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Case Report

Reidel's Thyroiditis: A Case Report and Review of Literature

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A B S T R A C T

Reidel's thyroiditis (RT) is a very rare form of chronic thyroiditis associated with fibrosis of the thyroid gland, surrounding cervical tissue and also with other forms of systemic fibrosis. It has a very complex presentation including both local and various systemic and biochemical alterations, but because of its rare occurrence, it is very difficult to suspect and diagnose it. It should be evaluated in every patient with hard thyroid mass having compressive symptoms. High dose steroids followed by tamoxifen should be started early to prevent the ongoing fibrotic process. Here there was a case of Reidel's thyroiditis in a 45 years old male patient who presented with a left-sided cervical mass with features of dysphagia and difficulty in breathing. This case highlights the difficulty in histological diagnosis and subsequent avoidance of aggressive surgery in this rare type of disease.

Keywords: Reidel's thyroiditis, Fibroinflammatory Process, Immunotherapy

Introduction

Reidel's thyroiditis (RT) is an extremely rare condition with an unknown aetiology. It was first described by Bernhard Reidel in 1883.[1] The prevalence of RT is 1 per 100000 inhabitants. Females of 30 to 50 years of age are the most commonly affected.[2] There is fibrosis of the thyroid gland along with the surrounding structures like trachea and oesophagus. The fibrotic process also involves the adjacent nerves, vessels, and parathyroid glands that may lead to compressive symptoms along with endocrine abnormalities. RT is a part of the spectrum of IgG 4 related systemic disorder (IgG4-RSD).[3] It has also been associated with a generalised fibro inflammatory process, commonly called "multifocal fibrosclerosis". There are no clear cut guidelines for the treatment of this disease, but it has been described that surgery, systemic steroids along with immunosuppressants like tamoxifen, rituximab, and mycophenolate mofetil may help to cure the disease.

Case History

A 45 years old male presented with a left side neck mass, dysphagia, and breathing difficulty for the last six months. He was having good general physical condition with normal vital signs. On examination, there was a 3*3 cm swelling present in the left cervical region which was hard in consistency, without any pulsation or murmur. He was having pulmonary tuberculosis 13 years back for which he took the full course of anti-tubercular therapy. Thyroid function tests were within normal range and anti-TPO antibodies were absent. Computed tomographic scan (Figure 1) of the neck was suggestive of non-specific enlargement of the left side thyroid gland along with widespread involvement of the surrounding structure. There was a postero-lateral displacement of the internal jugular vein and carotid artery. Fine needle cytology was inconclusive. An incisional biopsy was performed at the swelling. On histological examination, there were sections of

thyroid follicle of variable size containing colloid. The stroma was fibrous and showed lymphoid aggregates with germinal centre formation and dense plasma cell infiltration. There was no evidence of malignancy. Immunohistochemistry was positive for IgG4. After due consultation, he was started on systemic steroids and tamoxifen.

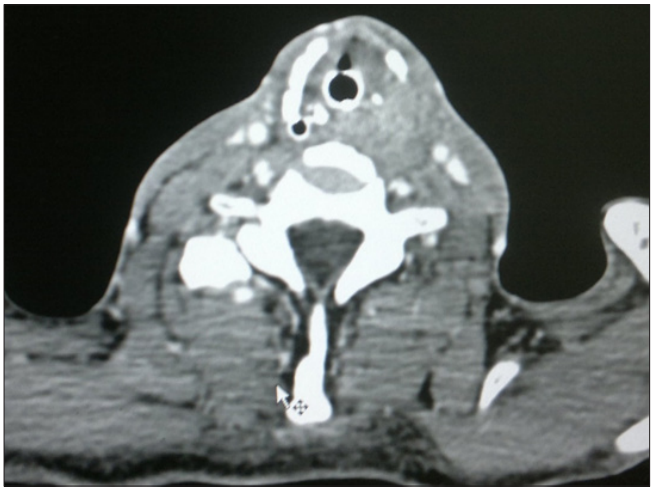

Figure 1.Computed Tomography Scan of Neck suggestive of Heterogeneously Enhancing Mass in the Left Lobe of Thyroid with Postero-lateral Displacement of Internal Jugular Vein and Carotid Artery with Compression of Oesophagus

Discussion

Reidel's thyroiditis is a very rare disease to encounter in clinical practice. The peak age for RT is the fifth decade with a male to female ratio of 1:4.[4] In contrast to it, our patient was a male who presented to us in his fourth decade. RT has varied clinical pictures. Most of the time it is associated with other disorders like concomitant hypothyroidism and Hashimoto's thyroiditis.[5] Local inflammatory condition and autoimmune immune process are the likely aetiology of this rare disease. The condition is characterised by the overgrowth of fibrosing connective tissue involving the surrounding tissue. The inflammatory process is so severe that it is very difficult to find a dissecting tissue plane during the surgery. Some evidence also suggests that RT may be due to organ-specific manifestation of multiple idiopathic fibrosclerosis because of its association with other similar conditions like pancreatitis, sclerosing cholangitis, and mediastinitis, but most recently it was linked to IgG-4 related systemic disorder[4,6] (IgG4-RSD).

Fine needle aspiration is the first diagnostic tool for any thyroid swelling, but its role in RT is controversial. We did FNA twice but with inconclusive results. The final diagnosis was established only after incisional biopsy and immunohistochemistry. Kumar in his review cases of RT from 8 years' data fails to distinguish amongst Riedel's thyroiditis, thyroid malignancy and other benign thyroid conditions through FNA.[7] Management of RT is a challenging task. This is mostly because of the rarity of the disease and the scarcity of adequate data in the literature. Lots of literature suggests that high dose corticosteroids as monotherapy or in combination with levothyroxine are very effective.[8] Steroids decrease the size and hardness of the goitre. Tamoxifen, a TGF-B stimulation modulator, also helps by inhibiting fibroblast proliferation. It is used in doses of 10 to 20 mg every 12 hours. It decreases the goitre size in patients who have poor responses to steroids. Our case responds to the combined treatment of steroids and tamoxifen. Patients who are refractory to both steroids and tamoxifen may be benefited from rituximab, a monoclonal antibody against protein CD20.[9]

Indication of surgery is basically for two reasons; one for obtaining the tissue sample for histopathological examination, and secondly for relief of compression or stenosis at the level of trachea or oesophagus. Low dose radiation therapy is useful if RT is totally refractory to the above medications.[10]

Reidel's thyroiditis is a progressive disease; the disease may appear in other extra thyroid regions after the discontinuation of medication. As per Papi et al.,[5] around 30% of RT patients develop other fibrosing disorders over 10 years of follow up, though the disease-specific mortality is very low. Our patient, though was healthy after 4 years, longer follow up is required to see any disease progression.

Conclusion

Awareness of RT should be suspected in patients presenting with hard thyroid mass with compressive symptoms. Diagnosis may be demanding due to unknown aetiology and non-specific overlapping symptoms. Medical management with high dose corticosteroid followed by tamoxifen should be used to control the disease. Extensive surgery is not required in most cases.

Conflict of Interest: None

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