

## Multiple Perforations of the Small Intestine during Systemic Sclerosis

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**Abstract:** Systemic sclerosis (SS) or scleroderma is a very rare systemic connective tissue disease.

Intestinal involvement is reported in 40 to 60% of cases of SS, and is by far dominated by intestinal hypomotility, malabsorption syndrome, and pseudo-obstructions. Intestinal perforation remains exceptional, dominates particularly in the colon, and is associated with a poor prognosis (6-12% of the overall mortality from this connective tissue disease).

We report the original observation of SS complicated by multiple perforations of the small intestine in 28-year-old Tunisian woman.

**Keywords:** perforation, small intestine, systemic sclerosis, scleroderma.

### 1. INTRODUCTION

Systemic sclerosis (SS) or scleroderma is a very rare systemic connective tissue disease [1,2]: prevalence estimated at 30 to 443 per million population [3].

It is characterized by a fibroblastic proliferation with accumulation of dense collagen in the connective tissue, a diffuse microangiopathy, and an immunological disorder [1,2]: presence of anti-nuclear auto-antibodies, of which the most specific of SS are anti-topoisomerase I, anti-centromere, and anti-RNA polymerase III antibodies according to the clinical form of SS[4,5].

Clinical manifestations are dominated by cutaneous, articular, pulmonary, gastro esophageal, cardiovascular, and renal involvement [1-3].

The digestive manifestations of this connective tissue disease can be seen in more than 90% of cases during the course of the disease [3,6]. All the structures of the digestive tract can be reached from the oral cavity to the anus [3,6,7]. Intestinal involvement is reported in 40 to 60% of cases, and is by far dominated by intestinal hypomotility, malabsorption syndrome, and pseudo-obstructions [3, 6]. Intestinal perforation remains exceptional, dominates particularly in

the colon, and is associated with a poor prognosis.

We report the original observation of SS complicated by multiple perforations of the small intestine.

### 2. CASE REPORT

28-year-old Tunisian woman, followed for two years for SS with skin (Figs. 1 and 2), esophageal, osteoarticular (Fig. 3), and cardiovascular involvement was hospitalized from the emergency department for acute abdominal pain worsening rapidly since the day before.



**Fig1:** Sclerodermic face.



**Fig2:** sclerodactyly.



**Fig3:** Subcutaneous calcinosis of the left elbow

The somatic examination noted a feverish patient at 38.7°C with a hardened anterior abdominal wall, not depressible, and painful on palpation. Hemodynamic, respiratory, and neurological status were preserved.

Basic biological tests showed leukocytosis at 12800/mm<sup>3</sup> with 80% of neutrophils, C-reactive protein at 49 mg/l, and erythrocyte sedimentation rate at 52mm/H1. The other tests were within normal limits: hemoglobin, platelets, amylasemia, lipasemia, glycemia, calcemia, ionogram, troponin Ic, transaminases, and creatinine. The ECG and chest X-ray did not reveal any abnormalities.

The abdominal ultrasound showed minimal peritoneal effusion and dilated small bowel loops with thickened walls.

The patient was transferred to the surgical department for urgent surgery with the diagnosis of acute peritonitis.

Intraoperative exploration showed generalized peritonitis with false membranes, multiple punctate ulcers of the small intestine with multiple perforations. The colon was normal. An enlarged resection of the small intestine was performed and broad-spectrum antibiotic therapy was prescribed.

The histopathological examination showed multiple punctate perforations of the terminal ileum and jejunum covered with fibrino-leukocytic material, several superficial ulcers of the small intestinal mucosa with angiodysplasia, microvasculopathy, and diffuse fibrosis. No micro thrombosis, specific vasculitis, or granuloma was noted.

Anti-phospholipid, anti-cardiolipin, anti-β-2-glycoprotein 1 (β2GP1), and anti-neutrophil cytoplasmic (ANCA) antibodies were negative. Likewise, no associated constitutional thrombophilia has been noted (normal levels of protein C, protein S, anti-thrombin III, and noctivated protein C resistance).

The subsequent course was favorable under systemic corticosteroids.

### 3. DISCUSSION

Gastrointestinal involvement is seen in more than 90% of cases of SS [3,6]. It mainly affects the esophagus: 90%, the anus and rectum: 50-70%, the stomach: 40-70%, the colon: 10-50%, and the small intestine: 40% [8].

It is the first sign revealing the disease in 10% of cases [3], and is responsible for 6-12% of the overall mortality from this connective tissue disease [3].

The main presentations of SS-specific intestinal involvement are: hypomotility, small intestinal bacterial overgrowth, malabsorption, chronic pseudo-obstruction syndrome, diarrhea, pneumatosis intestinalis, and bleeding by small intestinal telangiectasia/angiodysplasia [3,9,10].

Severe forms of intestinal involvement, as for our patient, are seen only in 8% of cases [6]. Ulceration and perforation remain exceptional in the small intestine [11, 12]; they are particularly reported in the colon as a consequence of the classic "stercoral ulcers" [13-15].

The causal mechanism of these complications is intestinal neuropathy associated with progressive fibrosis of the intestinal myocytes as well as the micro-vasculopathy that characterizes this disease [12]. However, anecdotal cases of spontaneous intestinal perforation in sclerodermic patients have been reported [11].

### 4. CONCLUSION

The intestinal involvement during SS deserves to be known by health professionals given its important prognostic implications.

Regular clinical and endoscopic screening is very useful for the early diagnosis and adequate management of this disorder, especially since it can remain completely asymptomatic.

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