CASE REPORT

Extensive sterile abscess in an invasive fibrous thyroiditis (Riedel's thyroiditis) caused by an occlusive vasculitis

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ABSTRACT. Riedel's thyroiditis is a rare disease determined by an invasive fibrosclerotic transformation of the thyroid gland. It may be one manifestation of multifocal fibrosis with still unknown etiology. Because it mimics carcinoma, a biopsy must be performed to get the correct diagnosis. The condition is self-limiting when confined to the neck. Prognosis depends on the extent of extracervical fibrosclerosis. We present a patient with a huge cervical and mediastinal, unilateral thyroid mass expanding to the aortic curve, which led to tracheal deviation and compression with symptoms of stridor and dyspnea. These symptoms continued under a course of high-dose steroids; thus an operation was necessary to relieve the airway obstruction and limit inflammation. Intraoperative and pathological findings showed an inflammatory infiltration of the adjacent neck muscles and a sterile abscess caused by an occlusive vasculitis. Therefore, hemithyroidectomy had to be performed instead of a local limited resection.
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INTRODUCTION

Riedel's thyroiditis is a chronic inflammatory disease of unknown etiology of the thyroid gland. The first two cases published were in 1896 by Professor Bernhard Riedel, the former chairman of the Department of Surgery in Jena, Germany (1). Both patients had tracheal obstruction caused by an extremely hard thyroid tumor without malignancy. Treatment was limited to simple wedge resection of thyroid isthmus to relieve tracheal obstruction.

Woolner defined Riedel's thyroiditis to be an entity characterized by the following histologic criteria: 1) fibrotic process of the thyroid gland, 2) extension of the fibrotic inflammation beyond the capsule with invasion and replacement of adjacent neck structures, and 3) complete destruction of the involved parts of the thyroid gland, without the giant cell reaction seen in granulomatous thyroiditis (2, 3). Therefore, the disease was also named "invasive fibrous thyroiditis" (2).

In a review of the literature up to 1986, 178 cases met the above mentioned criteria of Riedel's thyroiditis (4). In the last 13 years, another 54 patients were described (Medline®), making a total of only 232 published cases.

We report here a further case of Riedel's thyroiditis accompanied by a sterile abscess.

CASE REPORT

A 66-year-old man, with a history of a pre-existing goiter, presented to the emergency department in July 1999, with a sudden painless enlargement of his thyroid. His symptoms included progressive dyspnea and stridor over a one-week period.

On examination, there was an impressive enlargement in circumference of the neck with an extended, hard and fixed right thyroid lobe. The patient was afebrile, but had an elevated leukocyte count of 15.4/nl (normal range: 3 to 8/nl), as well as C-reactive protein of 25.2 mg/dl (normal range: 0 to 0.5 mg/dl). Antibiotic therapy with ciprofloxacin produced a slight improvement of laboratory parameters but did not significantly ameliorate clinical symptoms.

Key-words: Riedel's thyroiditis, multifocal fibrosclerosis, sterile abscess, vasculitis, surgery, hemithyroidectomy.

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Chest radiograph demonstrated a huge cervical and mediastinal tumor, leading to a deviation of the trachea to the left. In thyroid ultrasonography the right thyroid gland had an estimated volume of 180 milliliters partly localized in the upper mediastinum. A huge hypoechoic and heterogeneous nodule infiltrated the adjacent muscles. The left thyroid was normal in structure and size. A magnetic resonance scan revealed an enlarged thyroid mass of 8 x 8 x 12 cm expanding to the aortic curve and deviating the pharynx, esophagus, vessels, and trachea to the left. Also seen, was a tracheal-stenosis and thrombosis of the vena jugularis interna with inflow congestion. After application of contrast medium the tumor itself did not show an uptake while there was a slight enhancement of the surrounding thyroid. Altogether the scan missed the typical radiological findings of an abscess or malignancy (Fig. 1).
Thyroid laboratory examinations demonstrated normal hormone levels and the absence of antithyroid antibodies. The technetium uptake over the right gland at scintigraphy was low, with a normal left side.

With the tentative diagnosis of "carcinoma", a punch biopsy was performed. The histology hinted at the diagnosis of invasive fibrous thyroiditis without malignancy.
Multifocal fibrosclerosis with extracervical manifestations of the disease could be excluded by chest X-ray and abdominal ultrasonography.
A high-dose steroid therapy (100 mg prednisolone daily for one week) was initiated. Only a transient relief in breathing was noted with therapy. However, neck-swelling and inflammatory parameters increased dramatically. For this reason, the patient was taken to surgery. Neck exploration was carried out and a large, indurated grey mass was found that replaced most of the right thyroid lobe and invaded the adjacent muscles (Fig. 2). Upon removal of the mass, about 300 milliliter of unsuspected purulent fluid was noted to be leaking out of it. Subsequent microbiologic examination showed the fluid to be sterile. After abscess-evacuation the resection of the right thyroid lobe could be performed in the usual way without sternotomy. The

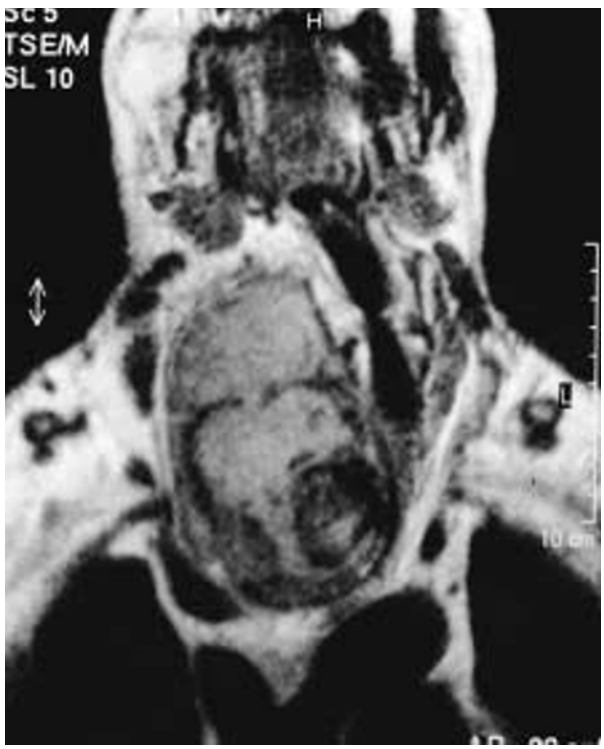

Fig. 1 - *Coronal magnetic resonance scan of the neck in a patient with Riedel's thyroiditis. Enlargement (8 x 8 x 12 cm) of the right thyroid lobe expanding to the aortic curve. Hereby dislocation of pharynx, esophagus, vessels, and trachea to the left with extensive tracheal-stenosis.*

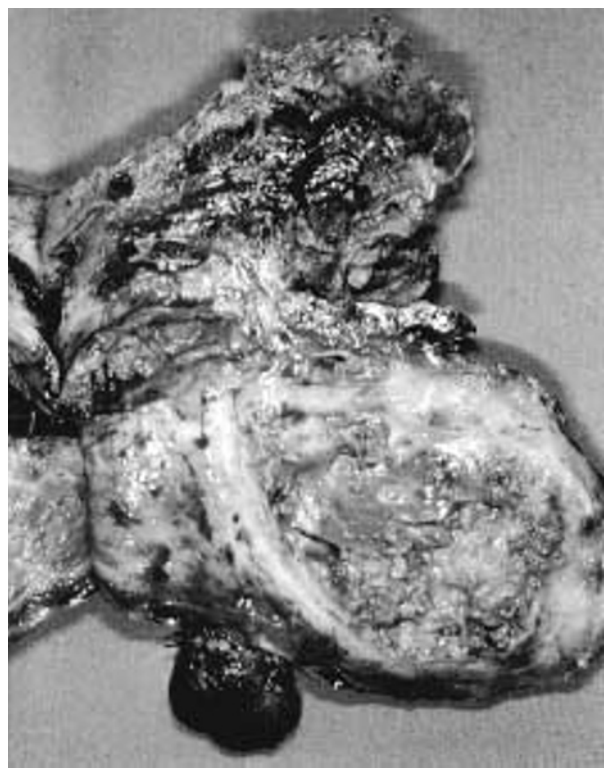

Fig. 2 - *Gross pathologic findings of surgical hemithyroidectomy specimen in Riedel's thyroiditis. The tissue is dense, white and avascular and shows an infarcted area with an abscess of a maximal diameter of 6 cm.*

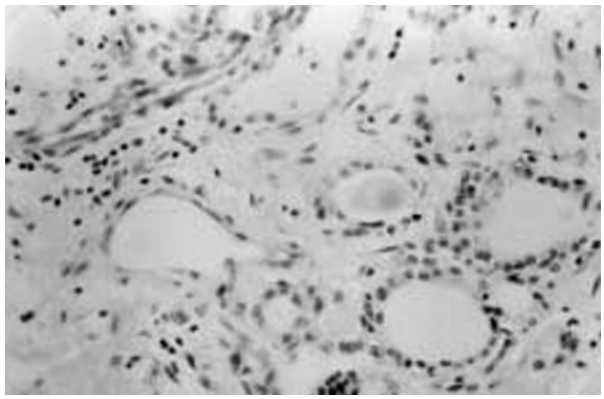

Fig. 3 - *Typical microscopic feature of Riedel's thyroiditis as manifested in our patient. The normal thyroid architecture is completely destroyed by hyalinized fibrous tissue with inflammatory cells (hematoxylin-eosin, x 40).*

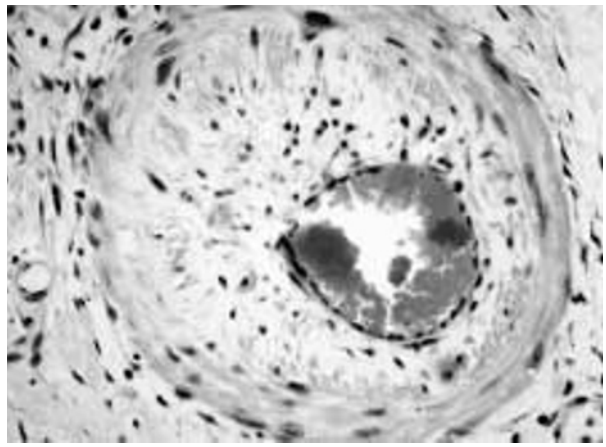

Fig. 5 - *Inflammatory vascular infiltration with resultant occlusive vasculitis (hematoxylin-eosin, x 40).*

parathyroids and the laryngeal nerve were identified.

Histologic examinations excluded malignancy and showed a fibroinflammatory process of thyroidal and perithyroidal tissue with infiltration of lymphocytes, plasma cells, and neutrophils (Fig. 3). Infiltration of adjacent muscles histologically imitated myositis, but without giant cells (Fig. 4). Histopathological findings detected an occlusive vasculitis causing infarction of the thyroid and resultant formation of the abscess (Fig. 5). Altogether, the diagnosis of Riedel's thyroiditis was confirmed.

In the postoperative period, the patient was supported by the respirator for two days, afterwards he was breathing very well on his own. A laryngeal examination revealed no paralysis of the vocal cords and the inflammatory parameters normalized rapidly. The thrombosis of the right subclavian vein was recanalized with anticoagulative therapy. A subcutaneous seroma resolved after repeated punctures. In a clinical follow-up visit four weeks later, the wound was completely healed, the patient could breath normally without stridor, and felt well. One year later, he was doing well without any symptoms of recurrence.

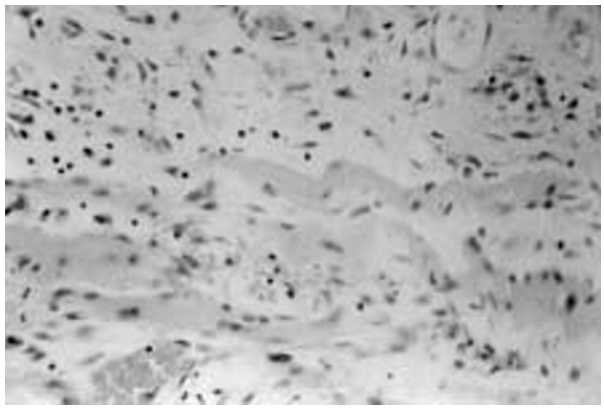

Fig. 4 - *Fibrosis and inflammation infiltrate adjacent sternohyoid muscles histologically imitating myositis (hematoxylin-eosin, x 40).*

DISCUSSION

Invasive fibrous thyroiditis is a very rare condition. The Mayo Clinic reported about 37 cases within 64 years and 56,700 thyroidectomies (5). This operative incidence of 0.06% corresponds with other investigations (2, 6, 7). We present the second case of Riedel's thyroiditis ever observed at our surgical department in over 15 years. During that time, approximately 4500 thyroidal resections had been performed so that the incidence of Riedel's thyroiditis in our patient-population is 0.04%.

As typical for Riedel's thyroiditis in our patient, the clinical appearance was characterized by a non-painful and hard enlargement of a pre-existing goiter (3-6, 8). Initially, these alterations are mostly localized unilaterally, but involvement of the contralateral gland is possible (4, 6, 8). The time span from the beginning of the first symptoms to diagnosis is about 4 to 10 months (2, 7, 9). Retrospectively, our patient remembers having increasing stridor and dyspnea for six months. Our observed symptoms of local pressure like dysphagia, dysphonia, and especially dyspnea and stridor were caused by a tracheal obstruction due to an increasing thyroid mass (3, 4, 6-9). A palsy of the la-

ryngeal nerve (6, 7), or a hypocalcaemia due to pressure on the parathyroid glands were lacking in our patient. These symptoms are usually reversible after decompression (3, 10, 11).

Clinical diagnosis of Riedel's thyroiditis is difficult. Besides physical examination findings of sonography, scintigraphy and even magnetic-resonance scan may be normal or mimic carcinoma (5, 9). The disease is characterized by a chronic inflammation but laboratory findings are non-specific (3, 8). However, as in our patient, increased inflammatory parameters are possible (3, 8). These were due to the systemic reaction to infarcted and necrotic thyroid tissue. Antithyroid antibodies were absent. In literature antibodies are found in 45 to 67% (4, 5), but it is not clear whether these are pathogenic or a phenomenon as a result of exposure of sequestered antigens to the immune system through tissue damage (4, 9). The diagnosis usually requires histologic verification (7). Samples received by needle biopsy are not diagnostic because the fibrotic reaction surrounding an undifferentiated carcinoma may be confused with the fibrotic reaction seen in Riedel's thyroiditis (3, 5, 12, 13). Though in our case a punch biopsy already hinted at the correct diagnosis, an open biopsy is always essential.

The main differential diagnosis of Riedel's thyroiditis is carcinoma, with same clinical features like dysphonia, stridor, and unilateral hard swelling of the thyroid gland (14). Besides carcinoma other entities of thyroiditis must be included in the differential considerations like chronic lymphocytic thyroiditis (Hashimoto's disease) or subacute thyroiditis de Quervain (5, 8). The acute suppurative thyroiditis is a very rare form of inflammation of the thyroid gland caused most often by hematogenic spread of bacteria, fungi or parasites (5). Besides the local inflammatory signs, the diagnosis would be based on fever, increased inflammatory parameters and the bacteriologic proof. In our case, the microbiologic examinations confirmed that the evacuated fluid was sterile. The abscess was most likely caused by vessel occlusion but not infection. Riedel's thyroiditis is self-limiting with a favorable prognosis (15). If the disorder is confined to the neck, complications are rare and include airway compression, hypothyroidism, and hypoparathyroidism (3, 5, 16).

A dramatic improvement has been demonstrated by systemic use of high-dose glucocorticoid even with recovery of parathyroid and vocal cord function (3, 4, 8, 9, 17-19). This and the finding of antithyroid antibodies in up to 67% of 178 reviewed patients support the hypothesis of an autoimmune reaction (3, 4). Other reports of no response to steroids confirm our experience. It is speculated that steroids may be helpful in the initial phase of the disease, but are without benefit in the end-stage where there is completely transformed fibrotic tissue (9). Nevertheless, a time-limited treatment with steroids should be performed in any patient (8, 9, 17, 18).

Because of the self-limiting character, only moderate surgical management should be chosen to relieve tracheal obstruction. A more extensive dissection increases the risk of damage to vital neck structures because of the altered anatomy caused by the fibrosclerosis (3, 5). Riedel's recommendation of a wedge-resection of the thyroidal isthmus is still preferred (3, 5, 15). This results in a release of tracheal compression. Simultaneously, an obligatory thyroid biopsy is obtained to confirm the diagnosis and to exclude carcinoma (15, 20). The presented patient showed an extensive abscess extending to the upper mediastinum. Therefore, a single wedge excision would not be sufficient. That was the reason to change the strategy from a minimal-invasive surgery normally performed in this disease to a hemithyroidectomy.

Riedel already theorized the accompaniment of endarteritis in his thyroiditis. Soon after, a detailed description of the vascular changes showed a diffuse infiltration of lymphocytes and plasma cells in the walls of small and medium-sized vessels (4, 21, 22). Meijer *et al*. reported five cases with inflammatory changes of small and large veins in the infiltrative, occlusive, and sclerosing stages. They considered this to represent an occlusive phlebitis, which could not be found in other thyroid diseases like Hashimoto or granulomatous thyroiditis "de Quervain". Therefore, an occlusive phlebitis was postulated as a diagnostic feature of Riedel's thyroiditis (22).

We postulate, that in our case inflammatory infiltration of the arteries leads to vessel occlusion with ischemia, infarction and subsequent formation of a sterile abscess. This supports Riedel's theory of an endarteriitis and favours the pathogenetical role of occlusive vasculitis.

CONCLUSIONS

The pathophysiology of Riedel's thyroiditis is most likely caused by an occlusive vasculitis. Surgical procedures depend on the extension of symptoms and inflammation. While simple wedge resection can relieve tracheal obstruction in chronic disease without risk to the parathyroids and the laryngeal nerve, extended inflammation and necrosis requires advanced surgical interventions, like hemithyroidectomy.

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