

Systemic sclerosis and sarcoidosis: a rare case of chronic intestinal pseudo-obstruction

Key words: Systemic sclerosis. Sarcoidosis and chronic intestinal pseudo-obstruction.

Dear Editor,

The coexistence of systemic sclerosis (SSc) and sarcoidosis is an extremely rare phenomenon; some studies question its existence. We report the case of a male with a diagnosis of sarcoidosis that was admitted due to abdominal distension and pain. After a thorough investigation, he was diagnosed with severe chronic intestinal pseudo-obstruction as a manifestation of SSc.

Case report

We report the case of a 62-year-old male admitted with abdominal distension, pain and no bowel movements. He had been diagnosed with pulmonary sarcoidosis three years previously. The physical examination identified

telangiectasia on the face, sclerodactilia, bilateral Raynaud's phenomenon (Fig. 1A), arrhythmic heart sounds and a distended abdomen with a decreased peristalsis. The blood tests were normal and an abdominal X-ray showed a marked distension of the large and small bowel. Colonoscopy and magnetic resonance (MR) enterography confirmed these findings in the absence of an organic obstruction (Fig. 1B). A diagnosis of chronic intestinal pseudo-obstruction (CIPO) was suspected. An extensive workup was performed and auto-antibody screening was negative. A capillaroscopy of the nail bed according to the suspicion of SSc was positive for advanced SSc and an esophageal manometry showed > 30% of non-transmitted waves. An atrial flutter and a restrictive cardiomyopathy were evident on the electrocardiogram (EKG) and echocardiogram. A diagnosis of SSc with severe gastrointestinal and cardiac involvement was suspected. Clinical resolution was observed with conservative treatment; there were no flare-ups during follow-up.

Discussion

SSc and sarcoidosis are rare diseases with an incidence between 50-400 million cases/year, 20 million cases/year, and the coexistence of both conditions is extremely rare (1). Only 30 cases have been reported in the literature (2) and some authors question the association of both con-

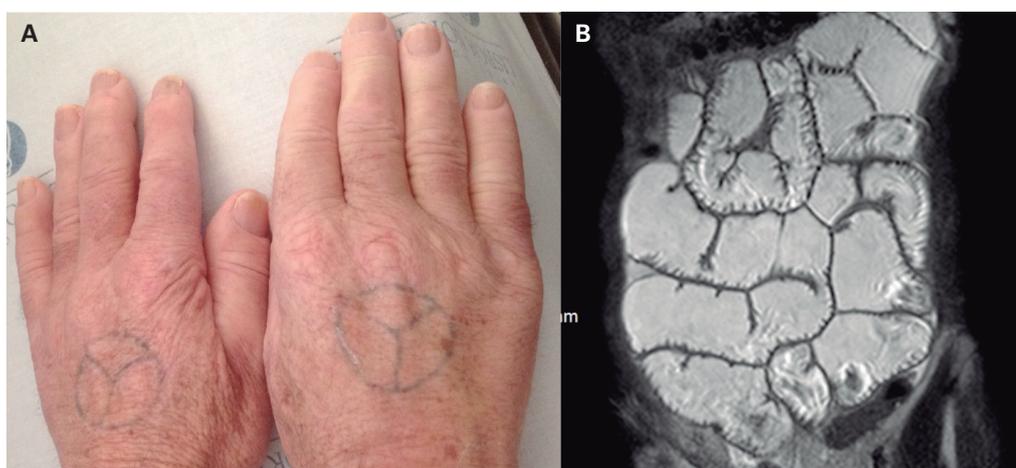


Fig. 1. A. Sclerodactylia and bilateral Raynaud's phenomenon. B. MR-enterography showing a marked distension of the large and small bowel without organic obstruction.

ditions (1,3). In our patient, a diagnosis of SSc was made based on the presence of the typical cutaneous findings. The gastrointestinal tract (GIT) is the most affected area (90%) and 75-90% of cases have esophageal involvement; dysmotility is the most common abnormality (1,4). Usually, there is a decreased or absent primary peristalsis, although 30% of cases may be asymptomatic (4). The small bowel is the second most common organ involved (20-60%) and CIPO rarely occurs (10% of cases) (4). Another unusual feature of this case is the fact that SSc usually affects females (70% of cases); SSc was diagnosed after sarcoidosis in only 20% of cases (2). The pathogenesis of GIT involvement is poorly understood. However, the most credible theory involves functional autoantibodies against the muscarinic-3receptor (M3R) that induces cholinergic dysfunction (4). SSc treatment is symptomatic. However, recent studies suggest the potential benefit of intravenous immunoglobulin (IVIG) infusions as it neutralizes autoantibodies to M3R (2,5).

In conclusion, we report a case of a rare cause of CIPO as a manifestation of SSc in a patient with a previous diagnosis of sarcoidosis. Gastroenterologists should consider SSc as a cause of intestinal pseudo-obstruction.

*Sónia Bernardo, Ana Rita Gonçalves and Luís Araújo Correia
Gastroenterology and Hepatology Service. Santa Maria Hospital. Centro
Hospitalar Lisboa Norte. Lisboa, Portugal*

DOI: 10.17235/reed.2018.5495/2018

References

1. Kobak S, Sever F, Sivrikoz O, et al. Coexistence of sarcoidosis and systemic sclerosis. *Case Reports Rheumatol* 2013;1-3. DOI: 10.1155/2013/684216
2. Senda S, Igawa K, Nishioka M, et al. Systemic sclerosis with sarcoidosis: case report and review of the published work. *J Dermatol* 2014;41:421-3. DOI: 10.1111/1346-8138.12438
3. Koguchi N, Okada A, Choh S, et al. Concomitant systemic sclerosis and sarcoidosis with combined pulmonary fibrosis and emphysema. *Intern Med* 2016;55:1331-5. DOI: 10.2169/internalmedicine.55.5903
4. Kumar S, Singh J, Rattan S, et al. Review article: pathogenesis and clinical manifestations of gastrointestinal involvement in systemic sclerosis. *Aliment Pharmacol Ther* 2017;1-16. DOI: 10.1111/apt.13963
5. Kumar S, Singh J, Kedika R, et al. Role of muscarinic 3-receptor antibody in systemic sclerosis: correlation with disease duration and effects of IVIG. *Am J Physiol Gastrointest Liver Physiol* 2016;310:G1052-60. DOI: 10.1152/ajpgi.00034.2016