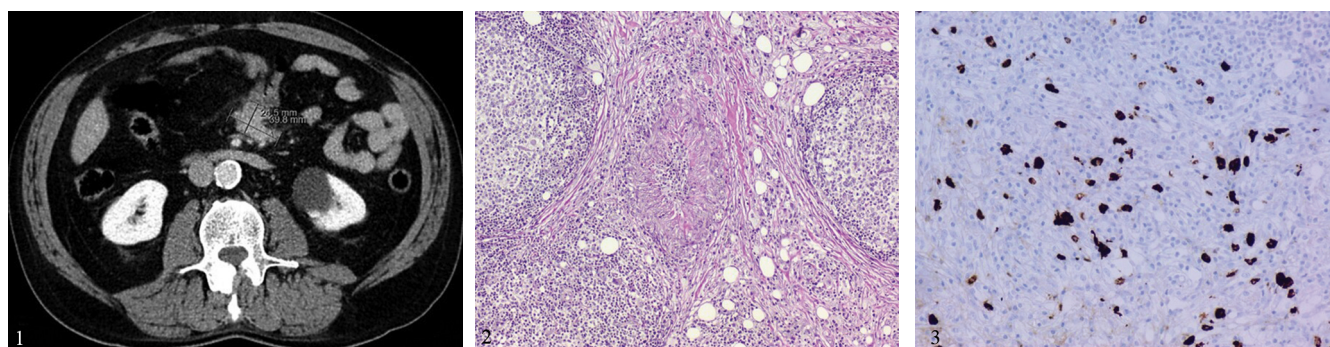


A Rare Cause of Abdominal Pain: IgG4-Related Sclerosing Mesenteritis

Sara Archer¹, Tiago Pereira Guedes¹, Tomás Fonseca², Diogo Carvalho Sá³, Isabel Pedroto¹

1) Department of Gastroenterology, Centro Hospitalar Universitário do Porto, Porto; 2) Unit of Clinical Immunology, Centro Hospitalar Universitário do Porto, Porto; 3) Department of Pathology, Centro Hospitalar Universitário do Porto, Porto, Portugal



A 67-year-old man with previous cardiovascular diseases was referred due to a 5-month history of recurrent epigastric pain. Esophagogastroduodenoscopy and full blood workup were unremarkable. Computed tomography scan showed an irregularly shaped mass at the root of the mesentery, measuring 40x25x47mm, with spiculated contours and retractile behavior (Fig. 1). Simultaneous densification of the adjacent fat and infracentimetric ganglionic formations scattered throughout the mesentery were shown. Surgical biopsy revealed extensive storiform fibrosclerosis, with the presence of interstitial lymphoplasmocytic infiltrate and obliterative phlebitis (Fig. 2); the plasma cells had mostly IgG expression, with IgG4:IgG ratio >40% (Fig. 3), accounting for more than 30-40 IgG4 plasma cells per field. The serum IgG4 level was 137 mg/dL. A diagnosis of IgG4-related sclerosing mesenteritis (IgG4-RSM) was made, without other organ involvement. Prednisolone (0.6mg/kg/day) improved partially the abdominal pain, so steroid sparing strategy with off-label rituximab was associated. Positron emission tomography scan after 1 year of rituximab treatment showed a reduction of the lesion, with no significant metabolic activity. The patient is asymptomatic to date.

IgG4-RSM is an uncommon manifestation of IgG4-related disease (IgG4-RD). Due to its low prevalence, the understanding of this entity is scarce, and its diagnosis is challenging. Unlike other manifestations of IgG4-RD, IgG4-RSM is identified in later stages and has unspecific symptoms [1]. It should be kept in mind in cases of chronic abdominal

pain and as a differential diagnosis of other mesenteric diseases like panniculitis due to chronic infections or lymphoma [2]. Serum biomarkers that can help a precocious diagnosis are also scarce and in IgG4-RSM serum IgG4 can be normal, making the diagnosis even more challenging. Although steroids are the standard of care in IgG4-RD, some disease phenotypes can be refractory, probably to its fibrotic nature. Rituximab showed some benefits in this kind of presentation, but randomized clinical trials are still lacking [3].

Corresponding author: Sara Archer, u13418@chporto.min-saude.pt

Conflicts of interest: None to declare.

REFERENCES

1. Liu Z, Jiao Y, He L, Wang H, Wang D. A rare case report of immunoglobulin G4-related sclerosing mesenteritis and review of the literature. *Medicine (Baltimore)* 2020;99:e22579. doi:[10.1097/MD.00000000000022579](https://doi.org/10.1097/MD.00000000000022579)
2. Bertoni M, Giani A, Tozzini S, Di Natale ME. Sclerosing Mesenteritis as an Uncommon Site of Involvement of IgG4-Related Disease: A Case Report With an Updated Review of the Literature. *Cureus* 2022;14:e25041. doi:[10.7759/cureus.25041](https://doi.org/10.7759/cureus.25041)
3. Tome J, Kamboj AK, Pardi DS. S2168 The Use of Rituximab for IgG4-Related Sclerosing Mesenteritis. *Am J Gastroenterol* 2022;117(10S):e1470. doi:[10.14309/01.ajg.0000865312.23249.61](https://doi.org/10.14309/01.ajg.0000865312.23249.61)