Tracheal invasion and cardiopulmonary compromise from primary thyroid lymphoma

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Abstract

Rapidly expanding thyroid lesions with tracheal invasion are typical characteristics of anaplastic and high-grade thyroid carcinomas. However, primary thyroid lymphoma (PTL) must also be considered as a differential. We present a rare case of PTL with direct invasion and compression of the trachea resulting in pulmonary oedema and cardiomyopathy.

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Key Clinical Message

This case highlights the diagnostic challenge of PTL in patients with a rapidly expanding anterior neck mass. In addition to tracheal invasion, progressive airway compression can lead to pulmonary oedema and cardiomyopathy.

Abstract

Rapidly expanding thyroid lesions with tracheal invasion are typical characteristics of anaplastic and high-grade thyroid carcinomas. However, primary thyroid lymphoma (PTL) must also be considered as a differential. We present a rare case of PTL with direct invasion and compression of the trachea resulting in pulmonary oedema and cardiomyopathy.

Introduction

Rapidly expanding thyroid lesions with tracheal invasion are typical characteristics of anaplastic and high-grade thyroid carcinomas. However, primary thyroid lymphoma (PTL) must also be considered as a differential. Aggressive thyroid lesions can compromise the airway through compression and/or direct invasion of the tracheal wall.

We present a rare case of PTL which presented with compression and invasion of the anterior tracheal wall causing acute respiratory distress secondary to negative pressure pulmonary oedema with cardiomyopathy.

Case report

A 57-year-old woman with known hypothyroidism presented to her GP with a few months' history of orthopnoea and intermittent hoarseness. An ultrasound showed right sided thyroid enlargement with an isthmus nodule extending to the left lobe. She developed progressive tightness in the neck and was seen in the rapid access ENT clinic. Flexible nasendoscopy examination was normal and the patient underwent an urgent ultrasound-guided Fine Needle Aspiration Cytology (FNAC), which suggested possible lymphocytic thyroiditis. Subsequent core-biopsy showed scanty lymphocytes and was non-diagnostic.

Nineteen days following ENT review, she presented to our Emergency Department with worsening shortness of breath. The patient was diagnosed with type 2 respiratory failure with acidosis and raised troponin. A computed tomography pulmonary angiogram (CTPA) on admission demonstrated severe ground glass shadowing. She was therefore admitted under the medical team with working diagnoses of atypical pneumonia. An echocardiogram demonstrated a reduced ejection fraction of 32% with left ventricle apical hypokinesia and lateral wall akinesia. Cardiac MRI demonstrated an apical left ventricle hypertrophy suggestive of cardiomyopathy. As a result, a diagnosis of Takutsubo-like stress cardiomyopathy was made.

Nineteen days from admission, the patient suffered a cardiac arrest. Whilst spontaneous ventilation was achieved through cardiopulmonary resuscitation, worsening type 2 respiratory failure required intubation and mechanical ventilation in the Intensive Care Unit (ICU). The cause of arrest was attributed to acute respiratory distress syndrome secondary to acute pulmonary oedema.

In light of her cardiorespiratory deterioration, the admission CTPA was re-reviewed where a large goitre with heterogenous enlargement was noted with significant tracheal compression resulting in a minimal lumen size of 6mm (figures 1 & 2). No direct invasion of the trachea was seen on imaging. The CT and clinical impression was of a rapidly progressive and aggressive thyroid malignancy such as anaplastic carcinoma, rather than thyroiditis as suggested on ultrasound. In view of the airway obstruction, the aetiology of the ground glass appearance of the lungs was revised to pulmonary oedema secondary to negative pressure respiration rather than an infective cause.

On day two of intubation, ENT were approached regarding surgical management of the airway as the patient was deemed unsuitable for extubation due to the degree of tracheal compression. Therefore, a hemithyoidectomy of the larger right lobe of thyroid was planned to decompress the trachea, gain histological diagnosis and allow extubation of the patient. Intraoperatively the thyroid lesion was more consistent with an infiltrative pathology involving the strap muscles. The thyroid was grossly enlarged, homogeneously firm with an appearance more suggestive of lymphoma rather than carcinoma. The trachea was identified distally,

and the thyroid cartilage was exposed to gain control of the field. The infiltration extended laterally toward the carotid sheath, hence an attempt at a formal hemithyroidectomy was not feasible. On dissecting the thyroid tissue from the trachea, it was evident on the right lateral aspect there was clear invasion into the lumen of the trachea. This was inspected using a 0-degree and 30-degree Hopkins Rod. We decided a wedge resection of the thyroid down to the trachea and the formation of a tracheostomy would be the safest option. A size 8-0 cuffed Shiley tracheostomy tube was inserted.

The patient stepped down to ward-based care after 17 days on ICU. Histological diagnosis confirmed diffuse large B-cell lymphoma. Since her discharge, she has completed six cycles of R-CHOP chemotherapy (four cycles at full dose) and will have two further cycles of Rituximab complete her chemotherapy treatment. She has been followed up closely by Cardiology, where her ejection fraction had improved from 32% to 62% and will undergo a repeat cardiac MRI and echocardiogram and decannulation of her tracheostomy upon completion of chemotherapy.

Discussion

Thyroid lymphoma can be classified into primary and secondary (non-thyroidal lymphoma metastasises to the thyroid gland). PTL is rare, accounting for less than 5% of all thyroid malignancies. The thyroid gland does not typically contain lymphoid tissue, therefore PTL tends to occur in pathological glands. Autoimmune thyroiditis (Hashimoto's thyroiditis) is the most common risk factor and is associated with 80% of cases of PTL and confers 40-80 times the risk of developing PTL compared to those without thyroiditis.

Whilst traditional FNAC plays an important role in diagnosing thyroid nodules, it is of limited value in PTL due to the challenge in distinguishing between lymphoma, lymphocytic thyroiditis and anaplastic thyroid carcinoma. However, advances in flow cytometry and immunohistochemistry have increased the sensitivity of FNAC in diagnosing PTL. Core needle and incisional biopsy techniques provide significant higher sensitivity and classifies 95% of lymphomas in typical cases. Computed tomography can suggest a diagnosis where anaplastic thyroid carcinoma displays more heterogenous attenuation within the lesion often with evidence of calcification and necrosis, whereas PTL and involved lymph nodes have a more homogenous appearance.

Conclusion

This case highlights the diagnostic challenge of differentiating between PTL and anaplastic thyroid carcinoma in patients with a rapidly expanding anterior neck mass. Tracheal invasion is a feature seen more commonly in anaplastic thyroid cancer due to release of proteinases from neoplastic giant cells, which resemble osteoclasts associated with tracheal cartilage. Necrosis is uncommon in PTL and whilst extension beyond the thyroid capsule is relatively common, erosion of adjacent structures is unusual.

Invasion of the trachea from PTL is very rare, especially prior to treatment, with fewer than 5 cases reported in the literature. Furthermore, this case demonstrates the potential cardiopulmonary sequelae of progressive airway compression which resulted in negative pressure pulmonary oedema and cardiomyopathy.

Author Contribution

Sean Fang: Literature review and discussion review.

Lisi Hu: write up of case report.

Karan Kapoor: Supervising Consultant.

Conflict of Interest and Ethics Approval

We have no conflicts of interest or financial sources to declare.

Ethics approval was not required for this article.

References

Figure captions

Figure 1: CT scan showing large thyroid lesion with significant tracheal compression

Figure 2: post-tracheostomy CT scan showing the thyroid lesion causing tracheal deviation and possible invasion.