonsteroidal anti-inflammatory drugs, low-dose steroids, and hydroxychloroquine, primarily for managing her SS-related arthritis. However, her medication

Table 1 Clinical details of reported cases of systemic sclerosis complicated by megacolon

	Age (year)/Sex	Disease history	Surgical intervention	Outcome
Shamberger et al. [8]	58/M	Newly diagnosed SS	Prophylactic resection because of perforation	Disease symptoms did not resolve after surgery
Ferreiro et al. [10]	59/F	Diagnosis of SS via necropsy	Conservative medication only	Died because of renal failure and heart failure
Norton et al. [11]	49/F	Newly diagnosed SS	Surgery to relieve symptoms	Discharged nearly 2 months later
Han et al. [12]	27/F	SS for 5 years	Conservative medication only	Required daily laxatives

F, female; M, male; SS, systemic sclerosis

adherence was poor. The diverse symptoms of SS among patients and lack of reliable objective measurement tools make it difficult to effectively assess and manage the progression of the disease. Severe manifestations include dysphagia, bacterial overgrowth in the small intestine, malnutrition, chronic intestinal pseudo-obstruction, and constipation, which can be life-threatening. Some studies have shown that risk factors for severe complications involving the GI tract, which may require supplemental nutrition, include male sex, myopathy, and sicca symptoms [3, 4]. Additionally, patients with severe GI dysmotility have a higher mortality rate than those with other mild GI symptoms [4]. Therefore, timely management of SS is very important.

The patient in the present case was not male, but she required supplemental nutrition because of poor disease control; males typically have higher risks of severe complications. SS affects the structure of the small bowel and colon and causes malnutrition and colonic dysfunction. This dysfunction, which typically manifests as constipation, is a common complication of SS occurring in approximately 50% of patients. Chronic pseudo-obstruction, which is a severe complication of colonic hypomotility, may develop in less than 10% patients and can significantly affect mobility and motility [5]. Studies have shown that the physiological process underlying colonic dysfunction can be divided into the following three steps: neural dysfunction, smooth muscle atrophy, and fibrosis [2, 3, 6]. Prokinetic drugs may effectively improve GI motility during neural dysfunction. However, the efficacy of these agents diminishes as colonic dysfunction progresses. Further, these agents are considered ineffective after fibrosis occurs. This physiological process is consistent with that observed in our patient.

The treatment for internal organ involvement in SS is organ-specific [7]. We prescribed medications such as stool softeners, prokinetic agents, and enemas for GI dysmotility; however, they were ineffective. Poor disease control and the ineffective drug treatment contributed to the eventual development of megacolon. Megacolon can be acute or chronic; however, both forms can progress to toxic megacolon if not well managed. The primary causes of toxic megacolon are systemic toxicity and severe colonic dilation [8]. Our patient developed fever, leukocytosis, an increased C-reactive protein concentration, anemia, and electrocyte imbalance. Additionally, computed tomography revealed dilation (7.8 cm) of the cecum region. The goal of chronic megacolon treatment is the prevention of toxic megacolon and avoidance of surgical intervention, if indicated. Surgery is a last resort if conservative therapy fails. We attempted decompression and administered medications; however, the patient had previously responded poorly to conservative therapy, and timely surgery was necessary.

SS complicated by megacolon is rare, and only a few case reports exist in the literature [9–13]. We reviewed the treatments and timing of surgical intervention described by those reports (Table 1). Controlling SS is important because it is associated with fatal complications, such as interstitial lung disease and chronic pseudo-intestinal obstruction, as in our case. Interstitial lung disease can cause poor coughing ability, resulting in pneumonia, which increases the risk of relapsing infection. However, chronic pseudo-intestinal obstruction can lead to malnutrition and electrolyte imbalance when the small intestine is affected and constipation that may progress to megacolon when the colon is involved. Both complications can be effectively avoided if SS is controlled with medication. Timely surgery may improve patient outcomes. The pathological findings of total colectomy performed for our patient revealed fibrosis of the colonic smooth muscle, which was consistent with the pathophysiology of colonic dysfunction, for which prokinetic agents can become ineffective. However, universal guidelines regarding the optimal timing of surgical intervention have not been established. Therefore, careful monitoring of the vital signs and repeated physical examinations are recommended to prevent progression to toxic megacolon for patients with severe colonic dysfunction associated with SS.

In conclusion, we encountered a rare case of SS that progressed to megacolon and ultimately required surgical intervention. Early identification of the optimal timing of surgery to prevent fatal complications is necessary.

Abbreviations

GI Gastrointestinal
SS Systemic sclerosis

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Author contributions
ZJL: Data collection and analysis and Writing–original draft; JCL: Clinical care of patient; YCC: Clinical care of patient and Writing–review and editing. All authors contributed to the article and approved the submitted version.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Informed consent was not required, and this study was approved by the Institutional Review Board of the Tri-Service General Hospital, National Defense Medical Center (Taipei, Taiwan; approval number: TSGHIRB no. B202415198).

Consent for publication
Not applicable.

Competing interests
The authors declare no competing interests.

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