Surgical management of a substernal goiter

Liang-Shun Wang a,b,*

a Division of Thoracic Surgery, Department of Surgery, Taipei Medical University-Shuang Ho Hospital, Taipei, Taiwan
b Graduate Institute of Clinical Medicine, College of Medicine, Taipei Medical University, Taipei, Taiwan

Received 16 October 2011; received in revised form 5 December 2011; accepted 21 December 2011
Available online 8 March 2012

Summary  Substernal goiter can be classified as primary or secondary, depending on the site of origin. Primary substernal goiters (< 1% of substernal goiters), also known as mediastinal aberrant goiters, arise from ectopic thyroid tissue in the mediastinum, and receive their blood supply from intrathoracic arteries instead of thyroid arteries. A secondary substernal goiter is defined as one that has descended from the neck to the plane below the thoracic inlet, or one that has more than 50% of its mass lying inferior to the thoracic inlet. Surgical resection should be considered even for elderly patients because of the risks of mass compression symptoms (e.g., dyspnea and dysphasia), malignancy, and low morbidity of surgery. Most of the primary substernal goiters can be resected through the cervical approach. In most instances, sternotomy or thoracotomy is needed only in cases of previous cervical thyroidectomy, invasive carcinoma, or ectopic goiter.

Copyright © 2012, Taiwan Surgical Association. Published by Elsevier Taiwan LLC. All rights reserved.

1. Introduction

The term goiter has been defined as an enlargement of thyroid gland, while some authors refer to a thyroid gland that is enlarged to twice its normal size or weighs more than about 40 g. Most of surgical goiters reported in the literature were nonendemic, although the classic pathophysiology of goiter development is dietary iodine deficiency, which leads to hypothyroidism with an increase in thyroid-stimulating hormone (TSH), and then causes thyroid gland hypertrophy (so-called endemic goiters). Substernal goiter was first described by Haller in 1749, and Klein was credited with removing the first substernal goiter in 1820. In addition to substernal goiter, there are other synonyms, including ‘retrosternal,’ ‘intrathoracic,’ or ‘mediastinal’ goiters in the literature.

2. Definitions and incidence

The terminology has not yet been standardized in the literature, nor has the proportion of the thyroid gland extending from the neck into the thorax been uniformly
defined. The most commonly accepted definitions describe a substernal goiter as one that has descended below the plane of the thoracic inlet or has more than 50% of its mass lying inferior to the thoracic inlet. Other definitions of substernal goiter consist of the following: the goiter with a major intrathoracic component that requires reaching into the mediastinum for its dissection, or if the goiter lies two fingerbreadths inferior to the thoracic inlet in the supine position, or if the goiter reaches the levels of the aortic arch or T4 on chest radiography. Given these myriad definitions, the incidence of substernal goiters as a percentage of thyroidectomies varies substantially in the literature, i.e., from 2%–19%. Based on a systematic review of surgical managements and its complications, Huin and colleagues defined a new classification system of substernal goiters from Grade 1 to Grade 3 according to their anatomic locations and the surgical approaches (including cervical, manubriectomy, or full sternotomy respectively).

3. Anatomy and clinical features
Substernal goiters can be classified as primary or secondary, depending on their sites of origin. Less than 1% of substernal goiters are thought to be truly primary in nature, also known as mediastinal aberrant goiters, arising from ectopic thyroid tissue in the mediastinum. These aberrant goiters have no connection with the cervical thyroid gland and receive their blood supply from intrathoracic arteries instead of thyroid arteries. Secondary substernal goiters, which develop owing to the downward growth of cervical thyroid tissue, account for most cases. These goiters tend to grow into two mediastinal regions: (a) the anterior mediastinum, anterior to the recurrent laryngeal nerve (RLN) and anterolateral to the trachea, and (b) the posterior mediastinum (10%–15%), descending posterior to the carotid sheath and the RLN, especially the right-side posterior mediastinum (> 90%). Almost all of the secondary substernal goiters, no matter anterior or posterior in location, continue to receive their blood supply from the superior and inferior thyroid arteries.

Generally, with female predominance, substernal goiters tend to grow slowly, and commonly leading to presentation in the fifth and sixth decades of life. In 20%–30% of cases, the goiter is not palpable or is barely palpable in the neck, with most of the tumor bulk in the chest. The most common presenting symptoms are due to mass compression effects, including (a) compression of the airway (most frequent), inducing the symptoms from a mild cough, upper respiratory tract infection, or isolated dyspnea to severe, life-threatening asphyxia, and (b) impinging on other nearby structures (e.g., esophagus and vessels), causing dysphagia, hoarseness, and other neurologic and vascular symptoms. Frequent infections of various degrees were the commonest clinical feature in our previous series. Precipitating factors for acute and severe respiratory distress may include a large substernal goiter with intrathyroid bleeding or upper respiratory infections. Although a few patients with substernal goiters may present with thyrotoxicosis and weight loss, most of them are in euthyroid status.

4. Risks of malignancy
Risk factors for malignancy in substernal goiters may include old age, a family history of thyroid pathology, a history of cervical radiation therapy, recurrent goiter, and the presence of cervical adenopathy, thrombotic material within the lumen of a vein in contact with a substernal goiter, and possible preoperative hoarseness. However, the incidence of cancer in substernal goiters is no higher than the incidence of cancer in cervical goiters; 25 series reported malignancy in 0%–22.6% of substernal goiters in literature review. The most common type of carcinoma was papillary, followed by follicular, medullary, mixed or coincident, and anaplastic. Most of the tumors (> 50%–60%) are microcarcinoma (< 1 cm). However, the inability to rule out malignancy in substernal goiters provides a further rationale for performing total or subtotal thyroidectomy in these cases.

5. Diagnosis
With addition of pertinent clinical manifestations, substernal goiter is a major diagnostic consideration in evaluating mass lesions in the upper mediastinal region. Neck and chest radiography as well as computed tomography (CT) scan and magnetic resonance imaging (MRI) are essential for diagnosis. Typical radiography discloses a mass with tracheal deviation or compression at and below the thoracic inlet, calcification within the tumor, and reflection of the mediastinal pleura below the goiter (Fig. 1A and B). Erbil and colleagues reported that chest radiography might provide the first evidence of a substernal goiter in 77% of patients. CT scanning can further permit detailed evaluation of the intrathoracic extent of the thyroid (for secondary substernal goiter) and displacement of the trachea, esophagus, and regional vessels. MRI may provide a critical tool in the visualization of tissue and local invasion of vascular structures by the mass. Fine-needle aspiration biopsy of substernal goiters for cytologic analysis may be helpful when a large cervical component exists, but this is not usually recommended because it can be dangerous and the material obtained is often inadequate for histologic diagnosis of malignancy. It is accepted that thyroid radionuclide scans are not particularly helpful although more than half of substernal extensions can be detected on scintiscans. Nuclear thyroid imaging may demonstrate thyroid activity in the mediastinum, but the absence of uptake in the mediastinum does not exclude a diagnosis of substernal goiter.

6. Managements for substernal goiter
There is general agreement that medical treatment is ineffective for substernal goiters; thyroxine suppression and iodine-131 ablation are not particularly useful. Moreover, radioiodine therapy may induce acute inflammation and swelling of the gland with the potential for airway obstruction. The treatment of substernal disease is clearly surgical, but there is no consensus on the indications for thyroidectomy, although many authors suggest that...
thyroidectomy should be performed in all patients with substernal goiter who do not have medical comorbidity excluding them from surgery.4,18,19 This recommendation is based on the risks of airway obstruction and of malignancy, the presence of symptoms in the majority of patients on direct questioning, and the tendency for the goiter to demonstrate time-dependent progressive growth. Besides, surgery is associated with low morbidity when performed by experienced thyroid surgeons.18,19 Nevertheless, controversy exists in surgical treatment of asymptomatic substernal goiter.19 A large observational study that included 32,777 thyroidectomies (of which 1153 were on substernal goiter) from multiple centers within the state of New York in the United States has convincingly revealed that substernal thyroidectomy (in comparison with cervical thyroidectomy) was associated not only with an increased risk of complications, such as recurrent laryngeal nerve damage (2.1% vs. 0.6%, respectively), hypoparathyroidism (5.5% vs. 3.5%, respectively) and postoperative bleeding (2.2% and 0.9%, respectively), but also with increased mortality (1.4% vs. 0.1%, respectively).20 However, it is generally agreed that radiologic evidence of significant tracheal narrowing and potential airway obstruction may be an indication for surgery in a clinically asymptomatic patient.

7. Surgical approaches and techniques

Surgical removal of a substernal goiter is a challenging procedure; it can be performed safely via a cervical approach in most cases (> 90%), with a slightly higher complication rate than the average rate for cervical goiter thyroidectomy, especially concerning hypoparathyroidism and post-operative bleeding.4 The most significant criteria for selecting patients requiring sternotomy (< 10%) are CT features, in particular, the presence of an ectopic goiter, the thyroid gland volume, the extent of the goiter to or below the tracheal carina, and those suspected of malignancy.16,21–23

As most substernal goiters are associated with tracheal deviation and compression, induction of general anesthesia in such patients has potential hazards. For patients with compression narrowing of the trachea, we suggest intubation of the alert patient for bronchoscopic confirmation that the endotracheal tube is placed distal to the stenotic area. To avoid traumatic injury of the stenotic trachea, fiberoptic endotracheal intubation during spontaneous respiration should be performed under adequate local anesthesia and sedation.

In our experience, more than 95% of substernal goiters can be resected via a low transverse incision, particularly in those with a cervical thyroid mass (secondary type). The blood supply of secondary substernal goiters almost always comes from the inferior thyroid vessels. After dissection and freeing of the cervical portion of the substernal goiter, the substernal part is easily delivered into the neck with gentle upward traction of the cervical thyroid and concomitant finger dissection, starting from the lateral surface, then to the base of the goiter mass, even if the goiter is so large to reach the subcarinal level, and even in those with previous thyroidectomy (recurrent substernal goiter).4 Besides, we strongly recommend suture obliteration of the substernal dead space after resection of the goiter mass.

The thoracotomy approaches may result in disastrous bleeding due to difficulty in controlling the cervical blood supply in addition to relatively poor identification of the recurrent laryngeal nerves and the possibility of simultaneous removal of the cervical portion of the goiter. On the other hand, thoracotomy or cervicothoracic or cervicomediastinal approaches have been advised when the goiter is mainly intrathoracic with little or no cervical component such as ectopic intrathoracic goiter.

8. Surgical results and comorbidity and mortality

A transverse collar incision is a simple and painless surgical wound, with much lower morbidity and mortality as compared with those after thoracotomy or median sternotomy approaches.4,19–22 The potential postoperative complications include vocal cord paralysis, hypoparathyroidism, and bleeding. Many authors have reported an incidence of complications after thyroidectomy for substernal goiters similar to that after standard thyroidectomy (for cervical goiter), except for postoperative temporary hypoparathyroidism.4,12,13,15,18,19 Intraoperative monitoring of recurrent laryngeal nerve may be helpful in preventing vocal

Figure 1 (A) The representative chest radiograph; and (B) chest CT scan of a giant substernal goiter. A woman 62 years of age was admitted with acute respiratory distress. Chest radiograph and CT scan revealed a giant substernal goiter from the cervical region to the level of tracheal carina with severe deviation and compression of the trachea and superior vena cava vein. Thyroidectomy was smoothly performed through the cervical collar incision. Patient was uneventfully discharged on the fourth postoperative day. CT = computed tomography.
cord paralysis. Such hypoparathyroidism occurs more often in surgery for substernal goiters (5%–10%) than in standard thyroidectomy. Most patients can leave hospital within a few days after surgery. Symptoms such as stridor and dyspnea subside soon after surgery. Despite remarkable tracheal compression at the time of diagnosis, tracheomalacia never occurred after thyroidectomy in our previous experience. Noninvasive positive pressure ventilation may be used in the management of stridor and airway compromise following early extubation in patients with post-thyroidectomy tracheomalacia. Radiographically, all the deviated tracheas can resume normal midline position, and the diameter of the compressed tracheal lumina will increase to almost normal within 2–3 months after surgery.

9. Conclusion

Substernal goiter is diagnosed essentially by chest radiography and CT/MRI scans. Surgery for substernal goiters should always be considered even in elderly patients because of the high risk of tracheal compression and the low morbidity of surgery. Most substernal goiters are benign and can be resected through a cervical approach. Sternotomy or thoracotomy is usually needed only in cases of previous cervical thyroidectomy, invasive carcinoma, or ectopic goiter.

References