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CASE REPORT

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A case of systemic sclerosis sine scleroderma associated with perforation of an afferent loop after subtotal gastrectomy with Billroth 2 anastomosis for its severe gastrointestinal involvement

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Abstract A 50-year-old man was admitted to hospital for dysphagia. The upper gastrointestinal series revealed esophageal stricture, pyloric stenosis, and hypomotile small intestine. He was diagnosed with systemic sclerosis sine scleroderma with gastrointestinal involvement. After subtotal gastrectomy with Billroth 2 anastomosis, he had recurrent intestinal pseudo-obstruction and perforation of the afferent loop. Our experience indicates that surgical procedures in bowel scleroderma, in which an afferent loop is reconstructed, could easily cause perforation of the afferent loop.

Key words Afferent loop · Esophageal stricture · Perforation · Pyloric stenosis · Systemic sclerosis sine scleroderma (ssSSc)

Introduction

Gastrointestinal (GI) involvement in systemic sclerosis (SSc) is common and may precede cutaneous manifestation.[1] Systemic sclerosis sine scleroderma (ssSSc), systemic sclerosis without scleroderma associated with organ system involvement, is proposed as a distinct category of SSc.[2] We describe a case of ssSSc associated with perforation of an afferent loop after subtotal gastrectomy with Billroth 2 anastomosis.

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Case report

A 50-year-old man was admitted to our hospital for dysphagia in May 2000. The patient had been suffering from dysphagia for 1 year with a decrease in body weight from 62.0 to 54.5 kg. At the age of 17 years, he had suffered from recurrent duodenal ulcers and had been told by his doctor that his duodenum was very hypomotile on upper GI series. On physical examination there were no abnormal findings with respect to his vital signs, skin, chest, or abdomen. Urinalysis, hematologic and serum chemistry evaluations, including total protein, lipid, and electrolytes, were normal except for positive results for the antinuclear antibody (ANA); 320-fold (normal range <40) and the anti-centromere antibody; 173 (normal range <10.0). Gastrointestinal endoscopy showed severe stricture of the distal esophagus with esophageal ulcers. On the upper GI series, the lesion was estimated to be 5 mm in internal diameter and 9 cm in length (Fig. 1). Retention of barium in the stomach due to gastroparesis and pyloric stenosis was also detected (Fig. 2). The duodenum and jejunum showed edematous, dilated loops and pooling of barium (Fig. 3). He was suspected as having gastrointestinal involvement of systemic sclerosis and treated with endoscopic balloon dilatation, which dilated the esophageal lesion to 10 mm in internal diameter, and additional medications of omeprazole and cisapride. Gastrointestinal endoscopy revealed no abnormal findings in the stomach. Dysphagia improved for 3 months until recurrence of the esophageal stricture. In January 2001, after the next balloon dilatation, he underwent subtotal gastrectomy with Billroth 2 anastomosis. The duodenum and jejunum were reconstructed as an afferent loop. Histopathological analysis revealed thinning of the muscularis propria without fibrosis in the duodenal bulb. Although dysphagia was subsequently alleviated, abdominal distension or diarrhea occurred repeatedly after the operation. He had Raynaud's phenomenon for the first time in October 2001, 2 years after the initial abdominal symptoms.

In August 2002, the patient was admitted for vomiting. His body weight was 45.0 kg. The abdomen was distended

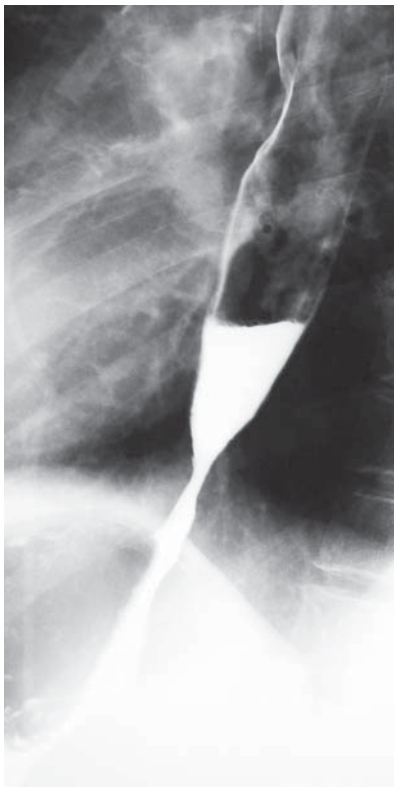

Fig. 1. The upper gastrointestinal series revealed severe stricture at the distal esophagus

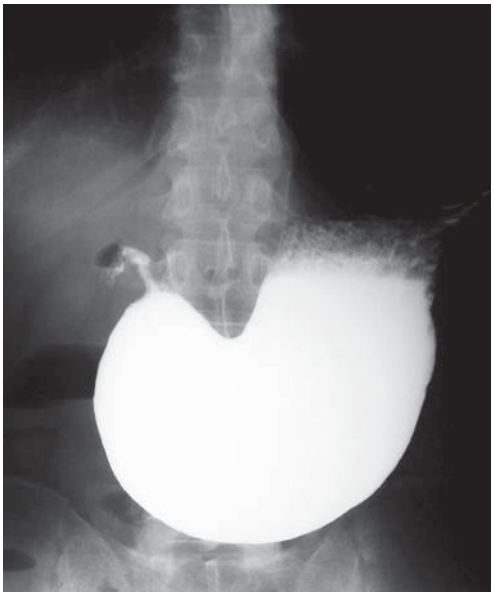

Fig. 2. Retention of barium in the stomach due to gastroparesis and pyloric stenosis was detected

with decreased bowel sounds. Computed tomography of the abdomen showed dilatation of small intestine, which implied intestinal pseudo-obstruction (IPO). His condition recovered within 1 week with nasogastric suction and bowel rest. One month later, he again had IPO and 3 days after treatment, he had sudden abdominal pain with shock. Computed tomography of the abdomen revealed intra-abdominal ascites, and an emergency operation was performed. Perforation of the afferent loop was detected; resection of the affected site of the jejunum and fistulization were performed. Histopathological analysis of the jejunum (at a site distinct from the perforation) showed edematous thickening of the submucosa and thinning of the muscularis propria without fibrosis (Fig. 4). The patient died of intra-abdominal bleeding due to dehiscence of the suture line in November 2002. On autopsy, muscle fibrosis was detected only at the middle and distal esophagus but not in other regions of the bowel (Fig. 5). There was no SSc-related organ involvement except for small lung fibrotic lesions.

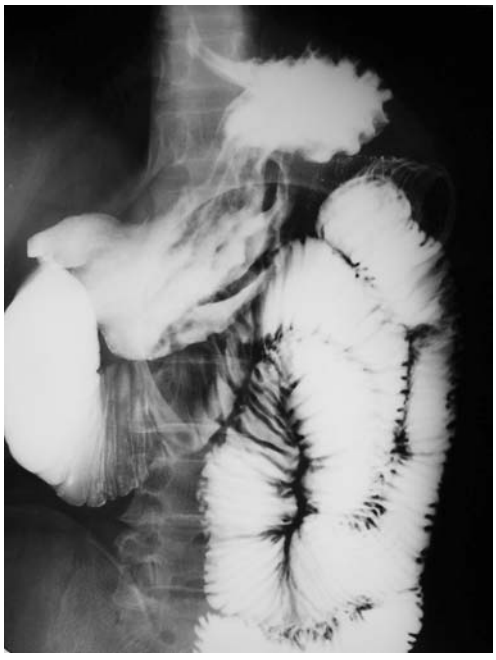

Fig. 3. The duodenum and jejunum showed edematous, dilated loops, and pooling of barium

Discussion

Gastrointestinal involvement occurs in up to 90% of patients with SSc.[1] This includes gastroesophageal reflux disease, intestinal pseudo-obstruction, malabsorption, and constipation.[1] Intestinal perforation was also found in 7 of 16 autopsy cases with SSc.[3] Our case was diagnosed as ssSSc based on GI involvement, positive ANA, and Raynaud's phenomenon according to the proposed criteria.[4] In 48 cases of ssSSc, GI involvement was observed in 31/39 (79%), while pulmonary, cardiac and renal involvement occurred in 32/47(68%), in 4/45 (9%), and in 0/48 (0%), respectively.[4]

In our case, esophageal stricture was associated with pyloric stenosis and severe gastrointestinal hypomotility, and endoscopic procedures and medications as first-line treatments[5] were not effective. We finally made the decision

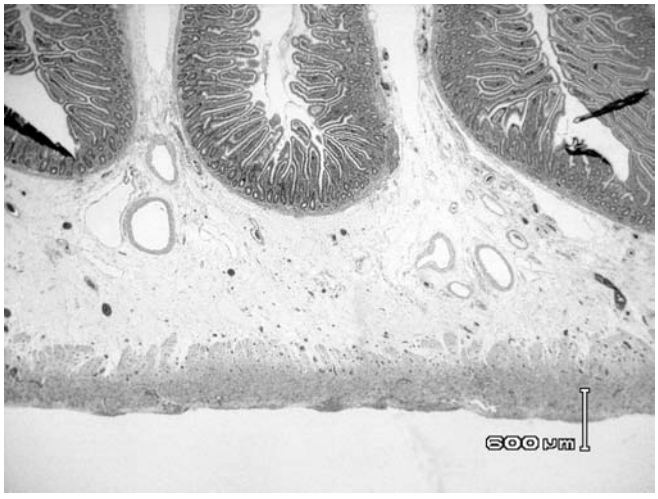

Fig. 4. Histopathological analysis of the jejunum (at a site distinct from the perforation) showed edematous thickening of the submucosa and thinning of the muscularis propria without fibrosis (H&E stain, ×2)

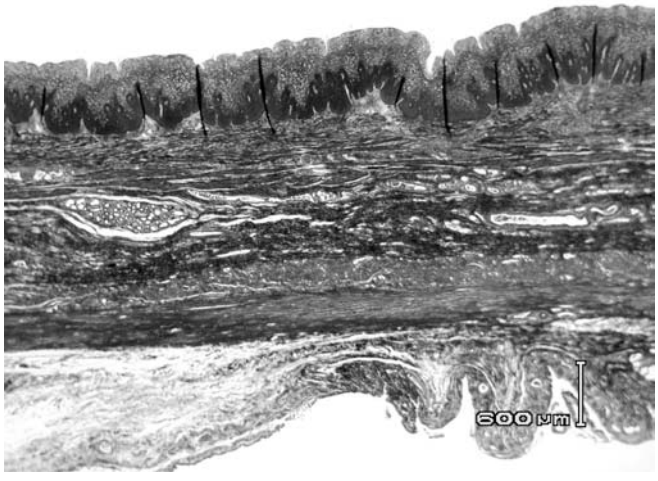

Fig. 5. Histopathological analysis of the middle esophagus revealed fibrotic lesion involving the submucosa and the muscularis propria (Azan stain, ×2)

that the patient should undergo the operation on pyloric stenosis, considering the adverse effects of pyloric stenosis and gastroparesis on gastroesophageal reflux. In a study of 14 patients with SSc, all of eight patients with prolonged gastric emptying also had delayed esophageal transit, and there was a significant correlation between esophageal transit and gastric emptying.[6] After the operation, our patient suffered from IPO followed by a fatal perforation of the afferent loop. We speculate that the rise in intraintestinal pressure at the afferent loop might have led to the perforation. In general, partial bowel resection in SSc is rarely indicated, as the disease is almost always generalized, and is often discouraged for a prolonged ileus.[5] We cannot justify our choice of operation, which might have caused IPO and perforation. Although we do not have any evidence to support other procedures, subtotal gastrectomy with Billroth 1 anastomosis or antireflux procedures such as Nissen fundoplication[5] might have been alternatives.

On autopsy, fibrosis was detected only in the distal esophagus, while thinning of the muscularis propria without fibrosis and atrophy of Meissner's plexus were found in the jejunum. In general, early GI lesions induced by SSc are neural dysfunction, causing smooth muscle atrophy; muscle fibrosis is the final lesion.[1] It was surprising that severe GI involvement in our case was not mainly due to muscle fibrosis. It has recently been suggested that functional antibodies against M3-muscarinic receptor-mediated enteric cholinergic neurotransmission may present in the serum of SSc patients.[7]

Although it is difficult to manage severe GI involvement of SSc, our experience indicates that surgical procedures in this disorder, in which an afferent loop is reconstructed, could easily cause perforation of the afferent loop.

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