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Intestinal pneumatosis as a manifestation of systemic sclerosis

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Dear Editor,

A 60-year-old female patient was admitted to the emergency room for a 7-day history of abdominal bloating, nausea, vomiting, constipation, and lack of flatus. She had been diagnosed with systemic sclerosis (SSc) 10 years ago and had been using methotrexate, sildenafil, and prednisone. She did not present any signs of instability, but physical examination showed malnourishment status and abdominal tenderness and distention. Plain abdominal radiography was suggestive of sigmoid volvulus, confirmed and successfully resolved after endoscopic decompression therapy. Eight months later, the patient developed a new episode of abdominal obstruction. Computed Tomography (CT) scan identified a distended sigmoid colon due to its torsion with gas areas within the bowel wall (Figure 1). This time, endoscopic decompression had failed to treat, so exploratory laparotomy was performed. Colonic distention and sigmoid volvulus were identified during the procedure, after which sigmoidectomy followed by primary anastomosis was performed. Neither perforation nor masses were found. Furthermore, the anatomopathological study was inconsistent with vascular, inflammatory, or neoplastic diseases.

DISCUSSION

Systemic sclerosis (SSc) is an autoimmune disease characterized by progressive fibrosis of the skin and internal organs and small vessel vasculopathy (1). Skin thickening pattern classifies the subset of the disease into two significant types: diffuse cutaneous SSc and localized cutaneous SSc. However, some patients lack skin involvement despite autoantibodies, microvascular disease, and internal organ fibrosis. Even though gastrointestinal tract involvement (GIT) is a common feature in patients with SSc, the 2013 American College of Rheumatology/European League Against Rheumatism (ACR/EULAR) classification criteria for SSc does not incorporate GIT manifestations in this scoring system (2).

GIT involvement occurs in up to 98,9% of SSc patients, and its severity varies from mild to severe GIT disease according to intensity and location of involvement. Severe GIT manifestations happen in 8% of patients and carry high morbimortality rates (3).

Intestinal Pneumatosis (IP) is a rare condition and represents a radiologic sign due to gas trapping into the bowel wall (4). In SSc, IP is very infrequent, and although there are mechanisms that try to explain its development, the pathophysiology still needs to be completely elucidated. A potential pathogenic role of intestinal ischemia in our patient can be present (5), a condition often associated with SSc. Imaging modalities such as radiography, CT scan, and CT angiography may help identify this complication (4); ultrasound or magnetic resonance imaging has little indication. Finally, the treatment for IP is based on supportive measures, and surgery might be required in some cases.

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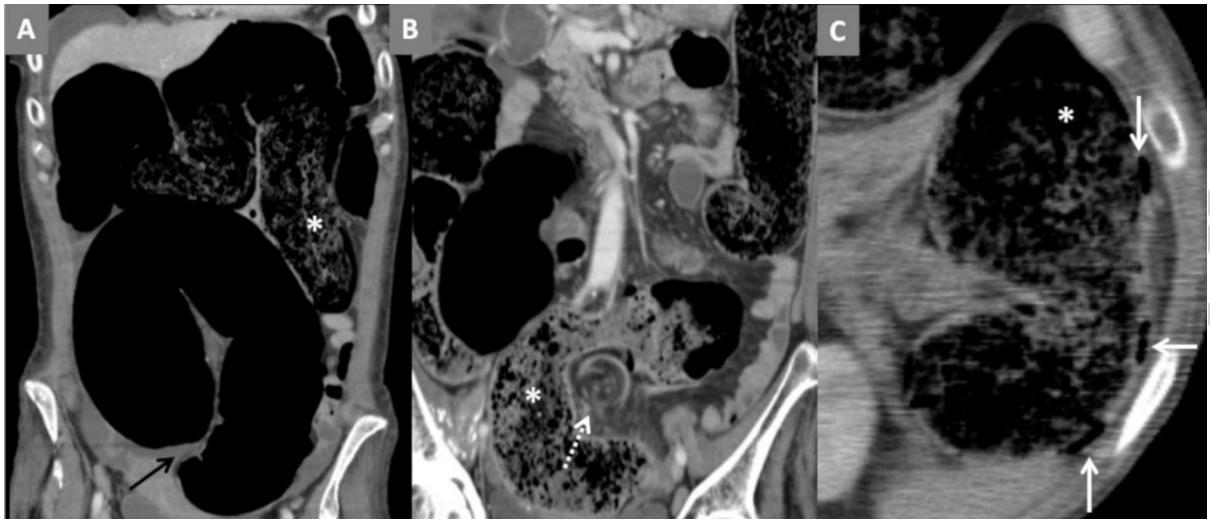

Figure 1- Computed Tomography: The distended sigmoid colon with the classic "coffee bean sign" configuration of sigmoid volvulus due to its juxtaposed walls (black arrow). There are two points of abrupt reduction with the torsion of the intestinal loop and the adjacent mesentery (“whirl sign”- dashed white arrows), determining marked distension of the upstream colic segments, with gaseous and fecal content (asterisks). Multiple gas-filled cysts in the submucosa of the bowel walls (foci of pneumatosis) in the descending colon are identified (white arrows).