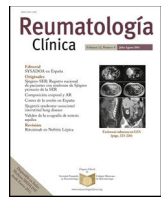




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Images in Clinical Rheumatology

Unusual presentation of IgG4-related disease

Presentación inusual de la enfermedad relacionada con el IgG4

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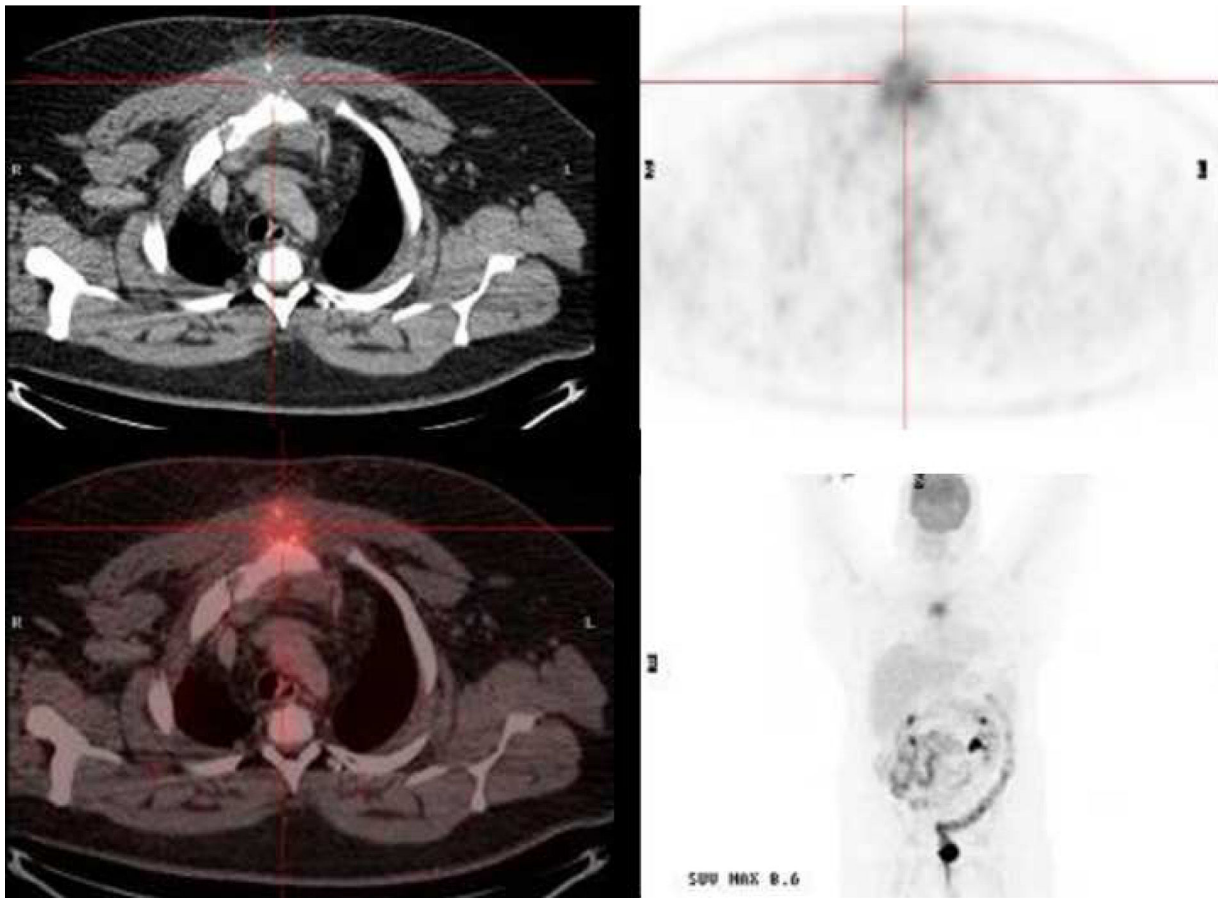


Fig. 1. Thoracic computed tomography scan shows destructive sternal bone lesion associated with a soft tissue mass (approximate size 96 mm × 71 mm × 53 mm) with a hypermetabolic lytic lesion at the manubrium-sternal junction, associated to a mass of perilesional soft tissues, and infiltration of the upper mediastinal fat with a SUV max value of 8.6.

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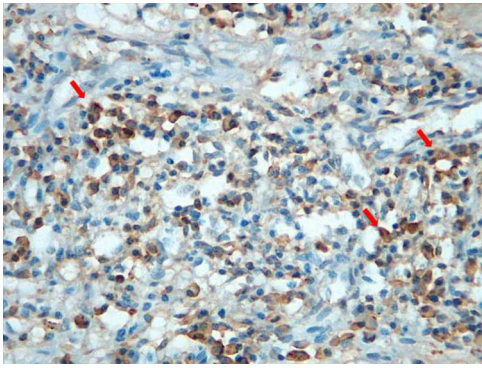


Fig. 2. Biopsy of the sternal lesion with immunohistochemical staining for IgG4 shows IgG4 positive plasma cells (arrows).

A 45-years-old male patient came to the Emergency Room due to a chest pain of a month of evolution, the physical examination was normal. The patient has a history of metabolic syndrome. Cardiac pathology was ruled out. A thoracic computed tomography (CT) scan was performed, where an destructive sternal bone lesion associated with a soft tissue mass located in the upper third of the breastbone was observed (approximate size 96 mm × 71 mm × 53 mm) associated to a soft tissue mass located at the upper third of the breastbone were found, in addition to sug-

gestive of malignancy bilateral axillary mediastinal adenopathy's (Fig. 1).

A PET-CT was performed, where a hypermetabolic lytic lesion at the manubrium-sternal junction, associated to a mass of soft tissue, with infiltration of the upper mediastinal fat with a SUVmax value of 8.6 were found (Fig. 1)

A biopsy of the sternal lesion and the soft tissue mass were performed, whose appearance was highly suggestive of IgG4-related disease, because a positive immunohistochemical study for IgG4 plasma cells was clearly present (Fig. 2). Elevated IgG4 levels in peripheral blood were also found. Treatment with dexamethasone was initiated, and a complete clinical response was seen.

IgG4-related disease is an autoimmune systemic disorder that can affect virtually any organ, most often affecting the pancreas, lymph nodes, and salivary glands.¹ The lesions have their own histopathological characteristics, mainly infiltration of IgG4 positive plasma cells in the affected tissue; usually an increase of IgG4 serum levels of is also present.^{2,3}

References

1. Stone JH, Zen Y, Deshpande V. IgG4-related disease. *N Engl J Med.* 2012;366:539–51.
2. Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. *Lancet.* 2014;6736:1–12.
3. Khosroshahi A, Stone JH. A clinical overview of IgG4-related systemic disease. *Curr Opin Rheumatol.* 2011;23:57–66.