

THE RETROSTERNAL (SUBSTERNAL) GOITRE

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Abstract

This is an overview of the various factors related to the substernal (retrosternal) goitre. The presentation and discussion include the terminology of the condition, its definition, type, the mechanism and way the goitre descends in the thorax, its incidence, clinical features, investigations, and possible challenges in anaesthesia and surgical exposures.

Introduction

Thyroid conditions and different types of enlargement of the gland are common events. Surgery remains having an important role in the management of these conditions. With advancement in contemporary peri-operative preparation, anaesthesia and surgery, excellent results with very low morbidity and almost zero mortality could be achieved. On the other hand large substernal goitres (SSG) continue to be challenging due to the distorted anatomy which they cause, the limited or difficult accessibility during their surgery and the potential injury to vascular, neurological, pleura and other structures.

Terminology and definition

Intrathoracic extensions of goitres are also named retrosternal, substernal and mediastinal goitres. There is no accepted agreement as to what is the best single definition of goitres in order to be called substernal. The commonly used definitions describe the following: (a) at least 50% of the thyroid tissue must be located below the sternal notch^{1,2}; (b) goitres extending to the fourth thoracic vertebra³; (c) Wu et al⁴ described three types depending on the clinical features and thyroid position as shown on imaging; type I goitres, when

the lower pole of the gland is over the arch of the aorta; type II goitres are those below the arch of the aorta and extending into the posterior mediastinum, while type III are giant goitres located in the chest or when the patient present with superior vena cava syndrome. (d) Others simply define these goitres as any thyroid enlargement identified below the thoracic inlet at operation, with the patient's neck held in extension⁵.

Classification and mechanism of extension from the neck

Substernal (SSG) goitres can be classified into primary and secondary. Primary SSG also named *aberrant* and represent about 1% of all SSG. It arise from an accessory (ectopic) thyroid parenchyma located in the chest^{1,6}, which do not have any connection to the cervical gland and do not receive a blood supply from the neck⁷.

Secondary SSG constitutes the majority of the cases. These goitres descend from a cervical goitre along the fascial planes of the neck of least resistance, and receive their blood supply from the superior and inferior thyroid arteries^{1,7}.

Extension of a cervical goitre into the mediastinum is attributed to many

factors. These includes: the increasing weight of the thyroid gland, negative intrathoracic pressure, respiratory movement, swallowing and shortness of the neck⁸. In addition it is thought that the absence of anatomical structure that anchors the thyroid between the lower neck and the inlet of the thorax will help migration⁸.

Seventy-five percent to 90% of SSG are located anterior to the recurrent laryngeal nerves and major vessels^{8,9}. However, the nerve may be lying anterior to the goitre, may course between lobules of these goitres, or may not always present posterior to all nodular components^{8,9}. Approximately 10% to 15% of SSG are located in the posterior mediastinum⁸, which tend to descend from the posterolateral aspects of the thyroid gland.

SSG usually descend to the right, because they take the path of least resistance and grow away from the aortic arch and its branches¹⁰. Goitres, which arise from the left lobe, may descend to the right due to the presence of major vessels.

Incidence, presentation and investigation

The incidence of SSG in several large thyroidectomy series varies between 1.7% to 21%^{2,11}. This wide range is due to how the SSG is defined by different centres, the geography of the practice and patients populations involved, and the pattern of referral of patients¹¹.

Although patients in their fifth decade of life are predominantly affected with this condition, patients as young as 15 years and old as 90 years were reported¹. The condition is commonly seen in women with a female to male ratio of 3 or 4:1^{11,12}. Five percent to 40% of patients with SSG are asymptomatic. Again this wide range of incidence is likely attributed to the same previously described reasons. The intrathoracic extension of the goitre in

these patients is discovered either on physical examination or as an incidental finding on a chest x-ray or CT scan performed for some other condition².

The variety of symptoms and signs, which patients with SSG may present with are mainly related to the pressure or compression of the airway, oesophagus, vascular structures, or nerves, separately or in combination^{8,9}. The most common symptoms include cervical mass (65%), dysphagia (36%), and dyspnoea (32%) in addition to cough, stridor, wheezing, and hoarseness^{11,13}. In a series of 175 patients with SSG laryngeal nerve palsy was found in 6% of the cases¹⁴. An abrupt deterioration of respiratory function may develop and usually caused by either respiratory tract infection, sudden haemorrhage in the nodules, or the position of the goitre in the mediastinum, or combination of any of these factors¹⁵. A rare neurologic presentation that may occur is Horner's syndrome (meiosis, anhidrosis, and ptosis)¹⁰. Pemberton in 1921 described a manoeuvre which involves "elevating both arms until they touch the sides of the head", if a substernal goitre is present, after a minute or so, congestion of the face, some cyanosis, and then distress become apparent. Some patients will complain of dyspnoea, stridor or flushing only when their arms are raised above their heads or when the neck is extended. These clinical features result from increased pressure in the thoracic inlet and impaired venous return from the head and neck¹¹. Toxic nodular goitre, which is a common complication of non-toxic multinodular goitre, usually occurs after the age of 50 in patients who have had non-toxic nodular goitre for many years¹⁷. Although no specific incidence of thyrotoxicosis is reported in patients with SSG it is expected to be no

different from that incidence of totally cervical nodular goitre. However, the incidence of cancer in patients with SSG is reported in several studies to be between 2.5% and 21%^{1,18,19}. Similarly this wide range is likely due to the same reasons described before.

Although a chest x-ray, ultrasound scan, and sometimes Magnetic Resonance Imaging (MRI) may be useful in the investigation of SSG, CT scan is the most valuable imaging study. Its importance relies on its ability to assess the extent of the intrathoracic component of the thyroid gland, its relationship to other mediastinal structures, and visualisation of any anatomical distortion such as tracheal deviation and/or compression that will help the plan for both the intubation and operation^{11,19}.

Thyroid function tests are useful to augment the clinical finding in assessing the function of the gland. Pulmonary function studies may indicate the presence and level of airway obstruction and serve as a baseline to measure improvement after thyroidectomy¹¹. A biopsy from the SSG is not indicated and it may cause unnecessary bleeding leading to airway obstruction and respiratory distress.

Anaesthesia and surgical approach

Long standing cervical multinodular goitres responds poorly to thyroid hormone suppression¹¹. In SSG, this line of treatment is even more difficult to monitor and is also expensive because it is impossible assessing the size of the SSG unless a regular CT scan is performed which is both costly and induce unnecessary radiation.

Many believe that mere presence of SSG by itself is an indication for operation²⁰. This is because of the possible abrupt deterioration in symptoms which needs an emergency operation. The other secondary

indication although not high is the possible co-existing malignancy. In addition the real risk for these patients who are not going to have their thyroid removed is unknown⁹.

For fairly asymptomatic patients with SSG surgery may be still offered due to the presence of significant finding by chest x-ray or CT scan⁹. It was also suggested that for a truly asymptomatic patient with SSG it is advisable to perform pulmonary function studies to help in deciding the severity of the physiologic problem because it was found that subjective symptoms may not be a good predictors of how severe the disease is anatomically and physiologically⁹.

Due to the significant distortion of the trachea in cases with SSG the usage of rigid endotracheal tube was recommended¹¹. Implementing the slow, awake intubation with variable degree of sedation was also mentioned⁹. However, Byrd in his discussion to reference⁹ thought that this way of intubation can aggravate obstruction of an already compressed trachea. He also thought that the site of narrowing or tracheal deviation in these patients is below the level of the larynx and therefore, a straightforward induction and intubation using a standard endotracheal tube and stylet is sufficient as the apparently narrow trachea will easily distend and allow passage of the tube. Probably one possible exception to that method is the rare patient with a true angulation of the trachea rather than the more common gentle curving deviation⁹.

All reporters^{2,4,8,9,11,18} agreed that cervical surgical approach is the preferred one. Through this approach a successful thyroidectomy could be achieved in all patients with SSG except in approximately 2% of the cases²¹. Other described surgical exposures are median sternotomy and right lateral thoracotomy. Median

sternotomy should be considered in patients who are known or highly suspicious to have invasive carcinoma, have had previous thyroid operations, those with primary mediastinal goitres with no thyroid tissue in the neck, and those patients who develop marked mediastinal bleeding that is difficult to control through the cervical approach^{22,23}. It was also reported that the presence of posterior mediastinal goitre greatly increases the chance of median sternotomy²⁴.

To make the most benefit of the cervical approach the subplatysmal flaps should be mobilised inferiorly and the strap muscles should be separated down to the sternal notch¹¹. Usually ligation of the middle thyroid vein and securing the upper pole vessels is recommended as an initial step in the dissection. The thyroid lobe is then mobilised medially and if needed the isthmus could be divided. This gives more space to manoeuvre and allows rotation of the cervical portion of the gland from lateral to medial, which provides further mobility and also help gentle, blunt finger dissection on the surface of the capsule along the carotid sheath into the mediastinum through areolar planes around the borders of the goitre. This permits superior retraction and delivery of the substernal part into the neck. If digital mobilisation is unsuccessful, special forceps and spoons can be used to deliver the substernal component of the goitre^{9,22}. It was also thought that because most of the blood supply to the SSG is still maintained through the cervical portion of the gland then by ligating as much blood supply as possible will result in some shrinkage of the substernal part because a substantial volume of the gland is from the presence of blood within the glandular tissue¹¹. Care must be taken not to rupture the capsule because it leads to a troublesome bleeding and

makes the subsequent dissection more difficult and complicated. If it became obvious that the goitre is unable to be delivered through the generous cervical approach then an additional approach, usually a median sternotomy should be undertaken, which could be a T-shaped skin incision caudally to split the sternum²⁵. Piecemeal removal or morselisation is not recommended because it could lead to serious bleeding and the violation of en-block dissection with potential spread of the disease in the presence of malignancy^{11,22}. If the somehow freed goitre was unable to be delivered through the thoracic inlet and became wedged between the bony structures and the trachea then an acute respiratory obstruction might develop. This necessitates an immediate median sternotomy¹¹. Clark et al²² described a technique to control continuous bleeding from the large substernal space occupied previously by the goitre. A Foley catheter with a 30-mL balloon was placed into the right mediastinum and inflated with 18-mL of water. The balloon deflated two hours later but left in situ till the next morning when it was removed after confirming that the bleeding was stopped.

Postoperative complications include recurrent laryngeal nerve injury, airway complications, haematoma, and hypocalcaemia. Other listed, possible complications are pneumothorax, pneumonia, pleural effusion, and cervical plexus neuropathy²⁶. The incidence of these complications is variable between different centres. For recurrent laryngeal nerve injury it ranges between 2.7% to 5.1%, transient hypocalcaemia between 3.4% to 13.3%, and haematoma between 2.7% and 3.4%^{19,27}.

Although there is a wide believe that management of the airway is difficult during the surgery for SSG, Bennett et

al²⁸ reviewed the literature, and identified 1969 patients with SSG, but found what they described a “scant evidence of difficult intubation or postoperative tracheomalacia resulting in tracheal collapse”. Shen et al⁹ reported that 7 (12%) of their patients developed postoperative airway complications, six of them could not be immediately extubated, but were successfully extubated after one to 10 days. They⁹ also found that patients with airway complications were older, had larger goitres, and were more likely to have tracheal compression on preoperative imaging.

Conclusions

Substernal goitres are not as common as the totally cervical goitres, but are important variety of thyroid gland enlargement because they could pose significant challenges in their management. The typical clinical features are due to pressure or compression of important structures in the mediastinum. The best imaging modality is the CT scan. In experienced hands of both anaesthetists and surgeons excellent results with minimal morbidity and no mortality could be achieved.

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