



RETROSTERNAL GOITRE WITH VARIED HISTOMORPHOLOGY: A CLINICOPATHOLOGICAL CASE REPORT

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ABSTRACT

The term such as retrosternal or substernal is used when a goitre extends below thoracic inlet. Retrosternal goitre is rare and accounts for 5–20% of thyroidectomy patients. Majority of the patients present with symptoms of dyspnoea as in our case. Also, patients can come with change of voice, stridor and a huge swelling. Most of the cases of retrosternal goitre can be removed with conventional collar incision, however sternotomy may be required in few cases. Our patient was euthyroid at presentation and a right hemithyroidectomy was done for her. A detailed histopathological evaluation (HPE) showed evidence of long-standing Adenomatoid goitre with extensive hyalinization, thickened capsule and secondary degenerative changes in the right lobe of thyroid and evidence of chronic lymphocytic thyroiditis in the sections examined from the portion of isthmus. The procedure was uneventful. The patient was doing well after three months of clinical follow up.

KEYWORDS

Retrosternal, dyspnoea, hyalinization, chronic lymphocytic thyroiditis, HPE

INTRODUCTION

Retrosternal, sub sternal or intrathoracic goitre is a rare form of thyroid presentation where the thyroid mass extends below the thoracic inlet. Some define that a thyroid gland should have extension more than 50% below the sternum to be labelled as retrosternal. They have a slow progression and longer course of illness. They present with symptoms of a huge swelling in the neck and because of its slow progression the peak incidence is 5th to 6th decade.^{1,3} Retrosternal goitre is more common in women.

Since mediastinum is a confined space, the enlarged substernal goiters can compress onto tracheal, esophagus and even vital large vessels causing respiratory problems, choking, change of voice, etc.^{4,6} With few exceptions they can be removed with conventional cervical approach. In few cases sternotomy may be required. Herein, we report a case of long standing right sided large retrosternal adenomatoid goitre in a 60-year-old female.

CASE REPORT

A 60-year-old female presented with history of a swelling in neck for last 10 years and breathlessness on exertion since past 6 months. The swelling was gradually progressive and was first noticed by the patient 10 years back. It was the size of a peanut then and it gradually increased to its present size. The patient also complained of fatigue and dyspnea during physical stress for last 6 months. Occasionally she had dry cough and choking like sensation.

General and systemic examinations were normal. On airway examination, neck movement and mouth opening were normal with Mallampati Grade III, thyromental distance of 7cm and intact teeth. Local examination revealed a diffuse swelling of right lobe of thyroid. The swelling was around 8 x 4 cm in size and lower border of the swelling could not be felt [Figure 1]. The swelling was firm to hard in consistency and moved up with deglutition. There was no cervical lymphadenopathy. On the left side no swelling or lymphadenopathy was seen. Ultrasound of the neck revealed multiple nodules in the right side of the thyroid gland.

Complete blood counts were within normal limit. The patient was euthyroid at presentation. Rest of the biochemical parameters were also within normal range for age. Fine needle aspiration cytology of the lesion was done. Smears examined were suggestive of multinodular colloid goitre with cystic changes on the right side.

Chest radiograph showed deviation of trachea to left. A detailed radiological evaluation was done. CT scan neck revealed thyroid gland on right side with areas of necrosis and calcification. The right lobe measured 8x4x4 cm; inferiorly right lobe was seen extending into retrosternal area. Medially the lesion was compressing the trachea (<15%) with narrowing and deviation (0.5-1cm) to the left. Widening of superior mediastinum was present left thyroid lobe was normal.

Indirect laryngoscopy and fiberoptic laryngoscopy showed bilateral mobile vocal cord movements during phonation and respiration. Baseline investigations were within normal limits.

Right hemithyroidectomy was planned for the patient. The patient was accepted for surgery under ASA class II risk status. Preoperatively, the patient was started on bronchodilator therapy, chest physiotherapy and incentive spirometry. Standard NPO orders were followed. Before shifting to operation theatre, nebulization of the patient with 3ml of 4% lidocaine and gargling with 2% lidocaine viscous was done. Awake fiberoptic intubation was done after adequate premedication (Inj. Glycopyrrolate 4mcg/kg i.v., Inj. Midazolam 0.5mg i.v.) and mild sedation (Inj. Dexmedetomidine 0.4-0.6mcg/kg/hr i.v.) of the patient after blocking glossopharyngeal, superior and recurrent laryngeal nerves. Very minimal indentation of the anterior tracheal wall, about 6 cm above the carina was seen. 7.0 mm ID cuffed flexometallic endotracheal tube was secured and bilateral equal air entry ensured after neck extension and positioning of the patient for surgery. Anaesthesia was maintained with N2O:O2:Isoflurane and i.v. Fentanyl and Vecuronium.

Surgery was uneventful. About 150 ml blood loss was replaced with crystalloids. On completion of the surgery, the patient was reversed with inj. Neostigmine i.v. Cuff leak test and tube occlusion tests were performed. Patient was extubated over fiberoptic after no tracheal wall collapse and normal movement of vocal cords visualized. No complications observed post-operatively. The surgical specimen was sent for histopathological examination [Figure 2A-B].

On gross pathological examination, the external surface of right lobe of thyroid was encapsulated and appeared asymmetrical with multiple lobules. The right lobe of thyroid measured 7.5x5x4 cm & portion of isthmus measured 4.5x1 cm. Cut section revealed grey-brown & grey-white areas along with cystic areas & haemorrhagic areas [Figure 3A]. Multiple sections showed involutional & hyperplastic nodules of

thyroid follicles, which were separated by thick and thin fibrotic septae [Figure 3B-D]. Few nodules showed extensive hyperplasia with mild overcrowding & overlapping of nuclei with mild atypia [Figure 4A]. Extensive hyalinization and bands of fibrosis were noted [Figure 3C]. Section from isthmus revealed lymphoid follicles with germinal center [Figure 4B]. Histomorphological features were consistent with diagnosis of Adenomatoid goitre with extensive hyalinization and secondary degenerative changes in the right lobe of thyroid. Also, section from isthmus showed evidence of chronic lymphocytic thyroiditis. On follow up, the patient was doing fine without any complications.

DISCUSSION

Goitre is considered retrosternal if the lower pole of thyroid is not palpable with the neck in hyperextended position. Most widely accepted classification of RG and management depending on its extension into mediastinum has been proposed by Huins et al.³ They defined three grades of goitre namely, grade I to the level of the aortic arch, grade II to the level of the pericardium and grade III below the level of the right atrium.³

Etiologically 85 – 95 % of retrosternal thyroid masses emerge as benign goitre. Retrosternal goitres are more common in women and symptoms appear mostly in 5th – 6th decade of life.^{4,6} There are some factors that help to understand the reasons and the etiology of extension of goitre downward the thoracic cavity. The mediastinum space size is not fixed yet it can lodge abnormally growing lesions. Hence, more the potential space in individual's mediastinum, higher the likelihood of retrosternal goitre. In addition, the intrapleural negative pressure and gravity could play a role in this mechanism. The vast majority of RG (85-90%) are in the anterior mediastinum. Posterior mediastinal goiters are uncommon comprising 10-15% of all mediastinal goiters.⁶

The patient becomes symptomatic depending on structures that are compromised. They may develop dyspnea and stridor as a consequence of slow progressive growth or sudden nodular hemorrhage resulting in tracheobronchial compression.^{4,6} Compression of vascular structures can cause vena cava superior syndrome with facial and upper body oedema. A positive Pemberton's sign could be seen due to superior vena cava obstruction. The sign is positive when bilateral arm elevation causes facial plethora.

The diagnostic methods of choice include traditional radiograph, ultrasound, CT scan. Retrosternal goiters are not easily imaged by ultrasound due to artefact generated by bony structures and needle biopsies are difficult to perform in routine practice. Value of CT is not only to confirm the presence of retrosternal goitre but also helps to define the location and extent of lesions into different mediastinal compartments, adjacent organ involvement or vascular involvement.^{4,6} The high iodine content of the thyroid gives it higher attenuation than the surrounding soft tissue, which is detected by CT and makes CT useful in identifying thyroid extension into the mediastinum.

Surgery is primary choice of treatment in which approximately 2% of substernal goitre is removed by extra cervical approach. El hammoui et al⁷ showed that treatment of choice in retrosternal goitre is early recognition and surgical removal by Kochers transverse collar neck incision with very low surgical complications. Sternotomy is more likely in case of growth beyond aortic arch. Casella et al.⁹ found that extension of the goitre below the level of the aortic arch appeared to be a significant predictive factor for the need for sternotomy. Conversely, the lack of radiologic extension beyond the aortic arch predicted successful trans cervical removal of mediastinal goiters without sternotomy.⁹

In such long-standing cases of goitre and lymphocytic thyroiditis, histopathological evaluation is very important to rule out any evidence of malignancy like follicular neoplasm, papillary carcinoma thyroid, or metastasis from any primary lesion. In our case, evidence of long-standing adenomatoid goitre was found on histology supported by the presence of extensive hyalinization, thickened capsule, atrophic thyroid follicles along with foci of chronic lymphocytic thyroiditis.

Massive retrosternal goiter not only increases the difficulty of surgery and postoperative complications but also poses a challenge to perioperative anesthesia management. It may cause a difficult airway, cardiovascular instability or postoperative tracheomalacia.⁴ The anaesthesiologist, thus, should know and be prepared to manage

mediastinal mass syndrome, difficult intubation, massive blood loss, prolonged surgical time, sternotomy concerns and post thyroidectomy complications.^{6,7}

CONCLUSION

Surgery remains the primary choice for retrosternal goitre. In most cases conventional cervical approach is sufficient, but sternotomy may be required in 2% of cases and without hesitation sternotomy should be done if growth extends beyond aortic arch to prevent complications.

Comprehensive preoperative planning including “context-sensitive airway management”, anticipation and preparedness for complications along-with interdisciplinary medical coordination & structured clinical protocols are required for safe management of such patients.

Figures with Legends:



Figure 1: Clinical image showing a diffuse swelling on right side of neck

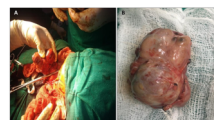


Figure 2A-B: Right hemithyroidectomy specimen.

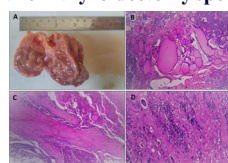


Figure 3A-D: A-Cut section showing grey-brown areas, thick fibrotic areas, cystic areas. A well-defined lobule is noted measuring 4x3.5cm; B: Involutional & hyperplastic thyroid follicles with luminal colloid [H&E-20X]; C: Image showing markedly thickened capsule surrounding the thyroid follicles [H&E- 20X]; D: Section showing extensive areas of hyalinised stroma & compressed thyroid follicles [H&E- 40X]

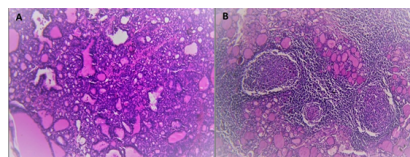


Figure 4A-B: A-Section showing few nodular areas showing extensive hyperplasia with overcrowding & overlapping of nuclei with mild atypia [H&E- 20X]; B: Isthmus showed prominent lymphoid follicles with germinal center [H&E- 20X]

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