Mod Rheumatol (2004) 14:419–421
DOI 10.1007/s10165-004-0334-1

CASE REPORT

Takafumi Chin · Kou Mochizuki · Hidetugu Murakami
Hidenori Tajima

A case of systemic sclerosis with abdominal surgery for stenosis of the terminal ileum

Received: February 14, 2004 / Accepted: June 7, 2004

Abstract A 71-year-old woman was admitted to our hospital with prominent abdominal distention. The diagnosis of systemic sclerosis (SSc) was made based on the sclerosis on her face, forearms, fingers, and toes as well as pitting ulcers on fingers and toes. After conservative therapy, she presented more severe signs of peritoneal irritation. Abdominal surgery was performed, revealing stenosis at the terminal ileum. Pathologically, this region showed muscularis propia becoming fibrotic. She was discharged from the hospital, returning about once a month as an outpatient.

Key words Abdominal operation · Peritoneal irritation · Pseudo-obstruction · Systemic sclerosis (SSc)

Introduction

Systemic sclerosis (SSc) is a systemic disease characterized by excessive deposition of collagen and other matrix elements by fibroblasts in the skin and sometimes in multiple internal organs. It is associated with prominent, often severe alterations of microvasculature, autonomic nervous system, and immune system. Clinically, symptomatic and significant gastrointestinal involvement occurs in approximately 50% of all patients with SSc.

Case report

A 71-year-old woman was admitted to our hospital with prominent abdominal distention and abdominal pain in June 2003. She was suffering from recurrent diarrhea, constipation, and subileus. SSc was diagnosed in 1993 based on sclerosis on her face, forearms, fingers, toes, and chest. The fourth toe of the right foot came off naturally. She underwent bypass surgery for occlusion of the left iliac artery in 2000. On admission, she presented with slight signs of peritoneal irritation. Laboratory data revealed a white blood cell count of 11 400/μl, C-reactive protein 2.56 mg/dl, and antinuclear antibody 235.8 index; the anti-DNA antibody assay was negative, as were the assays for anti-topoisomerase I antibody and anti-centromere antibody.

Abdominal roentgenograms showed dilation of the small intestine and an air-fluid level. Pneumatosis cystoides intestinalis and intraabdominal free air were not apparent. Computed tomography (CT) showed dilation of small intestine and tapering or occlusion of the terminal ileum (Figs. 1, 2). Enhanced CT showed no sign of embolus or dissection of the superior mesenteric artery.

After conservative therapy with nasogastric suction, parenteral hyperalimentation, and macrolide antibiotics, she presented with more severe signs of peritoneal irritation. There were no signs of intraabdominal free air by CT, but an emergency abdominal operation was performed for a diagnosis of strangled ileus. Iliocecal resection and ileocolostomy was performed, revealing stenosis at the terminal ileum (Figs. 3, 4).

Pathologically, this region showed the muscularis propia becoming fibrotic. The inner circular layer of the muscularis propia showed atrophy and fibrosis, but its outer, longitudinal layer did not exhibit fibrosis (Fig. 5). After the operation, an intestine–skin fistula was seen transiently with incomplete anastomosis. Temporary local care of the intestinal fistula[1] and parenteral hyperalimentation were provided. Over a period of a month the discharge from the intestine–skin fistula gradually decreased. The fistula finally closed, and good bowel movement was recovered. She was discharged from the hospital but returns about once a month as an outpatient.

T. Chin[1] (✉) · K. Mochizuki · H. Murakami · H. Tajima
Division of Surgery, Higashi Totsuka Memorial Hospital, 548-7 Shinano-cho, Totsuka-ku, Yokohama 244-0801, Japan

Present address:
[1] Department of Plastic and Reconstructive Surgery, Nippon Medical School Hospital, 1-1-5 Sendagi, Bunkyo-ku, Tokyo 113-8603, Japan
Tel. +81-3-3822-2131/Fax +81-3-5685-3076
e-mail: chin@peach.plala.or.jp

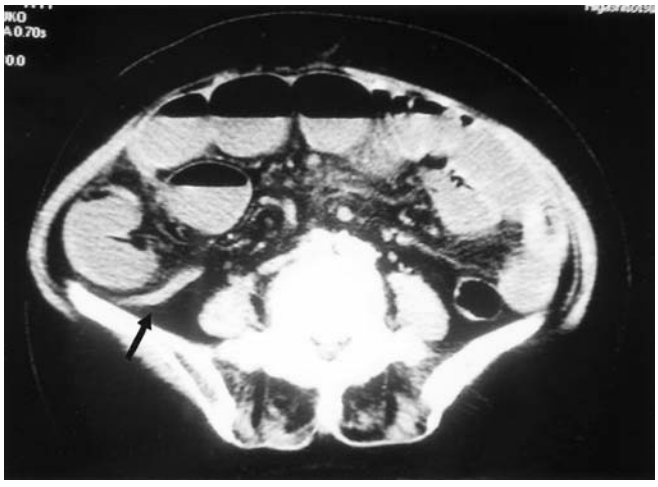

Fig. 1. Computed tomography (CT) scan (first medical examination) of the emptied ascending colon (*arrow*)

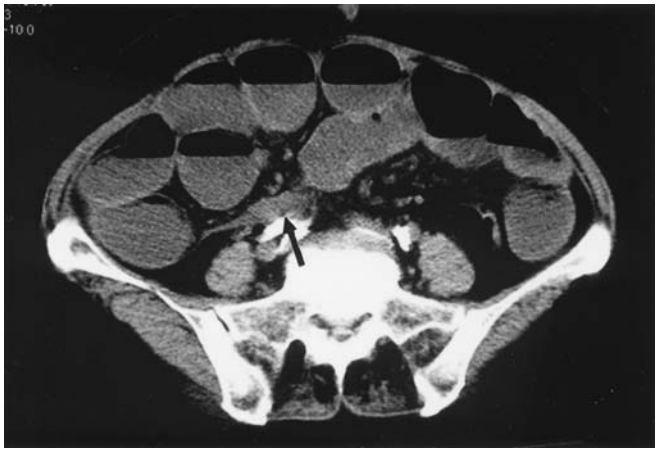

Fig. 2. CT scan (first medical examination) of the stenosis of the terminal ileum (*arrow*)

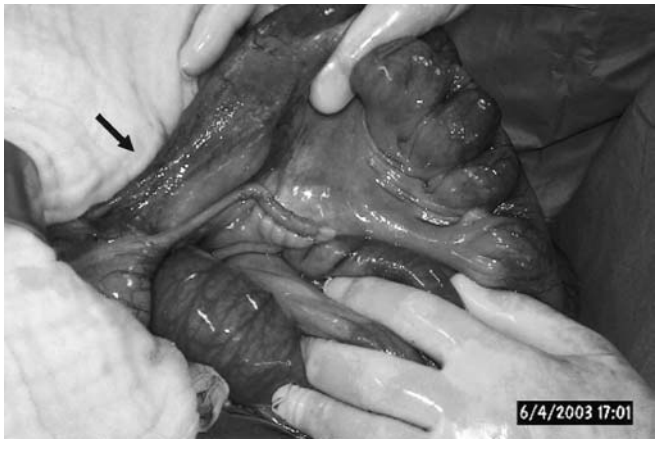

Fig. 3. During the operation an obvious stenosis in the iliocecal region became apparent (*arrow*). It was presumed that it was the cause of the mechanical ileus

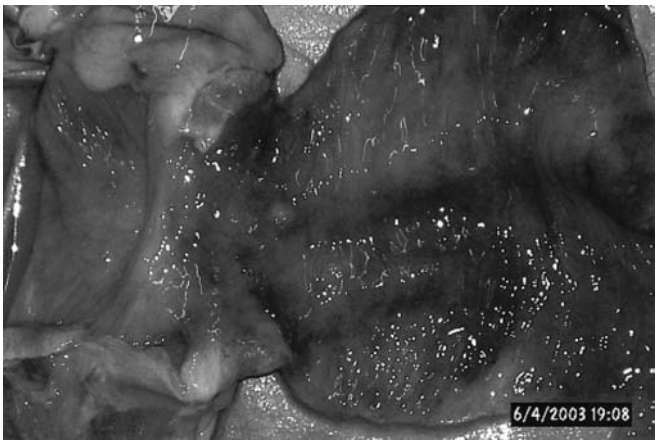

Fig. 4. Specimen. There were flat Kerckring's folds on the intestinal mucous membrane and a stricture in the terminal ileum. There was also an ulcer (0.5 × 0.6 cm) in the ileum

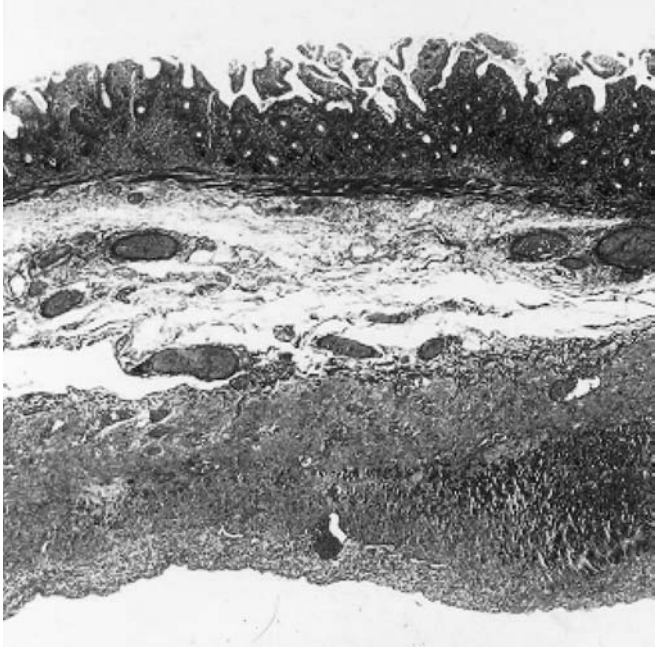

Fig. 5. There is fibrous replacement in the muscularis propia. The inner circular layer of the muscularis propia showed atrophy and fibrosis, whereas the outer longitudinal layer showed no fibrosis. These findings are compatible with the pathology of SSc. Azan stain

Discussion

Clinically symptomatic and significant gastrointestinal (GI) involvement occurs in approximately 50% of all patients with SSc. Many patients with SSc who do not have GI symptoms have subclinical GI involvement. Careful studies have revealed that as many as 75%–90% of patients with SSc have abnormalities noted on esophageal motility testing, although in many the involvement is not clinically significant.

Involvement of the anorectum is the next most frequent problem, occurring in 50%–70%. Small bowel hypomotility, known as scleroderma bowel, has been observed in 40% and colonic involvement in 10%–50%.[2] Scleroderma bowel is a common, life-threatening manifestation in patients with SSc. Once an advanced manifestation of scleroderma bowel occurs as a so-called intestinal pseudoobstruction, hospitalization is often required and a conservative approach to management is taken. This regimen includes nasogastric suction, bowel rest, antibiotics for bacterial overgrowth, and dietary supplements.

For chronic intestinal pseudoobstruction in early SSc, prokinetic drugs such as cisapride,[3] octreotide,[4] and macrolide antibiotics[5] are the treatment of choice. For chronic intestinal pseudo-obstruction in late SSc, the gastrointestinal muscle becomes refractory, and there is dramatic hypomotility. There is no contractile response of the stomach or the small intestine to a meal. At this stage, the intestinal myoelectric response to the regulatory hormones secretin and gastrin is impaired. In addition, while endogenous cycling of motilin and pancreatic polypeptide continues in patients with SSc, the levels of both substances are increased, suggesting end-organ refractoriness.[6]

Symptoms at this stage are often resistant to the effects of drugs compared to the early stage, and surgery is the only option other than nutritional support. Pathologically, the mucosa is characteristically normal, and the striking abnormality is in the muscularis propia, which shows extensive smooth muscle atrophy and replacement by fibrous tissue in the GI tract.[7] These changes are more severe in the circular layer than in the longitudinal layer. Based on these findings, although several causes were conjectured, it is valid to think that the stenosis in our patient originated from the SSc. According to a study of the pathophysiology of SSc bowel, such muscle atrophy and fibrosis lead to loss of the migratory motor complex.[6,8]

When surgical resection is considered, careful localization of the atonic segment to conserve the intestine and to ensure that newly apposed segments are functional provides good results. Inappropriate surgical intervention, however, which may aggravate the ileus and delay recovery even further, should be strongly discouraged. Patients at a late stage tolerate surgery poorly, and a conservative stance tends to be taken, even when surgery is considered. For nutritional support, parenteral hyperalimentation is usually required, although long-term hyperalimentation with SSc has been associated with complications and death rates as high as 40%.[9]

In a patient with SSc, a mechanical obstruction, as was seen in our patient, may be confused with severe pseudo-obstruction. The differential diagnosis of these two entities can be difficult. Patients with a pseudo-obstruction do not usually have fever, leukocytosis, or peritoneal irritation. In general, intraabdominal free air is one of the criteria for surgery. In a patient with SSc, however, intraabdominal free air is not a criterion for surgery because a patient with SSc usually has pneumatosis cystoides intestinalis (PCI) as the cause of intraabdominal free air without peritoneal irritation. PCI, which results in dissection of luminal gas in the bowel wall, is typically benign, does not require treatment, and only needs bowel rest for about 2 weeks.[10,11] Peritoneal irritation is an important criterion for abdominal operation in patients with SSc.

Acknowledgment We thank Hirokazu Yamamoto (Pathology, Itabashi Medical Laboratory).

References

1. Chin T. Application of a silicone gel sheet for the care of an intestinal fistula. J Jpn Soc Stoma Rehabil 2003;19:85–8 (in Japanese).
2. Sjogren RW. Gastrointestinal motility disorders in scleroderma. Arthritis Rheum 1994;37:1265–82.
3. Horowitz M, Maddern G, Maddox A. Effects of cisapride on gastric and esophageal emptying in progressive sclerosis. Gastroenterology 1987;93:311–5.
4. Ono M, Watanabe K, Aikoh T, Ono M, Omori K, Kohda M, et al. Effects of octreotide acetate treatment for scleroderma bowel. Kawasaki Med J 1996;22:233–7.
5. Dull JS, Rafuman JP, Zakai MD. Successful treatment of gastroparesis with erythromycin in a patient with progressive systemic sclerosis. Am J Med 1990;89:528–30.
6. Rees WDW, Leigh RJ, Christofides ND, Bloom SR, Turnberg LA. Interdigestive motor activity in patients with systemic sclerosis. Gastroenterology 1982;83:575–80.
7. D'Angelo WA, Fries JF, Masai AT, Schulman LE. Pathologic observations in systemic sclerosis. Am J Med 1969;46:428–39.
8. Frances JS, Harriette JK, Theodore MR, Russel ML. Advanced scleroderma bowel: complications and management. J Rheumatol 1988;15:869–74.
9. Grabowski G, Grant JP. Nutritional support in patient with systemic scleroderma. J Parenter Ent Nutr 1989;13:147–51.
10. London NJM, Bailey RG, Hall AW. Spontaneous pneumoperitoneum complicating scleroderma in the absence of pneumatosis cystoides intestinalis. Postgrad Med J 1990;66:61–2.
11. Jamart J. Pneumatosis cytoides intestinalis: a statistical study of 919 cases. Acta Hepatogastroenterol 1971;26:4.