

SCIENTIFIC LETTER

Invasive fibrous thyroiditis with severe acute compressing presentation



Tiroiditis fibrosa invasiva con presentación compresiva aguda severa

A 49-year-old woman was being followed up at our clinic for a euthyroid multinodular goiter. In 2009, an ultrasound (US) scan showed two well-defined hypoechoic nodules in the left lobe: the largest one was 33 mm in size and contained gross calcification; the other one was 15 mm in size. Fine needle aspiration of these lesions revealed benign histopathology, with a diagnosis of benign follicular hyperplasia. Anti-thyroid antibody levels were within the normal range. In 2015, a routine US scan showed significant enlargement of the goiter with intrathoracic extension and growth of the above nodules to 42 mm and 18 mm, respectively. A computed tomography (CT) scan showed predominant left lobe enlargement with tracheal displacement but no compression. Surgery was proposed but declined by the patient and she was followed up. In 2018, she presented urgently to our clinic complaining of progressive breathlessness and dysphagia over the last two weeks. The presence of dysphonia and severe dyspnea with stridor was evidenced. Physical examination revealed a large bilateral mass of woody consistency, which did not move on swallowing. There was also paralysis of the left vocal cord. A CT scan showed a diffuse infiltrative process that affected mainly the left thyroid lobe and ipsilateral surrounding tissues, with complete carotid involvement, tracheal compression and muscle damage (Fig. 1). Clinical suspicion of an aggressive type of thyroid carcinoma was established. Empirical treatment with intravenous dexamethasone (6 mg per day) was started to improve the compressive symptoms. Fine needle aspiration showed only inflammatory cells and she underwent surgery. Thyroidectomy was not possible due to hard fibrous adhesions. A biopsy specimen suggested invasive fibrous thyroiditis (IFT) as a first possibility. Glucocorticoid administration (dexamethasone 6 mg per day for 10 days and then prednisone 30 mg per day) led to a rapid and marked clinical improvement, with complete resolution of the compressive symptoms after a few days. A drastic reduction in the volume of the mass was also observed. A CT scan performed after six months showed an almost complete regression of the infiltrative process with recovery of the tracheal lumen (Fig. 1). Currently, the patient is still receiving low-dose oral prednisone (5 mg per day) and she remains asymptomatic. On physical examination, the neck appears similar to before the onset of IFT.

IFT was first described by Bernhard Riedel in 1894. Unlike other invasive thyroid diseases, IFT often involves adjacent structures and may cause hypoparathyroidism, laryngeal paralysis, esophageal and/or tracheal stenosis, and mediastinal injury. The estimated incidence is 1/100,000 patient-years, with a female to male ratio of 4:1 and a prevalence in the 30–50-year age group. It has been proposed that IFT may be a manifestation of various systemic fibrosing and autoimmune disorders, and it could be the specific thyroid lesion of the IgG4-related disease (IgG4-RD), although the etiopathogenesis remains incompletely understood.^{1,2} The typical clinical presentation is a slow-growing goiter of woody consistency that is immobile on swallowing. The enlargement may be asymmetric and occasionally only one lobe is involved. In advanced stages, when the infiltrative process is widespread, there is a hard diffuse thickening of the neck, sometimes associated with dysphagia, hoarseness, dyspnea, symptoms of hypocalcemia and Horner's syndrome. Approximately one-third of patients develop hypothyroidism and up to two-thirds have thyroid autoantibodies. There is often a clinical suspicion of aggressive thyroid cancer.³ A thyroid US scan shows hypoechoic areas without vascular flow, often infiltrating surrounding tissue. CT and magnetic resonance imaging show the lesions with greater accuracy. Complete involvement of the carotid artery has been reported in two cases. Fluorodeoxyglucose positron emission tomography shows IFT as a hypermetabolic process.⁴ Fine-needle aspiration samples are generally unsatisfactory. Open biopsies provide specimens with hyalinized avascular tissue with inflammatory cells, the number of which decreases in advanced stages. Paucicellular anaplastic thyroid cancer can mimic IFT.^{4,5} Without treatment, IFT is usually slowly progressive and potentially life-threatening, mainly due to tracheal damage. In a review of 212 cases, 190 had improvement or resolution, while 22 did not improve or worsened, of whom five died from IFT.⁶ As IFT is a rare condition, there are no clinical trials of treatments. The most common therapeutic approach is the administration of high doses of glucocorticoids, with doses up to 100 mg prednisone per day being described, the effect of which is variable. Some patients have experienced softening and reduction of the mass, especially in the early stages when inflammatory changes predominate over established fibrosis. Three cases of complete remission in response to steroids have been reported. There is a risk of recurrence when steroids are reduced or discontinued.^{7,8} Tamoxifen has also been used in IFT with variable results. One case of improvement with rituximab after no response to glucocorticoids and tamoxifen has been reported.⁹ The role of surgery is often limited to taking biopsies and relieving airway obstruction by isthmectomy, as the obliteration

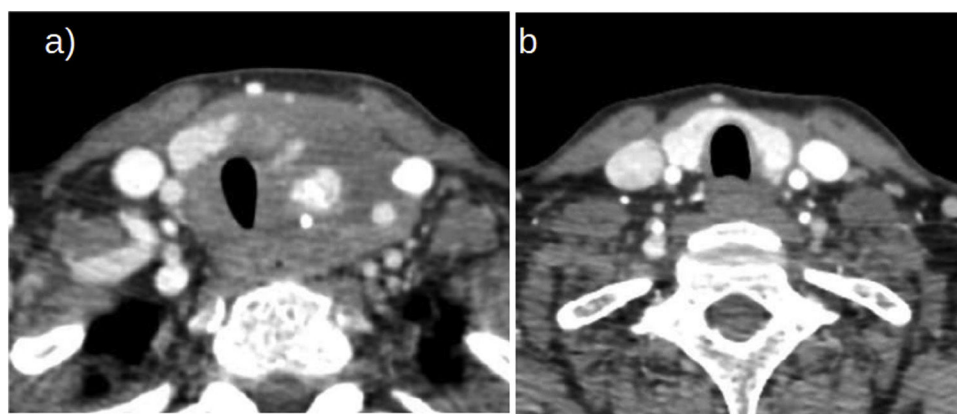


Figure 1 CT scan images at diagnosis of Riedel's thyroiditis (a) and at six months of corticosteroid therapy (b).

of tissue planes often precludes other procedures.³ Radiotherapy also has a role in cases of IFT refractory to steroids and tamoxifen.

We think that this case is of particular interest because it shows an extremely rare presentation of IFT, with a life-threatening rapidly progressive acute course with extensive involvement of cervical structures (even complete carotid involvement, which has been reported in only two cases besides this one). Probably due to it having this type of disease progression, in which the inflammatory component predominates over the fibrotic component, early treatment with corticosteroids was very effective and led to complete remission. Although discontinuation of treatment has been suggested by some authors, we prefer to maintain it, as we think that it may favour the permanence of the remission without exposing the patient to the typical adverse effects of corticosteroid therapy at higher doses. Finally, we would like to point out that although the area covered by our hospital includes 300,000 people, to our knowledge only this case of IFT has been treated during a period of 30 years, an incidence 90 times lower than that traditionally described in the literature. This would be consistent with a reported dramatic reduction in the incidence of this disease in recent decades.¹⁰

Conflict of interest

None of the authors have a conflict of interest regarding the content of this publication.

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