Colonic volvulus in systemic sclerosis: a rare complication managed with intravenous immunoglobulin

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Gastrointestinal (GIT) involvement is common in systemic sclerosis (SSc), affecting the entire digestive tract. Although dysmotility is the most frequent manifestation, mechanical complications such as colonic volvulus are rare but potentially life-threatening. We report what is, to our knowledge, the first described case of SSc complicated by colonic volvulus successfully managed with intravenous immunoglobulin (IVIG).

A 64-year-old woman presented to our department with a one-year history of severe nausea, recurrent vomiting and diarrhea, with a 30 kg unintentional weight loss. She denied Raynaud's phenomenon.

Physical examination revealed marked cutaneous sclerosis affecting the trunk, abdomen, face and all four limbs, with a modified Rodnan skin score (mRss) of 36. Antinuclear antibodies were positive (titre 1:160) with anti-threonyl-tRNA synthetase positivity. Cardiac, pulmonary and muscle assessments revealed normal. A diagnosis of diffuse cutaneous systemic sclerosis with predominant GIT involvement was established. The patient was started on subcutaneous methotrexate and domperidone for skin and GIT involvement, respectively.

Five months after diagnosis, the patient developed severe worsening of vomiting and constipation. Abdominal imaging revealed intestinal occlusion with dilation of the colon and small intestine, and pneumatosis intestinalis (Figures 1 and 2). Urgent surgical exploration confirmed torsion of the splenic flexure without neoplastic aetiology, and surgical correction was performed.

Symptoms recurred two weeks later, with computed tomography demonstrating similar colonic abnormalities. Colonoscopic decompression was only partially effective.

Given the severity of GI involvement, with recurrent pseudo-obstruction episodes and mechanical volvulus leading to severe malnutrition, off-label IVIG was initiated at a total dose of 2 g/kg administered for five consecutive days. Clinical improvement was reported after the third infusion, with no side effects reported. Post-treatment abdominal radiography revealed regression of pneumatosis intestinalis (Figure 3).

A second IVIG cycle was administered three months later, with further improvement of GI symptoms. Following Gastroenterology follow-up and dietary optimisation, symptoms stabilized, the patient progressively regained weight, and mRSS improved to 7 at the last evaluation, 2 year after diagnosis.

Colonic volvulus in SSc is rare: a systematic review identified only nine published cases, with surgical colonic resection required in the majority[1]. Absence of Raynaud's phenomenon at onset is significant, as data suggests this subgroup exhibits more severe diffuse cutaneous disease and greater GI involvement, consistent with the aggressive phenotype observed here[2,3].

IVIG has an established immunomodulatory role in SSc, with emerging evidence supporting its use in severe GI dysmotility. A multicentre observational study of 78 SSc patients demonstrated significant improvement in GI function as measured by the UCLA GIT 2.0 score, following IVIG at 2 g/kg per cycle[4]. A case series additionally described rapid radiographic resolution of intestinal pseudo-obstruction following IVIG in SSc-myositis overlap patients[5].

This case expands the potential application of IVIG to the most severe SSc-related GI disease, including mechanical complications such as colonic volvulus refractory to surgical and supportive management. Although a definitive causal relationship cannot be established, the clinical improvement, regression of radiological abnormalities, absence of further obstructive episodes, and sustained nutritional recovery support a potential therapeutic effect with a reasonable safety profile.

Tables and Figures

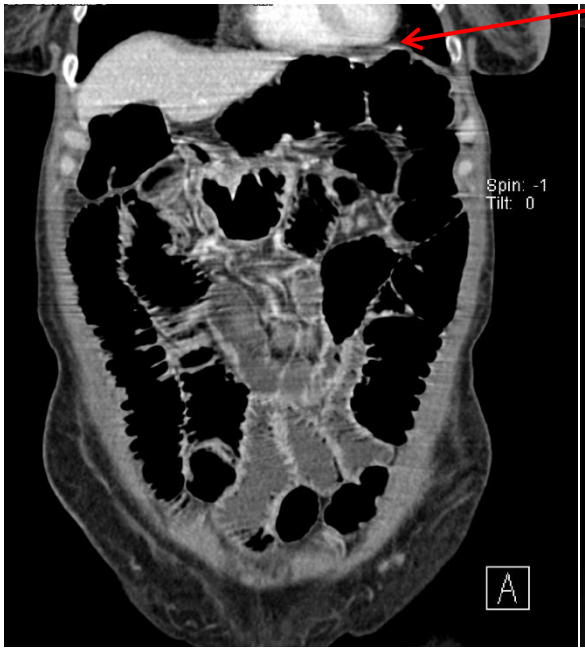

Figure 1 -Abdominal CT before scan IVIG therapy.

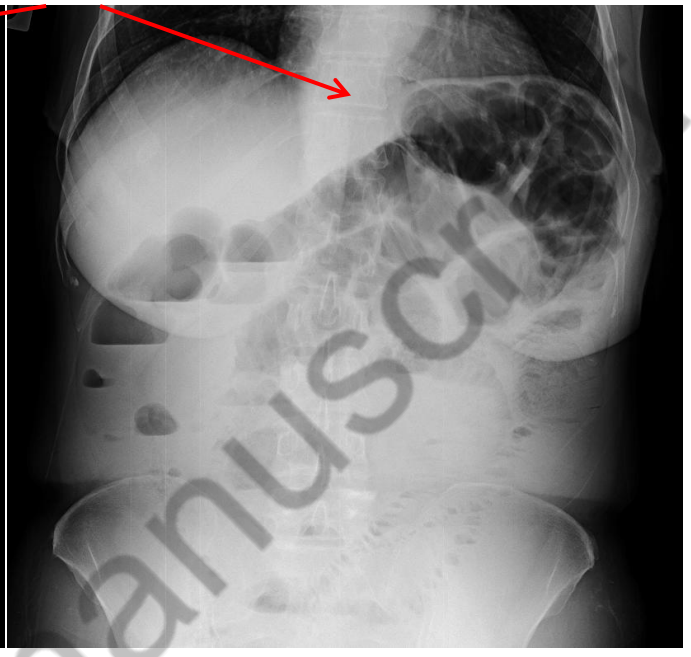

Figure 2 - Abdominal radiograph before IVIG therapy.

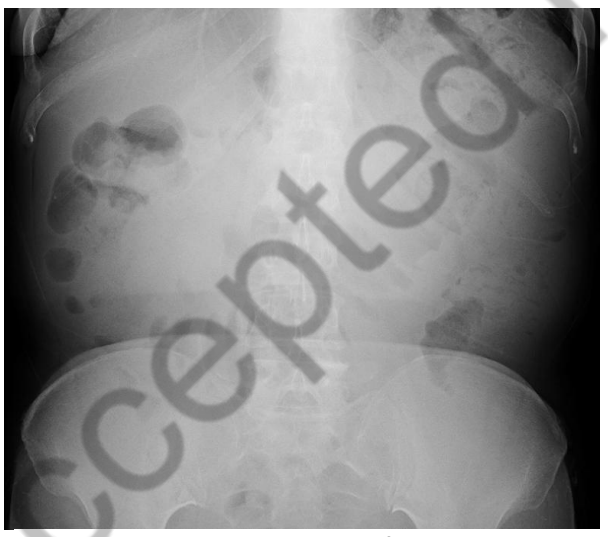

Figure 3 - Abdominal radiograph after IVIG therapy.

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