Case Report

Neglected Retrosternal Goitre: A management Dilemma

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Abstract
Retrosternal goiters, first described by Albrecht von Haller in 1749, defined as one that descends below the thoracic inlet, or has more than 50% of its volume below this level. Retrosternal Goitres with compressive respiratory symptoms may cause misdiagnoses and delay in the treatment. Here, we are reporting a young female with goitre who was taking treatment for respiratory symptoms was then diagnosed to have retrosternal extension. She was treated with surgical resection through median sternotomy approach. She was symptomatically relieved of respiratory symptoms.

Keywords: retrosternal goitre, median sternotomy

1. Introduction
Mediastinal goiters were first described by Albrecht von Haller in 1749, as the extension of the thyroid tissue below the upper opening of the chest.1 As goiters grow they attain a mediastinal position by extending through the thoracic inlet. As these mediastinal goiters grow very slowly and are present over many years, they often go unrecognized either because the thyroid mass is managed conservatively by suppressive therapy or the compressive symptoms misinterpreted as cardiopulmonary symptoms, leading to the late diagnosis or even misdiagnosis.3 Here lies the importance of taking a detailed history, carrying out a thorough clinical and investigative evaluation and maintaining an open mind. Advancing age is associated with increased medical co-morbidity, implying that operation at an earlier stage of goitre development may be associated with reduced complications related to co-existing disease2. Here, we report an interesting case of 37 year old female with neglected retrosternal goitre.

2. Case Report
A 37 year old female patient presented to our OP department 6 months back with a swelling over the front of neck for 15 years and breathlessness for 5 years. The swelling was gradually progressive in nature. It was associated with loss of weight, restlessness, tremors and anxiety, which were clinically suggestive of secondary thyrotoxicosis. She also had history of breathlessness for 5 years which was treated as Chronic Obstructive Pulmonary Disease from a local hospital. Local examination revealed the swelling to be a MNG with nodularity involving both lobes of thyroid extending deep to the anterior border of both sternocleidomastoid and lower border of the thyroid made out on deglutition (due to overhanging of the nodule). Her TFT showed hyperthyroid state and FNA and cytologic analysis were consistent with features of Colloid goitre-thyroid gland. Patient was treated with antithyroid drugs and biochemical euthyroid state was achieved. Surgery (total thyroidectomy) was planned. Routine X ray chest was done as preoperative work up which suggested widening in the superior mediastinum. Possibility of retrosternal extention was suspected and CT chest including neck was taken. CT scan suggested large nodular thyroid with retrosternal extension. Total thyroidectomy with median sternotomy by ‘T’ shaped incision was planned. Intraoperatively thyroid was seen extending into the retrosternal region. It was a dumb bell shaped
mass with constriction at the sternal notch. Postoperative period was uneventful and patient was symptomatically relieved of breathlessness and thyrotoxicosis.

3. Discussion

Retrosternal goitre is most commonly defined as one that descends below the thoracic inlet, or has more than 50% of its volume below this level. The natural history of retrosternal goitre is slow, progressive growth, commonly leading to presentation in the fifth or sixth decade of life. Women are affected 3-4 times more often than men. Presence of retrosternal goitre is documented in 2-19% of all thyroidectomy surgeries.

Retrosternal goiters can be either classified as primary or secondary. Primary retrosternal goiters are intrathoracic goiters that arise from aberrant thyroid tissue which is ectopically located in the mediastinum. They are rare, representing 1% of all retrosternal goiters. Secondary retrosternal goiters develop from the thyroid located in its normal cervical site. The downward migration of the secondary retrosternal goitres to the mediastinum is facilitated by negative intrathoracic pressure, gravity, traction forces during swallowing and the presence of anatomical barriers preventing the enlargement in any direction. The vast majority of intrathoracic goitres are located anteriorly in the visceral compartment and the undersurface of the manubrium of the sternum on the cephalad aspect of the great vessels. Goitres are more likely to extend to the posterior mediastinum in patients with a prior history of thyroidectomy, which could be owing partially to violation of fascial planes sealing this compartment by the previous surgical procedure.

The most reported pathology for intrathoracic goitre is nontoxic multinodular goitre followed by follicular adenoma. The disease carries a significant risk of malignancy, mainly papillary carcinoma, in 1.4% to 2.1% of patients with mediastinal goiters.

Mediastinal goitre is of clinical significance because its growth between the sternum anteriorly and vertebral bodies posteriorly leads to impingement of surrounding structures and compressive symptoms. Tracheal compression, especially dyspnea, is the most common compressive symptom. Patients with large substernal goiters often become symptomatic when lying down or when the neck is extended. Exacerbation of respiratory symptoms may also occur when patient turns the head towards the side of the goitre. Acute tracheal obstruction with severe respiratory compromise may be observed occasionally. Dysphagia is seen more in cases where the mediastinal goitre extends posteriorly. If dysphonia is the presenting symptom, it indicates malignancy, which in turn increases the risk of nerve injury and sternotomy during surgery. Moreover, a patient presenting with dysphagia with prior history of thyroidectomy gives a suspicion of posterior mediastinal extension.

Cervical goitres are amenable to careful clinical and ultrasonographic examination and needle biopsy of suspicious areas, with cytological determination of malignant nodules, leading to patient selection for surgery. Retrosternal components of goitres are not easily imaged by ultrasound due to artefact generated by bony structures. Similarly, intrathoracic nodules are inaccessible to needle biopsy in most instances. This makes the exclusion of malignancy fraught with difficulty in retrosternal goitres. CT scanning as the imaging method of choice, to assess clinically suspected retrosternal goitre. Although, it did not completely identify all those with retrosternal extension, it was useful for the detection of tracheal deviation and compression, which alone would be strong indicators for surgery.

Compressive symptoms resulting from a retrosternal goitre, such as cough, dyspnoea, choking symptoms and dysphagia are regarded as absolute indications for surgery. In addition, radiological indicators of compression, like tracheal deviation, would be seen as an indicator for surgery. But one should always remember that the correlation between symptoms and presence of tracheal deviation, size of goitre or extent of substernal extension as assessed by computed tomography may be poor. Hence, the policy of using radiological features as indicators for surgery, may not therefore, be appropriate in patients with subtle compressive symptoms. In addition to chronic compressive symptoms, the potential for acute airway obstruction needs to be considered. This may arise from haemorrhage within the thyroid gland or secondary to prolonged mechanical pressure with acute laryngeal oedema and congestion. Development of this complication can have catastrophic consequences, providing a clear rationale for thyroidectomy in patients with retrosternal goitre.

4. Conclusion

Thyroid swelling with doubtful retrosternal extension should be initially screened by a routine chest X-ray and may be confirmed by a CT scan if required. Early identification and proper management of the retrosternal goitre can avoid potential morbidity and mortality. Surgery is the only effective treatment for retrosternal goitre.
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