RETROSTERNAL GOITRE MASQUERADING AS OBSTRUCTIVE SLEEP APNOEA IN A MIDDLE-AGED FEMALE

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ABSTRACT

PRESENTATION OF CASE

Retrosternal goitre grows in a tight compartment between the sternum anteriorly and vertebra posteriorly and are symptomatic due to compression of airway and great vessels. The presence of RSG is, per se, an indication for surgical management. Surgery can most commonly be performed using the cervical access, but at times, a sternotomy or thoracotomy is necessary. The challenges encountered by the anaesthetist and the surgeon are- difficulty in intubation/ventilation due to compression of airways, hypervascularity and the proximity of the goitre to great vessels. We report a case of a large RSG in a middle-aged obese female who was on CPAP treatment from a physician for the symptoms of obstructive sleep apnoea for 2 years, but with no relief. There was no visible or palpable neck swelling. CECT neck and chest was done in view of worsening of symptoms, which showed heterogenous enhancing mass in superior mediastinum up to carina likely lymph node mass or exophyti thyroid mass after, which she was referred to Surgery Department. Ultrasound findings also suggested colloid goitre of both the lobes and swelling of isthmus, which was continuous with the retrosternal mass. FNAC proved it to be colloid goitre with cystic degeneration. She was euthyroid as per laboratory parameters. Patient was operated by transcervical approach and near total thyroidectomy was performed. Patient was discharged in satisfactory condition. A high index of suspicion for the presence of retrosternal goitre should always be kept in mind in endemic areas of goitre in patient of obstructive sleep apnoea, which is refractory to medical management.

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DIFFERENTIAL DIAGNOSIS

There was difficulty in diagnosis, since the patient had no swelling in the neck and she was being investigated for sleep apnoea because of her obesity.

CLINICAL DIAGNOSIS

Retrosternal goitre was first described by Hallen 1749 and first surgically removed by Klein in 1820.[1] No uniform definition of retrosternal goitre is described in the literature.[1] However, various criteria have been suggested by different authors, thyroid extending 3 cm below sternal notch, extension below thoracic inlet, goitrous mass 50% or more of which is in anterior mediastinum. Incidence of palpable thyroid nodule in general population ranges from 4-7%.[2]

Diagnosis of RSG is mostly made in 5th-6th decade of life with female-to-male ratio of 4:1.[1,3,4] 90% are noted in anterior mediastinum and the remaining having extension into posterior mediastinum.[1] CT is the most important imaging modality as it helps in the complete assessment with regards to the extent of goitre and compression effects on adjacent anatomical structures.

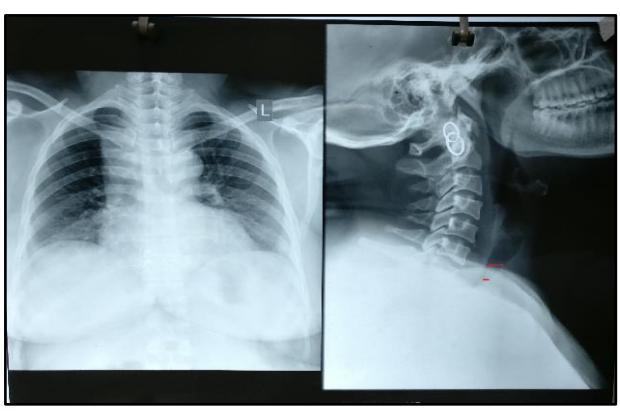

Figure 1. Preoperative CXR Showing Mediastinal Widening and Compressed Trachea Anteroposteriorly

There is a good consensus that most RSG should be removed for the possibility of potential airway compromise and association of thyroid malignancy.[1,5] In most of the cases, the retrosternal goitre maybe removed successfully by transcervical access, however, a sternotomy or lateral thoracotomy maybe required when there is involvement of posterior mediastinum, extension to aortic arch, recurrent RSG, SVC obstruction, malignancy with local involvement, emergency airway obstruction and excessive traction being required for surgery.[6] OSA occurs when there are repeated

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episodes of partial or complete upper airway obstruction during sleep. Described is a patient who was being managed previously on the line of OSA and subsequently diagnosed with RSG. The patient was operated and the goitre was removed via transcervical approach.

PATHOLOGICAL DISCUSSION: Histological examination confirmed colloid adenomatous goitre other space-occupying lesions in the mediastinum, including thymomas, dermoid, cysts, pleuropericardial cysts and neurogenic tumors should be considered in the differential diagnosis

DISCUSSION AND MANAGEMENT

A 45-year-old obese female presented with complaints of shortness of breath for 1.5 months with history of loud snoring, daytime sleepiness and abrupt awakening with sore throat for 2 yrs. Patient had episodes of choking during night, difficulty in lying supine and history of falling asleep while watching television with morning headache and shortness of breath. No history of pedal oedema or postural or diurnal variation of shortness of breath. She was a known case of hypertension and on tablet amlodipine 5 mg for 3 yrs. Patient was diagnosed as a case of obstructive sleep apnoea and was receiving CPAP for the same from a private practitioner. ECG and 2D echo done were normal. Due to worsening of symptoms, patient was referred to Department of Pulmonary Medicine, Government Medical College and Hospital, thyroidectomy was performed through transcervical approach (Figure 5).

At surgery, the upper right and left horn of both lobes were normal. Isthmus and lower part of both lobes were enlarged and going down into anterior mediastinum compressing trachea (Figure 6) anteroposteriorly (size 9*7*3 cm (Figure 3). During surgery, bilateral recurrent laryngeal nerves were identified and preserved. Postoperatively, after extubation, patient developed transient neurapraxia of right vocal cord, which resolved with prednisolone. Patient was discharged on postoperative day 10 in stable condition. Histological examination confirmed colloid adenomatous goitre. On followup, patient was relieved of the compressive symptoms and now could lie down supine without any difficulty.

Figure 2. Postoperative CXR showing Relieved Compression of Trachea

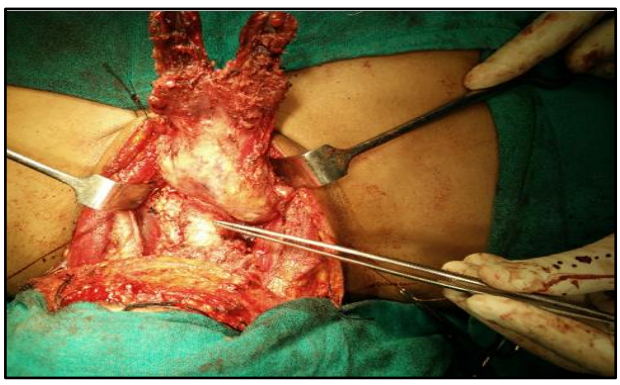

Figure 3. Intraoperative Specimen of the Goitre

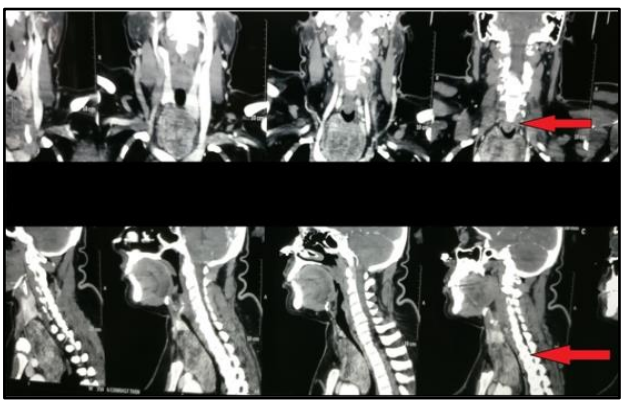

Figure 4. CT Showing Compressed Trachea

Figure 5. Transcervical Incision

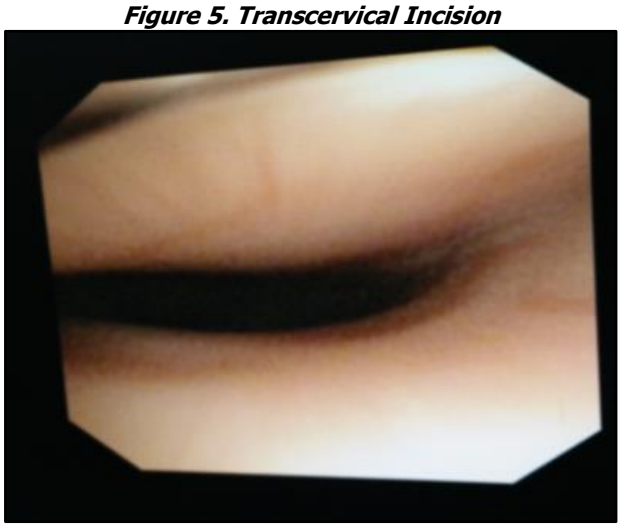

Figure 6. Intraoperative Fiberoptic Bronchoscopy

There is a lack of uniformity in definition of retrosternal goitre. Crile defined mediastinal goitres as those that extend to or are inferior to the aortic arch, whereas Lahey stated

that the greatest diameter of the goitre be inferior to the thoracic inlet on chest x-ray. More precise definitions of retrosternal goitre have been suggested, namely a goitre lying 2 fingerbreadths below the thoracic inlet with the patient in the supine position, a goitre reaching the aortic arch or the carina of trachea, a goitre with its inferior pole passing through the cervicothoracic isthmus below the subclavian vessels.[7]

The term true or aberrant or intrathoracic should be reserved for goitres that neither receive a cervical blood supply nor maintain any connection to the cervical gland. Their blood supply should be from thoracic vessels.[8] As per another classification, goitres maybe primary, arising from embryonic remnants and have no connection with the cervical thyroid, or secondary, when they represent a downward growth of the thyroid gland.[9] By far, the vast majority are of the secondary variety, which derive its blood supply from the thyroid arteries in the neck.[10] In contrast, the primary type has separate blood supply from intrathoracic vessels.

The reason of such enormous dimensions of retrosternal goitre remains unexplained. As described by Lahey and Swinton, due to the anatomical confinement of the thyroid, the path of least resistance, the enlarging thyroid can take it downwards through the thoracic aperture to lodge in the anterior or rarely the posterior mediastinum.[11] This descent is aided by the negative intrathoracic pressure, the action of swallowing, and more importantly, the effect of gravity. Progressive enlargement over the years is due to resilient nature of the lungs and other thoracic contents, which allow the goitre to reach large dimensions before causing pressure symptoms.[12]

Retrosternal goitres are generally seen in the 5th decade of life with a predominance in women.[6] In endemic goitre areas, 20% of the population over 70 will have retrosternal goitre.[1] It is common with a reported incidence of 1-20% of all patients undergoing thyroidectomy.[5] Among the space occupying mediastinal abnormalities, the incidence of retrosternal goitre is 10-15%. The risk of malignancy in RSG is between 3-21%.[5,13]

Mediastinal goitres can remain asymptomatic until compression of structures located in the tight compartment. The most common symptoms are dysphagia, cough and hoarseness of voice. Other modes of presentation include neck mass (75%), hoarseness of voice (37.5%), dysphagia (31.3%) or stridor/wheezing (19%).[14] There is risk of life threatening acute airway obstruction due to sudden haemorrhage within the thyroid gland or secondary to prolonged mechanical pressure with acute laryngeal oedema and congestion.[15] This demands the requirement of emergency tracheostomy in acute presentation in patients of RSG. Horner's syndrome can also be a presenting symptom resulting from cervical sympathetic chain invasion by malignancy, but secondary to sympathetic chain compression with multinodular goitre has been illustrated in the literature.[8] Some patients present with superior vena cava syndrome.[16] The most common symptom complex of obstruction of SVC is oedematous swelling and cyanosis of the head, neck and upper extremities accompanied by dilated collateral veins of the neck, thorax and upper abdomen. The syndrome is more pronounced in acute obstruction, as is the case in malignancy, when venous collaterals have not had time to develop.[17]

In symptom-free patients, the diagnosis is generally established by incidental finding in chest x-ray investigation for some other complaint. The exact location and extent of the goitre can be detected by means of CT. Retrosternal components of goitres are not easily imaged by ultrasound due to artefact generated by bony structures.[15] Other space-occupying lesions in the mediastinum, including thymomas, lymphomas, dermoid cysts, pleuropericardial cysts and neurogenic tumours should be considered in the differential diagnosis.[18]

Literature supports surgical removal of RSG due to the possibility of compression symptoms, potential airway compromise and association of thyroid malignancy. Most thyroid malignancies encountered do not have obvious predisposing features and, therefore, lack of risk factors alone cannot ensure the benign nature of the goitre. Because, there is no effective treatment other than surgery and because thyroidectomy is felt to be both safe and effective, the presence of retrosternal extension is an indication for surgery in an otherwise healthy patient even in the absence of clinical symptoms.[6] Surgical removal of retrosternal goitre can be performed by a cervical approach in majority of patients.[5] Only around 2% of patients undergoing thyroidectomy for RSG will require surgical access other than a standard collar incision (either manubriotomy, sternotomy or thoracotomy).[19] The most important predictive factor as to whether a goitre can safely be removed through a cervical approach is the presence of a clear tissue plane around the nodule in the mediastinum on preoperative imaging. If such a clear plane is not present, preparations should be made for sternotomy.[5]

Considering their hypervascularity and proximity to the great vessels, anticipation of major bleeding is prudent and precautions should be taken. Additionally, due to compression and distortion of the trachea and diminution of the lung fields, difficult intubation and ventilation should also be anticipated. These two points, the difficulty in intubation/ventilation and the proximity of the goitre to great vessels provide real challenges to the anaesthetist and the surgeon.[11] Chances of recurrent laryngeal nerve injury, hypoparathyroidism and tracheomalacia after retrosternal goitre excision always should be kept in mind and for timely management.[15]

FINAL DIAGNOSIS

The possibility of a retrosternal goitre should be kept in mind in patients presenting with respiratory difficulty even without an obvious neck swelling, especially in endemic areas. Mainly, due to respiratory symptoms, patient may be treated initially by the physician on the lines of asthma or obstructive sleep apnoea. Huge retrosternal goitre presents as a challenge to the surgeon and the anaesthetist. Difficult

intubation and ventilation should be anticipated. The recommendation of thyroidectomy even in asymptomatic patients of RSG is based on the risks of airway obstruction and malignancy, the presence of symptoms in the majority of patients on direct questioning and the tendency for the goitre to demonstrate time-dependent progressive growth.

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