CASE REPORT

RIEDEL'S THYROIDITIS IN AN ETHIOPIAN PATIENT

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ABSTRACT

Riedel's thyroiditis is a very rare fibroinflammatory disease of the thyroid gland of an unknown etiology. We describe this rare disease in a 40 year old Ethiopian woman who presented with a painful anterior neck swelling. With a suspicion of thyroid gland malignancy, the patient underwent a subtotal thyroidectomy. The histopathologic examination showed dense fibrous replacement of the thyroid gland and a diagnosis of Riedel's thyroiditis was confirmed. This is the first documented case of Riedel's thyroiditis in Ethiopia and we recommend the disease should be in the list of differential diagnosis of a thyroid mass which is hard with fixation to the surrounding structures. Additionally surgery should be considered to make a histologic diagnosis and relieve compressive symptoms.

Key Words: Riedel's thyroiditis, fibro-inflammatory diseases

INTRODUCTION

Riedel's thyroiditis is a very rare chronic inflammatory disease of the thyroid gland. It was first recognized in 1896 by Bernhard Riedel who described two patients with a hard goiter and tracheal compressive symptoms (1, 2). The etiology is not clear, but inflammatory, autoimmune and infectious factors are incriminated. It is usually difficult to diagnose Riedel's thyroiditis clinically, and definitive diagnosis is made after histopathologic evaluation.

The disease has some relation with retroperitoneal, mediastinal and retro-orbital fibrosis, and it is considered by some as a systemic fibrosclerosis process that involves the thyroid. Women are 4 times more likely to be affected than men, and it most commonly occurs between 30 and 50 years of age (3). Modes of treatment include surgery to relieve pressure, and although no extensive trials exist because of its rarity, good results have been reported with steroids and tamoxifen.

The purpose of this case report is to describe this rare disease in Ethiopia, and discuss the diagnostic difficulties and clinical presentation so that similar cases could be picked early and managed appropriately.

CASE REPORT

A 40 year old female patient presented to the Gondar University Hospital with a complaint of anterior neck swelling of 9 months duration .The swelling was initially small but progressively increased. She had progressive dysphagia and pain but had no change in her voice. She had no symptoms of hypothyroidism or hyperthyroidism. Her physical examination showed a stable patient with a 6 x 6cm thyroid swelling .The swelling was stony hard in consistency and had mild tenderness. The overlying skin was not fixed to it, but the mass hardly moved from side to side and longitudinally. There were no associated enlarged lymph nodes and no evidence of thyrotoxicosis was noted during her examination.

All her laboratory investigations were within the acceptable range, and her thyroid function tests were normal. Fine needle aspiration biopsy from the thyroid gland was done and showed scanty aspirate consisting of few follicular cells with no atypical features, but conclusive diagnosis could not be rendered. With all the above findings, a clinical diagnosis of thyroid carcinoma was suspected and the patient was subjected to surgery. Intra operatively, there was dense adhesion between the thyroid gland and strap muscles which were dissected with difficulty.

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After releasing the adhesions, the carotid arteries and internal jugular veins were found bilaterally attached to the gland and they were carefully released from the gland. The parathyroid glands were visualized with a lot of difficulty, and the thyroid was adherent to the trachea which was difficult to dissect. Near total thyroidectomy was done with a significant difficulty and a small normal looking part of the gland was left untouched. Post operatively the patient showed uneventful recovery and was discharged improved on the 7th post operative day. The patient was appointed for follow up in two weeks with thyroxine supplementation but didn't show up on the appointment date.

Pathologic findings

Macroscopic

The gross specimen was a grey white solid unencapsulated mass measuring 6x6x4cm, and weighing 56gm .The cut surface revealed a greyish-white solid mass which was firm to hard. There was no normal looking thyroid tissue visualized. (Figure1, 2)

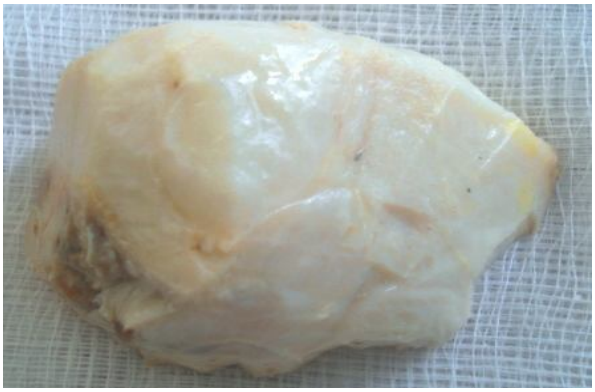

Fig.1 Gross picture of the thyroid showing a grey white solid unencapsulated mass

Fig.2 Cut surface of the gross specimen showing a grey white solid lesion

Microscopic

There was dense collagenous background with fibroblastic spindle cell proliferation and scattered lymphocytic and plasmacytic infiltrates. There were few scattered vessels with an inflammatory infiltrate in the wall without giant cells (Figure 3,4). The fibrous tissue involved almost all parts of the specimen except at the upper pole where few scattered solitary follicular cells and few thyroid follicles with a small amount of colloid were seen embedded in the fibrous tissue. No malignant cells were observed.

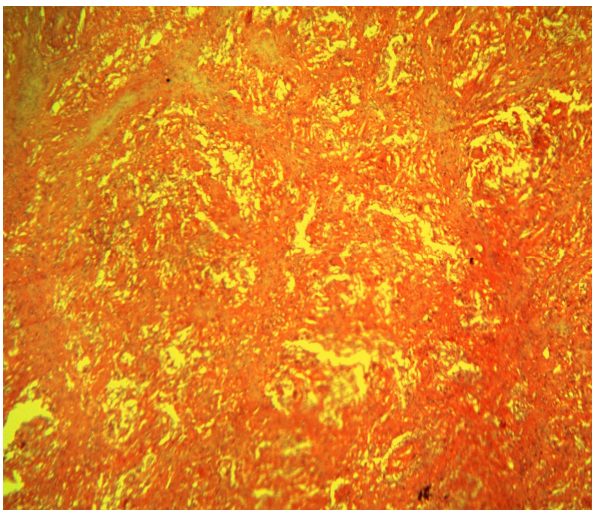

Fig 3. Low power view showing fibrotic lesion which replaced the thyroid (Hematoxylin & Eosin 200X)

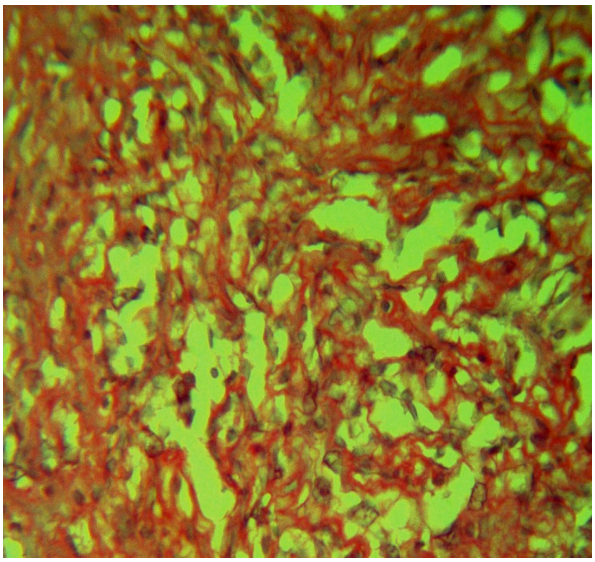

Fig. 4. High view of a densely fibrotic tissue (Hematoxylin & Eosin 400x)

DISCUSSIONS

Riedels thyroiditis is an extremely rare disorder characterized by invasive fibrous tissue proliferation (1, 2). It is considered as a local manifestation of a systemic fibrotic process, and is a progressive fibrosis of the thyroid gland that may extend to surrounding tissues. The prevalence of this disease is only 0.05 percent among patients with a thyroid disease requiring surgery, and its cause is unknown (2, 3). High serum thyroid antibody concentrations are present in up to 67 percent of the patients, but it is unclear whether the antibodies are the cause or an effect of the fibrotic thyroid destruction (2, 4).

Patients with Riedel's thyroiditis present with a rock-hard, fixed, painless goiter. They may have symptoms due to tracheal or esophageal compression or hypoparathyroidism due to extension of the fibrosis into adjacent parathyroid tissue. Riedel's thyroiditis itself may be confused on clinical grounds alone with malignant neoplasms because of its invasive features. Most patients are euthyroid at presentation but later become hypothyroid once replacement of the normal thyroid tissue is nearly complete (5, 6 and 7).

Riedel's thyroiditis is often suspected on clinical grounds of being an undifferentiated carcinoma. However, when surgical exploration is performed, the finding of a hard fibrous mass replacing the thyroid, firmly adherent to the surrounding planes, the diagnosis can be suspected. A definitive diagnosis is made by open biopsy, and a microscopic pattern of interstitial fibrosis with a lymphoplasmacytic infiltrate and atrophic thyroid follicles establishes a definite diagnosis (8).

The criteria used to make the histologic diagnosis of Riedel's thyroiditis were defined first by Woolner et al and subsequently improved by Meijer and Hausman (8) and Schwaegerle et al(1). These are: 1.fibroinflammatory process involving all or a portion of the thyroid gland; 2.evidence of extension into surrounding tissues, including strap muscles; 3.

Infiltrates of inflammatory cells without giant cells, lymphoid follicles, oncocytes or granulomas; 4.evidence of occlusive phlebitis; 5. absence of neoplasm.

Our patient presented with the classical clinical features of hard and fixed thyroid enlargement of a relatively short duration .The intraoperative finding of dense adhesion which was present between the thyroid and the strap muscles was typical of the disease .The histologic appearance was also classical with dense fibrosis which replaced a large part of the thyroid .All these findings are consistent with the established features of Riedel's thyroiditis which have been reported in different reports(1-9).

Diagnosis of Riedel's thyroiditis is usually difficult by cytologic examination. Fine-needle aspiration experience is limited in Riedel's thyroiditis and only a few reports are available in the literature. In most cases, acellular or paucicellular, often nondiagnostic, smears are obtained (2). This is the probable reason for the misdiagnosis in our case as the diagnostic element was scanty.

The treatment of Riedel's thyroiditis is surgical although therapy with glucocorticoids, methotrexate, and tamoxifen has been reported to be successful in the early stages of the disease. The main purpose of surgery is to confirm the diagnosis and relieve compressive symptoms (9). Our patient disappeared before the histopathologic diagnosis was ready and didn't receive any medical therapy except the thyroxine supplementation.

In conclusion, this case report calls attention to the existence of Riedel's thyroiditis in Ethiopia, and it should be in the list of differential diagnosis of a thyroid mass which is hard and fixed to the surrounding structures. Moreover, surgery should be considered for therapeutic reasons and making tissue diagnosis as cytologic examination might not be revealing.

ACKNOWLEDGEMENTS

We would like to acknowledge our patient and all the physicians and nurses involved in the care of the patient.

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