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Case Presentation
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A Large Riedel's Thyroiditis Causing Impending Upper Airway Compromise in a 55-Year-Old Female: A Case Report

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Abstract

Riedel's thyroiditis is a diagnostic dilemma. It is a rare disease made up of an invasive fibrotic and inflammatory process of the thyroid gland that can lead to destruction and extend into the adjacent neck structures. It has been associated with systemic fibrotic disorders. We report a 55-year-old female who presented to the emergency department with a history of painless enlarged neck swelling and compressive symptoms (dyspnea and dysphagia) in a short period. The patient was admitted to the ICU for monitoring and observation. A CT neck and chest was done, and the patient was scheduled for biopsy in the next day. Laboratory tests of TFT and auto-antibodies were euthyroid, and no antibodies were detected, respectively. Histopathology results revealed only dense fibrosis, no malignant cells, and extension into surrounding tissues. The patient started on conservative management, including hydrocortisone and anticoagulants. A few days her symptoms dramatically improved and transferred to the ward. Then the patient was discharged home on prednisone 40 mg orally once daily for two months and to be followed up in the clinic after one month. During her follow-up, she was compliant with her medication, and no relapse symptoms were noticed.

Keywords: Riedel's; Thyroiditis; Fibrosis; Airway; Lymphoma; Anaplastic; Prednisone

Introduction

Riedel's thyroiditis, or invasive fibrous thyroiditis, is a rare condition thought to originate from an autoimmune disorder [1]. It may appear as new or because of a prior diagnosis of Hashimoto's Thyroiditis (HT) or Graves' disease. In a series from the Mayo Clinic, nearly all individuals reported were women with an average age of 42 years and a significant smoking history [2,3]. Riedel's thyroiditis is now regarded as part of the spectrum of IgG4-

related sclerosing diseases. Histologically, it demonstrates dense keloid-like fibrosis that disrupts the normal lobular architecture of the thyroid gland while extending into neighboring extra thyroidal tissues, and over time, normal tissue is replaced by dense fibrous tissue. Compared to Hashimoto's Thyroiditis (HT) fibrous variant, Riedel's is notable for more pronounced invasion **[4]**. Accompanying inflammatory infiltrate varies but consists mostly of lymphocytes and plasma cells. This study aims to highlight Riedel's thyroiditis in the differential diagnosis of any sudden progressing neck swelling and the outline of challenging management options.

Case Presentation

This 55-year-old woman, with no known chronic illness, arrived at the emergency department reporting a rapid increase in neck swelling along with compressive symptoms. She has experienced neck swelling for over two years, which then suddenly enlarged, was painless, and accompanied by shortness of breath and difficulty swallowing a few weeks prior to the presentation. There is no significant family history of thyroid cancer or endocrinopathy, nor any history of radiation exposure. Upon assessment, the patient was positioned at 45 degrees on the bed, with vital signs showing SPO_2 at 93% on 5L of O_2 delivered through a face mask. A prominent, diffuse stone-like swelling on the right side extended retrosternal, featuring a hard surface. No cervical lymphadenopathy was noted, and the trachea was shifted. The patient was admitted to the ICU for monitoring and additional assessment of airway safety. The lab results featured Thyroid Function Tests (TFT), TSH, T4, T3, euthyroid status, absence of auto-antibodies, and normal IgG4 levels. The chest and neck CT scan revealed a significant thyroid enlargement displacing the trachea to the left, mild tracheal narrowing with patent airway, and encasement of major blood vessels **(Figure A and B)**. The patient underwent a US-guided Tru-Cut biopsy, and the findings indicated dense, fibrotic tissue that disrupted the normal tissue without signs of malignant cells. The morbidity committee discussed the case for additional management, and it was unanimously decided to initiate hydrocortisone 200 mg IV OD for the patient for several days. The patient showed improvement following the hydrocortisone, and her shortness of breath subsided, then the patient was transferred to the wards in stable status and remained in hospital for one week, and thereafter, The patient was sent home on prednisone 40 mg orally once daily for two months and scheduled for a follow-up in the clinic after one month to evaluate the treatment's effectiveness or necessity for surgical intervention for decompression effect. There was notable progress in medical treatment, and no signs of relapse after two months.

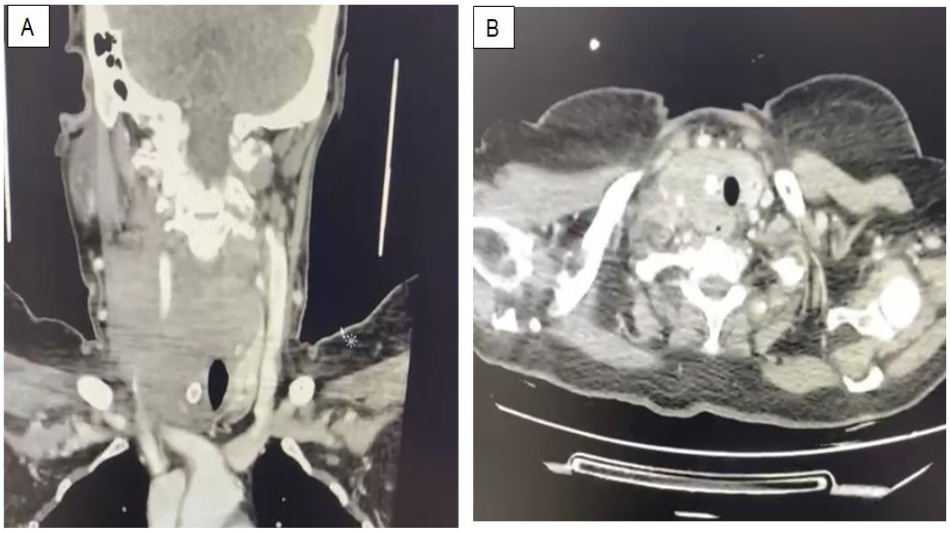

Figure A and B: Coronal and axial cut showed huge mass from the thyroid lobe and isthmus that measured around 5*6.3*4.5 cm not separable form the sternocleidomastoid muscle and tracheal division to the left side and but patent airway this mass effect causing major blood vessels encasement and compressed included (right jugular vein, common carotid, and the bifurcation) with multiple sub-centimetric cervical lymph node

Discussion

The hallmark of Riedel's thyroiditis is the replacement of normal tissue with dense fibrotic tissue, which involves extra-thyroid structures, e.g, trachea, esophagus, recurrent laryngeal nerve, major vasculatures, and neck musculatures [5]. This can cause thyroid and extra-thyroid structures to be immobile and fixed, and described as stone-hard or woody palpation. The most important differential diagnosis in our case was thyroid lymphoma and anaplastic thyroid cancer the Riedel's thyroiditis can be misdiagnosed with the cause mentioned earlier, and the vital step in such a case is to secure and protect the airway and to get a final diagnosis [6,7]. The options for airway protection are either intubation or tracheostomy to secure the airway. The definitive diagnosis is made by biopsy in such a case, by Tru-Cut vs. open, depending on the individualized case-by-case [8]. The FNA in such patients as ours, which is present with an enlarged thyroid hard mass like a stone, might be inadequate. There is no standardized management due to its rarity in the literature, and the outcome. The mainstay of treatment in our case was prednisone 40 mg orally once daily. In general, steroids are the cornerstone in management due to their anti-inflammatory effects on tissue. *Valeria Navarro-Sánchez* et al. in their study, we found that oral tamoxifen is another interesting option in Riedel's thyroiditis. Tamoxifen is an antiestrogen. This medication has been shown to enhance tumor growth factor-β, and a well-recognized suppressant of cell proliferation [9-19]. Another option for the agent is Mycophenolate mofetil is an immunosuppressant with anti-fibrotic effects that is used therapeutically in systemic fibrosis. It transforms into mycophenolic acid, which prevents the production of antibodies by T and B lymphocytes. Levy et al. effectively treated a case of Riedel thyroiditis resistant to tamoxifen and prednisone using a combination of mycophenolate

and prednisone [10,11]. The crucial aspect is to continue the patient on anticoagulants to prevent venous thrombosis caused by the mass effect impacting the major blood vessels [12,17]. In such cases, the surgical intervention is warranted only if there are symptoms of compression or concerns about malignancy, as the absence of tissue planes between the thyroid and the fibrosis complicates the procedure, resulting in a complication risk of 99-100% [18,19].

Conclusion

Riedel's thyroiditis can be misdiagnosed as an aggressive thyroid cancer (anaplastic thyroid cancer) or thyroid lymphoma. The successful treatment is prednisone alone or oral tamoxifen +/- immunotherapy can be added if there is no improvement or relapse symptom. The surgical role in such cases is a decompressive effect only, and can be considered if there are compressive symptoms or suspicion of malignancy. The surgery here is extremely risky and difficult due to the lack of tissue planes between the thyroid tissue and the fibrosis.

Highlights

- The thyroid biopsy is a vital tool to rule out anaplastic cancer or thyroid lymphoma
- Prednisone is the choice of treatment
- Surgical option is the 2nd line in case there are a compressive symptoms or suspicion of malignancy

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