Formosan Journal of Surgery (2012) 45, 41—44

Available online at www.sciencedirect.com

SciVerse ScienceDirect

journal homepage: www.e-fjs.com

MINI-REVIEW

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

KEYWORDS
goiter;
intrathoracic;
substernal

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.

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.[1] 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[2] in 1749, and Klein[3] 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

* Taipei Medical University-Shuang Ho Hospital, Number 291, Jhongjheng Road, Jhonghe City, New Taipei 235, Taiwan.
E-mail address: wangls72269@yahoo.com.tw

1682-606X/$ - see front matter
doi:10.1016/j.fjs.2012.02.001

defined.[4] 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.[5–8] 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.[9,10] Given these myriad definitions, the incidence of substernal goiters as a percentage of thyroidectomies varies substantially in the literature, i.e., from 2%–19%.[11] Based on a systematic review of surgical managements and its complications, Huin and colleagues[12] 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.[13] 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%).[14] 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.[6] 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.[4,6] Frequent infections of various degrees were the commonest clinical feature in our previous series.[4] 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.[11] 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).[15] 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[16] 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.[4] 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.[16]

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.[17] 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