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Riedel's Thyroiditis: Diagnostic and Therapeutic Difficulties

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ABSTRACT:

Riedel's thyroiditis: Diagnostic and therapeutic difficulties

Objective: We aimed to discuss the diagnostic and therapeutic difficulties of Riedel's thyroiditis case which is a rarely seen disease of thyroid gland.

Case: A 43-year-old female patient who admitted to the hospital with a firm, painless midline mass, and shortness of breath. Diagnosis was made by radiographic imaging, fine needle aspiration biopsy, tru-cut biopsy and open biopsies. Surgical treatment was not planned due to extensive extrathyroidal spread of the disease. Systemic steroid treatment was decided to be initiated to the patient, who had tracheotomy due to tracheal compression, after examining the medical treatment protocols reported in the literature. A distinct decrease in the size of the thyroid mass was noticed at follow up.

Conclusion: Riedel thyroiditis is diagnosed in the presence of histopathological findings as well as radiological and physical examination findings. For diagnosis, primarily, it is important that the disease is suspected and that thyroid malignancies are excluded. Surgical treatment is not recommended because of the high morbidity except for the symptoms of compression, but there is also no standard medical treatment protocol. Therefore, reporting and discussion of the treatment choices of Riedel's thyroiditis on an individual basis will be beneficial.

Keywords: Riedel's, thyroiditis, thyroid gland

ÖZET:

Riedel tiroiditi: Tanı ve tedavide karşılaşılan güçlükler

Amaç: Tiroid bezinin nadir görülen bir hastalığı olan Riedel tiroiditi tanısı konulan bir olgunun tanı ve tedavi seyri sırasında karşılaşılan güçlüklerin tartışılması amaçlandı.

Olgu: Boyunda orta hatta ağrısız, sert kitle ve nefes darlığı şikayeti ile başvuran 43 yaşında kadın hastaya yapılan radyolojik görüntülemeler, ince iğne aspirasyon biyopsisi, tru-cut biyopsi ve açık biyopsiler ile tanı konuldu. Hastalığın yaygın ekstratiroidal yayılımı nedeniyle cerrahi tedavi planlanmadı. Trakeal kompresyon nedeniyle trakeotomi açılan hastaya, literatürde bildirilmiş medikal tedavi protokolleri incelenerek, sistemik steroid tedavisi başlanmasına karar verildi. Takiplerinde tiroid kitlesi boyutlarında belirgin regresyon izlendi.

Sonuçlar: Riedel tiroiditi tanısı histopatolojik bulguların yanısıra, radyolojik ve fizik muayene bulguları eşliğinde konulmaktadır. Tanıda öncelikle, hastalıktan şüphelenilmesi ve tiroidin habis hastalıklarının dışlanması önemlidir. Cerrahi tedavi, bası semptomları dışında, yüksek morbidite olasılığı nedeniyle önerilmezken standart bir medikal tedavi protokolü de bulunmamaktadır. Bu nedenle olgu bazlı tedavi seçimlerinin bildirilmesi ve tartışılması yararlıdır.

Anahtar kelimeler: Riedel, tiroidit, tiroid bezi

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INTRODUCTION

Riedel's thyroiditis is a rare, invasive thyroiditis with normal thyroid tissue displaced by dense fibrous connective tissue. Its incidence was reported as 1.06/100.000 (1). Women are three times more affected than men (2-5). Pathophysiology and etiology of the disease are still unknown. There is no standard protocol for Riedel's thyroiditis since there are only a few published case reports and case series.

In the presented case, it is aimed to examine the diagnosis and treatment of this rare disease and to discuss the difficulties encountered.

CASE

A 43-year-old female patient admitted to our outpatient clinic with complaints of firm, painless swelling at her neck and shortness of breath for 3-4 months. The patient's history revealed no any other characteristics except the use of 10 pcs/day cigarettes. On the ear, nose and throat examination, the thyroid gland was diffusely enlarged and as stiff with palpation. An expansile hypoechoic mass surrounding the left main carotid artery, which enlarged both lobes and isthmus of the thyroid gland and narrowed the tracheal air column, was observed in the neck ultrasonography of the patient (USG). Thyroid lymphoma and anaplastic thyroid carcinoma were suggested to be of differential diagnosis and biopsy was recommended. In color Doppler USG, the lesion observed in the thyroid parenchyma was stated to be hypovascular.

It was stated that homogeneous parenchymal echogenicity was preserved only at the subcapsular area at posterolateral part of the thyroid's right lobe.

Contrast-enhanced neck tomography (CT) revealed a 40x62x39 mm soft tissue mass lesion that was epicentrically located in the left lobe bed of the thyroid and medially invaded the thyroid right lobe by surrounding the trachea. The mass was surrounding the left main carotid artery and displacing the jugular vein towards anterolaterally. The lesion which was also surrounding the trachea, was narrowing the lumen at advanced level. Severe stenosis was observed at the level of trachea segment of 2 cm, starting from the cricoid cartilage level up to the caudal. No border distinction could be made between the mass and cervical esophagus at the posterior (Figure-1,2). No lymphadenopathy was observed in the pathologic aspect and size at the neck. Magnetic resonance imaging (MRI) could not performed due to claustrophobia.

In the presence of these findings, the patient was hospitalized to our clinic. In routine laboratory tests, leukocyte counts were within normal limits, while C reactive protein (CRP) was increased (25.12 mg/L). TSH was high (6.7 uIU/ml), free T3 and T4 were within normal limits. Anti-thyroid peroxidase (anti-TPO) was measured in the normal range and anti-thyroglobulin was measured as high (545.6 IU/ml). Calcitonin levels were within normal limits.

The imaging features of the lesion were primarily arousing a malignant tumor. For this reason,

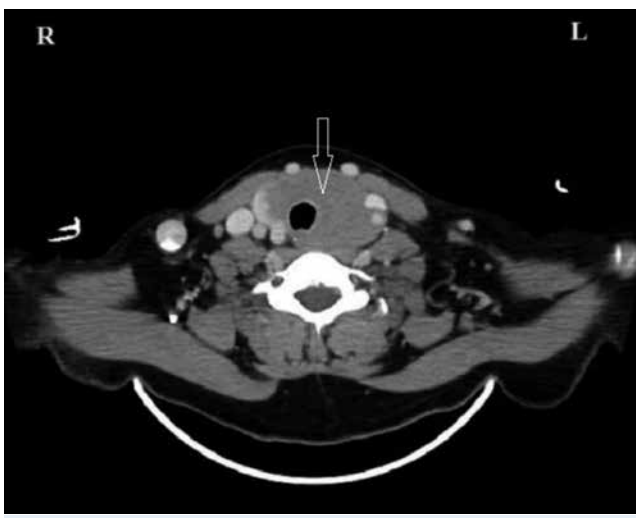

Figure-1: CT image of the mass located at the thyroidal lodge, before medical treatment

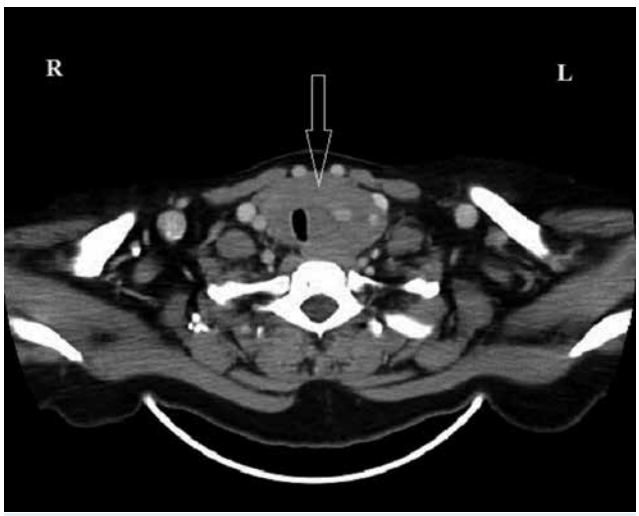

Figure-2: Tracheal pressure and repulsion towards right before medical treatment. A hypodense area of 15x15 mm in the central part of the lesion. Deletion of the border between esophagus and trachea. Mass surrounding the left main carotid artery and anterior displacement of the left jugular internal vein

cytopathological sampling was decided. Fine needle aspiration biopsy (FNAB) of the right and left lobe under USG reported hypocellular smear and diagnosis could not be made. A true-cut biopsy was planned upon this, and the results of the biopsy revealed active chronic inflammatory findings accompanied by intense fibrosis. Thyroid tissue was not seen in the tissue samples. Immunohistochemical staining showed no staining with pancytokeratin, TTF-1, and CD30. Staining was observed with CD3 and CD20 in lymphocytes and histiocytes. Neoplasia could not be diagnosed with the present findings and it was indicated that the findings may be related to fibrosis due to Riedel's thyroiditis (Figure-3). It was suggested that the lesion should be evaluated in the presence of clinical and radiological findings.

However, due to the compression of the trachea, for the safety of the respiratory tract, a tracheotomy was performed. During the tracheotomy, an open biopsy was also made of the thyroid gland. Pathology results of open biopsies were also reported as dense fibrosis and mild lymphocytic infiltration. Patient who was thought to have Riedel thyroiditis in the light of all findings was also referred to endocrinology and her treatment options were examined. The surgical treatment has been decided to be limited to tracheotomy because of the invasion of neck soft tissues by extrathyroidal spread of the mass and the compression would have increased the morbidity. Because there was no standard medical treatment protocol, it was decided first to initiate systemic steroid therapy, by screening the literature and reviewing the case-based treatments, then repeating the imaging techniques at 4th week to follow up the size of the mass. The patient was treated parenterally with methylprednisolone 60 mg/day, reduced by 4 mg per week, and levothyroxine 50 mg/day, and esomeprazole 40 mg/day. A salt-free diabetic diet was suggested. The blood sugar of the patient was also followed with 4x1 protocol. At the CT performed at 1st month of medical treatment, it was decided to continue the current medical treatment protocol on the basis of the fact that the size of the mass in the thyroid lodge is regressed (29x33x52 mm). The patient was discharged at the 6th week of treatment to be followed up by endocrinology and otorhinolaryngology outpatient clinics after decannulation and closure of the tracheostomy.

The patient's systemic steroid therapy has been completed at 13th week. The size of the mass was measured as 20x28x48 mm in the post-treatment CT of the patient who came to the outpatient clinic after the end of the medical treatment. Tracheal air column was open. In addition to the decrease in mass size, the surrounding of the left main carotid artery and the border between the trachea-esophagus

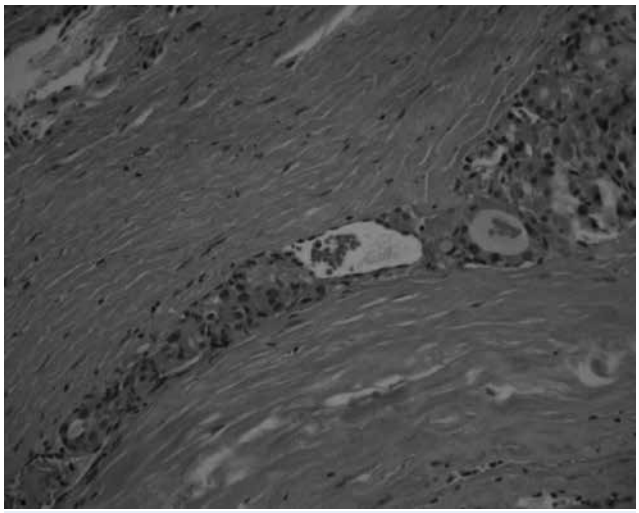

Figure-3: Histopathological appearance of biopsy material taken from lesion (Microscopic magnification x200, Hematoxylin-Eosine)

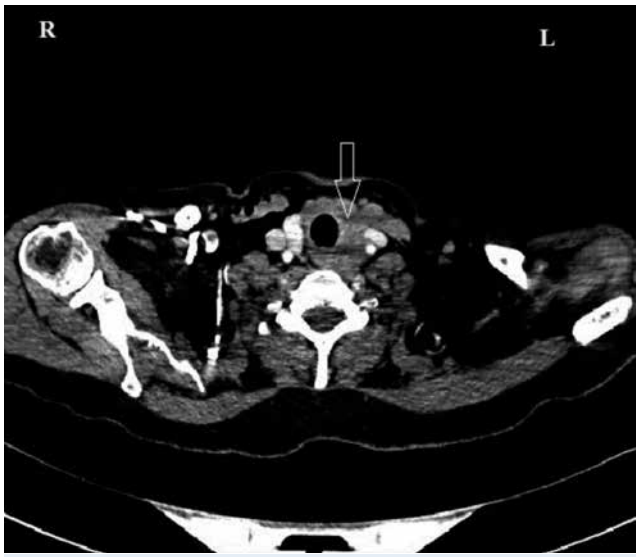

Figure-4: CT images of the mass located at the thyroid lodge at the end of systemic steroid therapy (13th week)

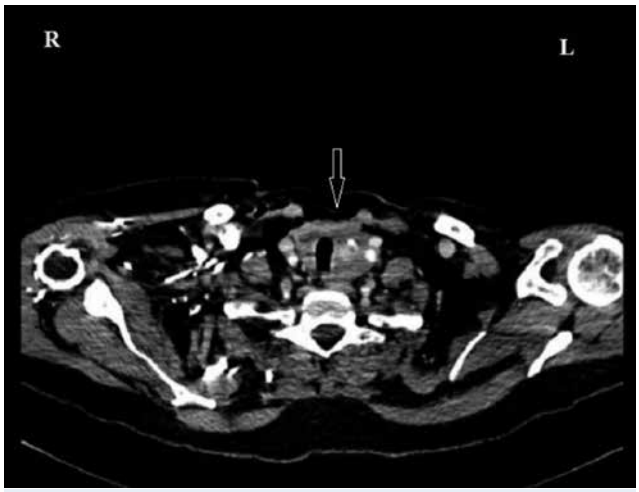

Figure-5: Significant regression in mass size and decrease in tracheal compression following medical treatment. Clarification of the left main carotid artery and the border between esophagus and trachea

can be seen more clearly (Figure-4,5). On physical examination, it was observed that there was a significant decrease in the stiffness of the thyroid gland with palpation. CRP levels reduced to normal range in blood samples (0.32 mg/L). A decrease in anti-thyroglobulin level was also observed (415.6 IU/ml). The TSH level of the patient who had thyroid hormone replacement also turned to normal levels (2 uIU/ml).

DISCUSSION

Riedel's thyroiditis is a rare, invasive thyroiditis in which normal thyroid tissue is displaced by dense fibrous connective tissue. Its incidence was reported as 1.06/100.000. It was found in 37 cases of 57,000 thyroidectomies performed in the Mayo clinic (1). Women are affected three times more than men (2-5). Although the pathophysiology and etiology of the disease is still unknown, autoimmunity is found responsible (2).

The main complaint of the patient is the presence of painless, stiff mass. This mass characteristically extends beyond the thyroid capsule and involves the adjacent neck tissues such as carotis, trachea, strep muscles, and nerves. This appearance is confused with neoplastic diseases of the thyroid (6). Similarly in the imaging technique of the presented case, in the thyroid and perithyroidal regions, structures such as carotis, trachea, cervical esophagus and juguler vein were seen to be affected the fibrotic process and were giving an impression of neoplasm. In order to distinguish between neoplastic masses, diagnosis was tried to be made by FNAB, true-cut biopsy and open biopsy.

The macroscopic and histopathologic appearance of the disease described in the literature is as follows: fibrous thyroid gland stiff as wood, fibrosis extending beyond the thyroid capsule to the perithyroidal neck structures, microscopically without accompanying granulomatous inflammatory component, displacement of fibrous tissue with normal parenchyma of the thyroid (6). Lymphoma, anaplastic carcinoma, sarcomas, and Hashimoto's thyroiditis should definitely be included in the differential diagnosis of Riedel's thyroiditis (7-10). In the present case, the absence of atypical cells in the biopsy specimens, intense concentration of fibrosis almost as if displaced totally by normal thyroid tissue, extansion to the outward of the thyroid capsule and neck and the fibrotic process affecting the adjacent neck structures at a level of compression made us think of Riedel's thyroidits as the first choice.

Compressive symptoms are common in patients and usually manifest as dyspnea, dysphagia, stridor, and hoarse voice. In the present case, there was the trachea being pushed to the right and complaint of dyspnea due to compression; the respiratory tract was taken under control by opening a traceostomy. In Riedel's thyroiditis imaging tools usually give nonspecific findings. Thyroid gland in USG appears homogeneous hypoechoic due to massive fibrosis (11). This appearance is nonspecific and can also be seen in Hashimoto thyroiditis, lymphoma, or carcinomas. The lesion was hypoechoic in evaluarion with USG in our case. Doppler examination revealed a hypovascular appearance. Biopsy was recommended to exclude malignancies such as lymphoma and anaplastic carcinoma.

CT is valuable in evaluating the extension of fibrosis, compression and repulsion of trachea or esophagus, and involvement of the large vessels such as carotis and juguler vein. The lesion can be

seen at different densities from hypodense to normal. Invasion of adjacent tissues shows a mild positivity after contrast agent administration (12). CT scan of the presented patient clearly showed repulsion and compression of the trachea to the right, deletion of the border between the trachea and cervical esophagus, and all around involvement of the left main carotid artery.

There is no standardized treatment protocol in Riedel's thyroiditis, as there are few case reports and small case series in the literature. Surgical approach is thought to be the first choice in the majority of patients, to relieve obstructive symptoms and for diagnosis. If total thyroidectomy is not possible in surgical treatment, debulking surgery is performed, in which the compression is reduced and the surgery is usually limited to thyroid isthmusectomy (2,13-15). The obliteration of tissue planes due to fibrotic process increases the risk of permanent hypoparathyroidism and recurrent nerve injury. Thus, the surgical procedure becomes problematic. Despite limited surgical treatment, permanent nerve injury and hypoparathyroidism have been reported in 7 (39%) patients of a case series of 18 patients. The general opinion is that a large surgical resection in Riedel's thyroiditis is not appropriate (4,15-17).

After diagnosis is confirmed, stopping the progression of the disease is the main purpose of medical treatment. However, lack of a validated treatment protocol with controlled clinical trials makes medical treatment difficult. However, the use of glucocorticoids and tamoxifen has been reported in the literature (18-21). Glucocorticoids are preferred in first-line treatment (22). There are studies reporting that steroid therapy is effective in improving symptoms and reducing mass volume (20,23,24). Steroids were administered as 100 mg/day, 15-60 mg/day, with variable doses, and positive results were obtained (2,10,19,24-26). The use of tamoxifen in medical treatment is also an effective treatment (21,25,27). Potential mechanism is thought to be the inhibition of induction of autocrine secretion of TGFβ1, a potent growth inhibitor, and inhibition of fibroblastic activity (14,28,29). Successful results were obtained with 10-20 mg dose of tamoxifen in combination with steroid therapy or as monotherapy (21,26,30,31). In cases accompanied by hypothyroidism or hypoparathyroidism, L-thyroxine replacement, calcium and calcitriol should be included in the treatment. This case is important from the point in presenting the diagnostic stages of Riedel's thyroiditis and the course of the treatment. Due to the small number of cases and case series in the literature, it is difficult to establish an appropriate treatment protocol. A multidisciplinary approach with endocrinology should be preferred. It has been reported that successful results are obtained with medical treatment. Surgical treatment involves potential risks because the disease can invade the neck tissues and delete the tissue planes. For this reason, surgical resections should be done in a limited fashion only to relieve pressure symptoms.

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