IgG4-related Hashimoto's thyroiditis

An emerging variant of a well known disease

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INTRODUCTION

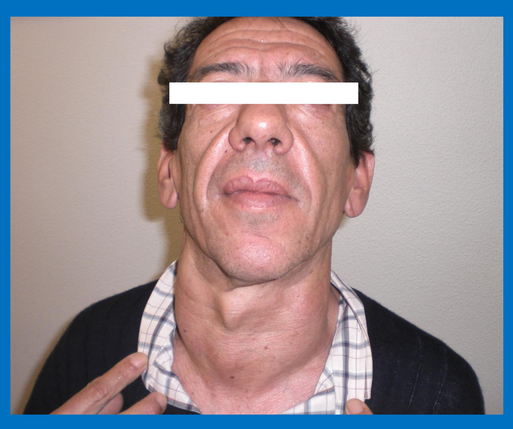
IgG4-related Hashimoto's thyroiditis (HT) is a very recently reported entity, characterized by thyroid inflammation rich in IgG4-positive plasma cells associated with marked fibrosis. It may be part of the systemic IgG4-related sclerosing disease and is associated with younger age, lower female-male ratio, higher levels of thyroid autoantibodies, low echogenicity on ultrasound (US), rapid progress requiring surgery and more subclinical hypothyroidism, when compared with non-IgG4 HT.

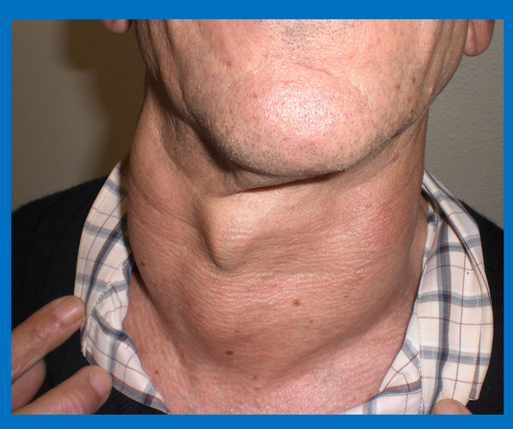
CASE REPORT

Identification: 56-year-old Portuguese man

Present illness: Four-month history of progressive neck swelling, dysphagia, and weight loss of 7 kg

Past medical history and family diseases: unremarkable

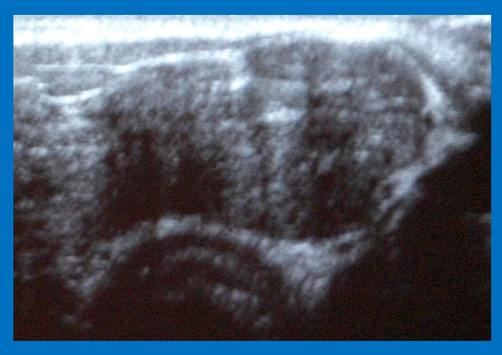




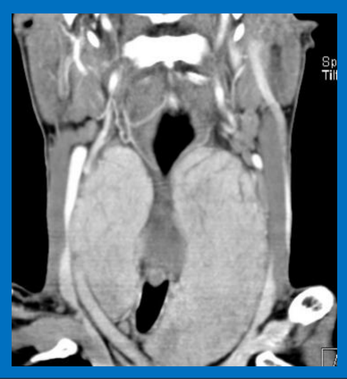
Cervical palpation: enlarged, hard and painless thyroid gland

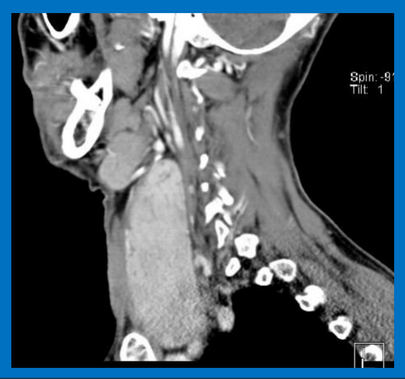
Laboratory testing: ESR 81 mm/h (<31), CRP 8.2 mg/dL (<1), TSH 19 mIU/L (0.1-4), free T_4 0.99 ng/dL (0.93-1.7), anti-Tg >4000 IU/mL (<40), anti-TPO >600 IU/mL (<35)

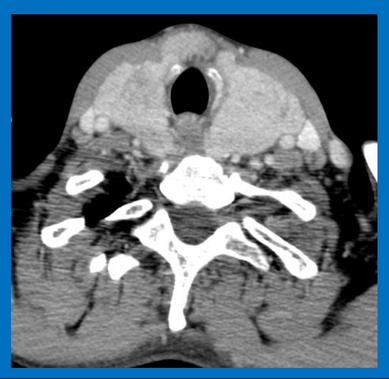
Cervical US: enlarged and heterogeneous thyroid gland, with a substernal component, both lobes with maximal dimension >10 cm and two hypoechoic nodules with 3.2 and 2.2 cm located on the isthmus and right lobe, respectively



Cervical CT: thyroid gland with increased dimensions and a substernal component



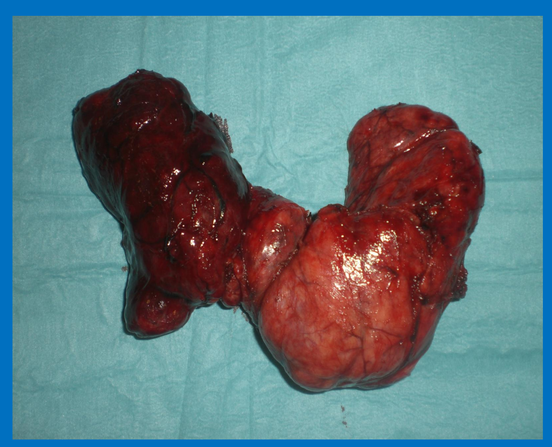




US-guided fine needle aspiration cytology of both nodules: lymphocytic thyroiditis

Treatment: total thyroidectomy

No surgical complications



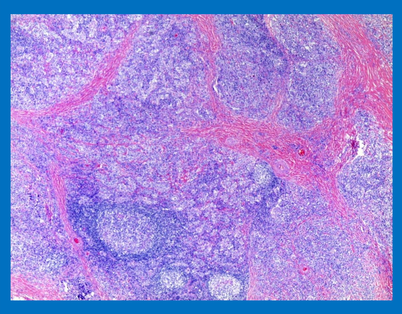
Weight: 284 g

Right lobe: 10 x 6 x 4.5 cm

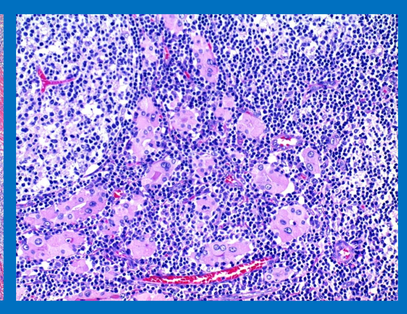
Left lobe: 12 x 6 x 6 cm

Hard consistency

Histological findings: lymphoplasmacytic infiltration, lymphoid follicles with germinal centers and marked fibrosis limited within the thyroid capsule. Increased number of IgG4-positive plasma cells (> 50 cells/high-power field)



hematoxylin/eosin



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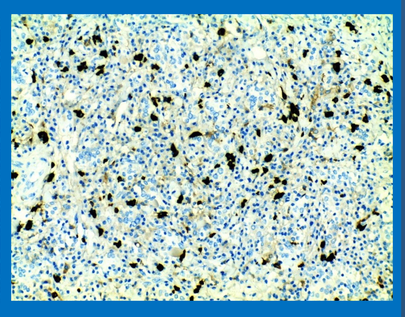

IgG4 immunostaining

Follow-up: one month after surgery, serum IgG4 concentration high-normal, 165 mg/dL (3-201)

Symptoms relief and reduction in laboratory inflammatory parameters

Thyroid function controlled with Levothyroxine 137 µg/day

ESR 20 mm/h, TSH 2.9 mIU/L, free T_4 1.2 ng/dL, anti-TPO >600 IU/mL

CONCLUSIONS

We report a typical case of IgG4-related HT. The presentation form suggested the diagnosis, which was confirmed based on histological data.

Our case highlights this new variant of the well known HT, and helps physicians in recognizing its main clinical features.

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