Riedel's Thyroiditis: A Diagnosis to Remember
Taleuan A*, Bamine H, Elatiq H, Oudidi A, Elalami MN

Department of ENT, UHC Hassan II, FES, Morocco

DOI: 10.36347/sasjs.2020.v06i04.005 | Received: 13.03.2020 | Accepted: 20.03.2020 | Published: 08.04.2020

*Corresponding author: Taleuan A

Abstract

Riedel's thyroiditis is a rare benign pathology of unclear etiology, characterized by a densely fibrotic inflammatory process involving the thyroid gland and the adjacent neck tissues. This rare form of chronic thyroiditis often leads to clinical evidence of anaplastic thyroid carcinoma or lymphoma. We discuss, based on an observation of an unusual case of Riedel's thyroiditis, the clinical characteristics and management modalities of this rare entity.

Keywords: Riedel's Thyroiditis, Diagnosis, Histology.

Copyright © 2020: This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

INTRODUCTION

Riedel's thyroiditis is a rare benign condition of unknown etiology characterized by extensive fibrosis of the thyroid and adjacent tissues [1]. This rare form of chronic thyroiditis was first described in Germany in 1896 by Bernhard Riedel [2]. The great characteristic of Riedel's thyroiditis is that it can mimic an anaplastic carcinoma or lymphoma [1, 3], leading to mutilating interventions with the morbi-mortality they can generate. Hence the importance of educating practitioners about clinical presentation, which would enhance their ability to make this diagnosis quickly.

We report an observation of a rare case of Riedel's thyroiditis, in order to clarify the clinical features and management modalities of this rare entity.

OBSERVATION

This is a 38-year-old patient with no significant pathological history of basal cervical mass, which has increased rapidly over the past 2 months, associated with dysphonia and exertional dyspnea, with no signs of dysphagia or dysthrodism, all evolving in a context of apyrexia and preservation of general condition. Clinical examination noted a basal cervical mass, hard, mobile, and painful, with normal looking skin. Ganglia areas were clear. Nasofibroscopy revealed a paralysis of the right vocal cord. The thyroid hormones were correct, and the cervical ultrasound showed a multinodular goiter (the largest nodule measuring 49 mm), heterogeneous, and poorly limited. Cervical CT scan showed a multinodular goitre (GMHN), predominantly right-sided (measuring 68×52×50 mm overall) with bumpy contours and heterogeneous density. The GMHN exceeds the cervico-thoracic orifice by 43 mm and exerts a mass effect on the trachea, which is compressed and displaced to the left, and on the esophagus and jugulocarotid axis, which are also compressed and displaced (Fig. 1).

Based on these clinical and paraclinical characteristic, the malignant nature of goiter was strongly suspected, and the diagnosis of invasive thyroid cancer (anaplastic carcinoma) was suggested. On surgical exploration, the thyroid gland was hard, fibrotic, fixed, and adherent to the trachea and the right vagus nerve with invasion of the subhyoid muscles (Fig. 2). Thyroid extirpation was difficult, due to its multiple adhesions, which led us to perform a subtotal thyroidectomy extended to the subhyoid muscles (Fig. 3). Extemporaneous examination of the surgical specimen (Fig. 4) revealed fibrous, inflammatory and congestive reorganisation. The definitive histological analysis showed Riedel's thyroiditis which was confirmed by the immuno-histochemical study. The patient was put on hormone replacement therapy and then referred to the endocrinology department where low dose corticosteroid therapy (20mg prednisone/d) was instituted, with a good clinical evolution with a 16-month follow-up.
Riedel’s Thyroiditis is extremely rare (1.06/100,000) [1, 3]. The operative incidence among goitres operated on at the Mayo Clinic between 1920 and 1984 was 0.06%: 37 cases out of 56700 thyroidectomies [4]. Women are 4 to 5 times more affected than men [3, 4]. The average age at diagnosis is 30-50 years, with an average diagnostic delay of 4-10 months [3, 4].

This rare form of chronic thyroiditis involves a possibly autoimmune inflammation and fibrosis of the thyroid gland and its surrounding tissues, with possible involvement of distant organs [4, 5]. The presence of IgG4+ plasma cells and thyroid autoantibodies in some patients with Riedel’s thyroiditis has led some authors to consider it as part of multifocal fibrosclerosis, linking it to the group of IgG4 sclerosing diseases [3, 5].

Riedel’s Thyroiditis has no pathognomonic clinical signs. Usually the goiter has a remarkably hard consistency, often associated with compressive signs, such as a gene in the anterior part of the neck, dysphagia, a feeling of suffocation and dyspnea, especially in the supine position [1, 3-6]. This clinical picture is caused by tracheal and/or esophageal compression by fibrosis and can sometimes lead the practitioner to confuse this form of thyroiditis with a malignant tumour. Other clinical features of Riedel’s thyroiditis resembling a malignant tumour can be found, such as lesions of the inferior laryngeal nerve [6, 7] causing vocal cord paralysis and consequently dysphonia, or a lesion of the cervical sympathetic nerve responsible for Claude Bernard Horner syndrome [7]. Progressively Riedel’s chronic thyroiditis will destroy the thyroid gland itself. This is why thyroid function is
most often normal at first, and then a phase of hypothyroidism occurs. In the series of Schwaegerle et al. 64% of subjects had euthyroidism, 32% had hypothyroidism and 4% had hyperthyroidism [8].

Scintigraphic, ultrasound and cytological data are rarely conclusive [1,7-9]. Computed tomography is the imaging of choice, and is recommended firstly for diagnosis by showing a hypodense appearance without enhancement when administering a contrast agent, except sometimes at the level of adjacent tissues with regard to fibrosis; and secondly as part of monitoring for extension assessment, which must be systematic in the search for retroperitoneal fibrosis [3, 8, 10].

The only way to arrive at a conclusive diagnosis is by pathological examination from a biopsy, or after decompressive surgery [1, 9-11]. To avoid aggressive surgery that will be potentially morbid, Riedel's thyroiditis must be differentiated histologically from malignancies, particularly anaplastic thyroid cancer, thyroid lymphoma, sarcoma and fibrotic variant of Hashimoto's thyroiditis [3, 5-7]. In anatomopathology; the cell population is composed of polymorphic inflammatory elements, dense collagen fibres separate the thyroid into small lobules [1, 6]. Fibrosis is often hyaline in appearance and replaces the thyroid parenchyma. This fibrosis extends to fatty tissue, muscles and may sheathe vessels [1, 3-5]. In the absence of an understanding of the pathogenesis of the disease, the therapeutic approach is empirical and imperfect. Corticosteroid therapy is effective in the constitutive phase of the disease, which is of more controversial interest beyond this [9, 12, 13]. Small doses are recommended (10 to 20 mg per day) for 4 to 6 months, possibly maintained over the long term if they prove to be effective. Tamoxifen is a good alternative, starting at 20 mgx2 per day and decreasing according to adverse events to 10 mgx2 per day [12, 13]. The place of anti-TNFα therapeutics, immunosuppressive in localized or more general forms of fibrosis disease, is still imperfectly codified [12, 13].

Surgery can be performed to lift tracheal or esophageal compression, although extensive surgery is not recommended and complete resection of the thyroid gland is almost always impossible. Surgery has no curative indication, except exceptionally at a very early non-extensive stage of the disease [3, 13-14]. Riedel's thyroiditis does not seem to affect life expectancy. So far, only one fatal case of Riedel's thyroiditis has been described [6]. However, in approximately one-third of patients with Riedel's thyroiditis, one or more localizations of fibrosis develop over a 10-year period, and complications due to retroperitoneal disorders and/or pulmonary and mediastinal fibrosis may occur and cause death [15, 16].

CONCLUSION
Our case highlights the difficulty of diagnosing Riedel's thyroiditis, which should be suspected in all patients with a hard thyroid mass and compression symptoms. In any case, Riedel's thyroiditis remains a diagnosis of elimination; the removal of a malignant tumor must be systematic. Future efforts should focus on improving preoperative diagnosis, either by fine needle aspiration or imaging.

Declaration of links of interest
The authors declare that they have no links of interest.

Authors' contributions
All authors contributed to the conduct of this work. All authors also declare that they have read and approved the final version of the manuscript.

REFERENCES