European Annals of Otorhinolaryngology, Head and Neck diseases xxx (2015) xxx–xxx

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Original article

Substernal goiter: Experience with 50 cases

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ARTICLE INFO

Keywords:
Goiter
Substernal
Fiberoptic endoscopy
CT
Surgery

ABSTRACT

Introduction: Goiter is localized or generalized thyroid hypertrophy. It is usually cervical, but may show intra-thoracic development beyond the thoracic inlet and down to the mediastinum: i.e., substernal goiter. The purpose of this study was to analyze the epidemiological, clinical and paraclinical profile of substernal goiter and the difficulties of management.
Material and methods: A retrospective study included 50 cases from our ENT department.
Results: Mean age was 47 years, with a sex ratio of 0.06. Clinical symptomatology was dominated by medial anterior cervical swelling. Compression signs were observed in 26 patients, and cervical lymph nodes in 3. Mean time to consultation was 7 years. Fiberoptic endoscopy found vocal cord palsy in 3 patients. Cervical ultrasonography was the first-line diagnostic test. Cervical-thoracic CT scan was requested in 45 patients to study thoracic extension and the relation of the thyroid mass with the supra-aortic vascular axes. Treatment systematically comprised total thyroidectomy on a cervical approach. There were 6 cases of malignancy.
Discussion-conclusion: Substernal goitre is fairly frequent. Despite particularities, an exclusively cervical approach is sufficient in a large majority of cases. The substernal nature of the goitre did not have major impact on postoperative complications.

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1. Introduction

Goiter is localized or generalized thyroid hypertrophy. It is usually cervical, but may show intra-thoracic development beyond the thoracic inlet and down toward the mediastinum: i.e., substernal goiter. Management of substernal goiter is specific, due to the size of the thyroid and the mediastinal location.

The purpose of this study was to analyze the epidemiological, clinical and paraclinical profile of substernal goiter and the difficulties of management.

2. Material and methods

A retrospective study included 50 patients undergoing resection of substernal goiter in our ENT and Head and Neck Surgery Department between January 1, 2010 and December 31, 2013. Inclusion criteria were: inferior end of goiter non-palpable on clinical examination; substernal goiter on ultrasonography; substernal goiter on CT scan; and/or substernal goiter discovered intra-operatively ($n = 5$, 10%). Exclusion criteria were: goiter not confirmed to be substernal on intraoperative observation (false substernal goiter), and exclusively intrathoracic goiter.

Analysis focused on clinical data (age, gender, history and functional and physical signs), imaging, surgical data (approach, operative difficulties, type of resection), follow-up and pathology results.

3. Results

Only 6.5% of goiters operated on in the department were substernal. Mean age was 47 years (range, 22–72 years); the predominant age group was 43–52 years (18 patients, 36%). Sex ratio was 0.06 (47 females, 3 males). Sixteen percent of patients had familial history of goiter.

All patients had first consulted for progressive medial cervical swelling, at a mean 7 years after onset (range, a few months to 25 years).

Clinical symptomatology predominantly comprised compression signs, in 33 patients (66%). Dyspnea was frequent ($n = 23$, 46%); dysphonia affected 13 patients (26%), dysphagia 4 (8%), and superior vena cava syndrome 1 only (Fig. 1).

All patients were euthyroid on admission to surgery; 3 (6%) had shown clinical signs of hyperthyroidism confirmed on biological

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http://dx.doi.org/10.1016/j.anorl.2015.09.007
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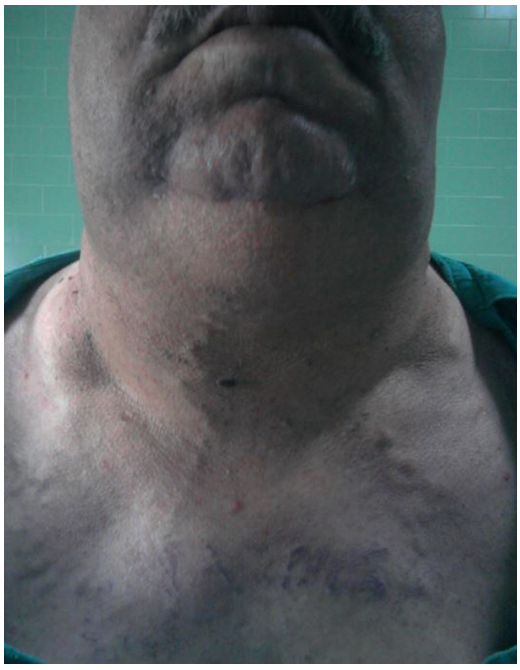

Fig. 1. Frontal clinical examination showing telangiectasias.

analysis, and received synthetic anti-thyroid drugs with or without associated β-blockers.

Goiter could be palpated in the neck in all cases (Fig. 2), and presented a firm consistency in 36 patients. Cervical adenopathies were found on palpation in 3 patients. Systematic fiberoptic endoscopy found paramedial vocal fold palsy in 3 cases (6%).

Systematic chest X-ray found superior mediastinal opacity in 36 patients (72%), tracheal deviation in 44 (88%), and tracheal deviation and compression in 8 (16%).

Cervical ultrasonography was performed in 42 patients (84%) but confirmed the substernal form of the goiter in only 23 cases (46%).

CT was performed in 45 patients (90%) and systematically confirmed the substernal form of the goiter. Patients were classified according to the location of mediastinal extension with respect to the trachea and aortic arch (Table 1).

Fig. 2. Lateral clinical examination: large substernal goiter.

All patients were operated on under general anesthesia by experienced thoracic surgeons. The approach was exclusive anterior cervicotomy. Surgery consisted in total thyroidectomy in 49 cases (98%); left lobo-isthmectomy was performed in 1 case. Retrograde recurrent nerve dissection was performed in all cases (Table 2). No patients required sternotomy.

Histologic analysis found benignity in 88% of cases, 4 papillary carcinomas (2 of which associated with adenopathy at admission), 1 medullary carcinoma, and 1 anaplastic carcinoma (associated with adenopathy at admission). Malignancy was not diagnosed preoperatively due to the difficulty of performing fine-needle aspiration in substernal goiter; this was true even for the medullary carcinoma, which was revealed by histology, with no familial history and hence no biological assessment. Mean substernal goiter weight was 205 grams (maximum, 820 grams); size ranged between 5.3 cm and 19 cm.

There were no postoperative compression hematomas. One case (2%) of transient hypoparathyroidism, revealed by paresthesia of the limbs, was managed by calcium supplementation. There were no cases of permanent hypocalcemia. Two patients (4%) showed postoperative recurrent nerve palsy despite previous normal fiberoptic endoscopy and benign histology.

4. Discussion

There is a strong correlation between the frequencies of cervical goiter and substernal goiter; the latter is therefore very common in regions where goiter is endemic [1,2]. Depending on reports, substernal goiter frequency ranges from 2.5% to 20% of thyroidectomies [3]; in the present series, it was 6.5%. Mean age at onset of cervical goiter is 40 years but, for substernal goiter, is reported to be 56–72 years [1], perhaps due to the slowness of cervical goiter migration [4]; in the present series, it was 47 years. There is clear female predominance in substernal as in cervical goiter: 87% for Rios [5], 86% for Parra-Membrives [6] and, in the present series, 94%.

Anatomic and topographic conditions and goiter volume explain the diversity in discovery of thoracic goiter: from total clinical latency with intra-operative revelation, to discovery on systematic X-ray, signs of mediastinal compression or thyroid dysfunction, or revelation by asphyxia [7]. The most frequent circumstance is evolution of known anterior goiter and onset of signs of compression. Respiratory signs predominate in clinical symptomatology: dyspnea, whether on effort or at rest, is the main functional sign of compression; in the present study, it was found in 46% of patients, comparably to most series.

Nerve compression, especially frequent in posterior goiter, is not restricted to malignant goiter. It may involve the recurrent, sympathetic cervico-thoracic or cervico-brachial or equally the phrenic nerve [8]. In the present series, frequency was second only to tracheal compression (26%).

Esophageal compression is less frequent than tracheal or neural compression, as the esophagus is easily pushed back, but induces dysphagia, in 5–20% of thoracic goiters in the literature [9–11] and 8% in the present series.

The rate of venous compression ranges from 3% to 19% [7,9]. It almost always involves the superior vena cava confluent. Signs comprise increased neck volume and a greater or lesser collateral circulation, depending on the compression site. In the present series, only 1 patient presented superior vena cava syndrome.

Most authors recommend CT on clinical or ultrasound suspicion of substernal goiter [12,13]. CT assesses extension toward the aortic arch [14,15], locates the goiter (anterior, posterior or mixed), and assesses tracheal impact and any stenosis as well as relations with neighboring organs, and the oesophagus in particular [16]. The Mercante classification [17], based on CT slice reconstruction, identified

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Table 1
Patient distribution according to mediastinal extension location with respect to the trachea and aortic arch.

	Grade 1: goiter above aortic arch	Grade 2: goiter reaching aortic arch	Grade 3: goiter below aortic arch
Type A: prevascular goiter	27	7	0
Type B: paratracheal retrovascular goiter	6	3	0
Type C: retrotracheal goiter	2	1	2

Table 2
Summary of study results.

Number of patients	Mean age	Sex ratio	Functional signs			Physical signs	
			Dyspnea	Dysphonia	Dysphagia	Anterior cervical tumefaction	Recurrent nerve palsy
50	47 years	0.06	23 (46%)	13 (26%)	4 (8%)	50 (100%)	3 (6%)

Approach	Histology		Postoperative complications	
Anterior cervicotomy	Benign	Malignant	Transient hypothyroidism	Recurrent nerve palsy
50 (100%)	44 (88%)	6 (12%)	1 (2%)	2 (4%)

risk factors determining the choice of an extra-cervical approach in retrosternal goiter: grade 2 (goiter reaching or extending below the aortic arch craniocaudally), and type C (goiter with posterior extension to the trachea anteroposteriorly). For Burns [18], sternotomy is indicated if the goiter touches or extends beyond the aortic arch and infiltrates the mediastinum on thoracic CT. In the present study, cervico-thoracic CT was performed in 90% of cases, and systematically confirmed the substernal nature of the goiter.

Substernal goiter can usually be removed by cervicotomy [2,4,19,20] and resorting to sternotomy or thoracotomy is exceptional. Even large retrotracheal posterior goiter can in some cases be removed via a cervical approach [21]. The difficulty with substernal goiter is to determine extension so as to be able to use the least invasive and safest surgical approach possible. It is generally agreed that a single cervical approach is sufficient in a large majority of cases [4,22,23]; in the present series, a cervical approach was used in all cases. The first steps are classical: the superior thyroid pedicle is ligated first, to exteriorize the goiter; after ligating the middle thyroid vein, dissection identifies the capsule contact giving access to the cleavage plane, which is followed digitally. Identifying this plane is the key to the procedure [6,19]. The index finger in contact with the capsule traces the outer side of the lobe, around the inferior edge, assessing the arterial pulses and resistances [24], enabling the goiter to be drawn out upward after ligating the inferior thyroid vessels. Dissection of the parathyroid glands is often complex, especially for the inferior parathyroid, which lies against the deep side of the intra-thoracic extension [25], and should be performed in contact with the capsule. It is preferable to leave some fatty periglandular tissue, so as best to conserve the inferior parathyroid and its vascularization. The scale of the thyroidectomy is determined by the pathology, and consists at least of lobectomy and usually of total thyroidectomy. Recurrence is not rare after partial thyroidectomy, studies reporting rates of 15% to 23% [16]; some authors therefore recommend systematic total thyroidectomy, even if the contralateral lobe appears healthy [4]. Resort to sternotomy is rare and usually unpredictable; most authors give it a limited role [18]. As it is usually unpredictable, however, the surgeon should be prepared to perform sternotomy in case of substernal goiter, having a suitable clamp and sternotome to hand and alerting the thoracic surgery team.

Postoperative mortality usually implicates cardio-respiratory failure [22], or sometimes a hemorrhagic, infectious or thyrotoxic event or pulmonary embolism. Rios et al. [5] reported zero mortality, as in the present series. The most frequent complications are vascular and tracheo-esophageal wounds. Recurrent nerve lesions are seldom recognized before extubation.

5. Conclusion

Substernal goiter is fairly common and positive clinical and radiological diagnosis is straightforward. Surgery is mandatory and should be undertaken before onset of compressive complications, which can be very serious. Isolated cervicotomy is the approach currently recommended by most authors. A regulated and codified procedure is the best means of avoiding neural and endocrine complications.

Disclosure of interest

The authors declare that they have competing interest.

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