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# Patient-tailored management of an asymptomatic massive substernal goiter presenting as brachiocephalic vein occlusion. Report of a case and review of sternotomy indications



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## ABSTRACT

**INTRODUCTION:** Substernal goiters are characterized by the protrusion of at least 50% of the thyroid mass below the level of the thoracic inlet. Still their definition is controversial.

**CASE PRESENTATION:** The case refers to a 44-year-old male who presented to our department due to swelling and a feeling of 'heaviness' of his left upper extremity for the past 6 months. CT scan revealed a massive substernal goiter extending to the great vessels. Intraoperatively, a median sternotomy was performed due to the size of the gland and the close adhesion of the isthmus and lower left thyroid lobe to the brachiocephalic vein. Resection of the gland revealed the vein to have a cord-like shape, leading to reduced venous return and upper extremity symptoms. Recovery was uneventful for the patient who was discharged on the 7th postoperative day.

**DISCUSSION:** While most substernal goiters can be surgically managed through a cervical incision, there are cases in which a median sternotomy is indicated. Those cases include excessive gland size, thoracic pain, ectopic thyroid tissue and the extent of the goiter to the aortic arch. Median sternotomy is associated with a number of intra and postoperative complications, although when performed by an experienced surgeon, mortality and morbidity rates along with long-term recovery are not affected.

**CONCLUSION:** The lack of a uniform definition and variety of indications, lead to a patient-tailored approach regarding the execution of sternotomy during surgical management of massive substernal goiters.

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## 1. Introduction

Substernal goiter refers to the intrathoracic descent of a portion of thyroid mass. Those goiters are usually overly enlarged and their clinical presentation may include symptoms associated with the close proximity of the substernal part of the gland and the surrounding visceral and vascular tissues [1,2]. We present a rare case of a patient with known Grave's disease for the past 20 years due to an enlarged substernal goiter, the lower left lobe of which was constricting the brachiocephalic vein, causing upper extremity heaviness and swelling.

## 2. Case presentation

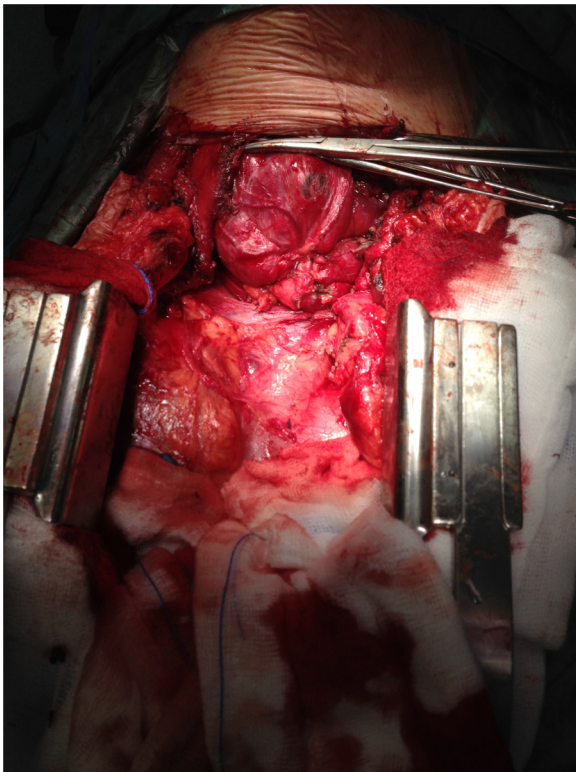
The case pertains to a 44-year-old Caucasian male, who was admitted to our department regarding a mild swelling of his left upper extremity, along with a feeling of 'heaviness' for the past 6 months. Patient exhibited these symptoms during the day, while they were subsiding after night rest. His previous medical history was free. Physical examination revealed a large multilobular goiter, possibly extending below the level of manubrium, while his left arm had no visible findings. Laboratory exams were within normal limits. A CT examination and U/S scan of his cervical region were performed, revealing a massive substernal goiter with its isthmus and left lobe extending in the anterior mediastinum. Maximum thyroid diameter was approximately 15 cm. The trachea was severely stenosed and dispositioned to the right side (Fig. 1). After written consent, patient was admitted for surgery.

Under general endotracheal anesthesia, a cervical Kocher incision was performed. After dissection, the upper anterior part of

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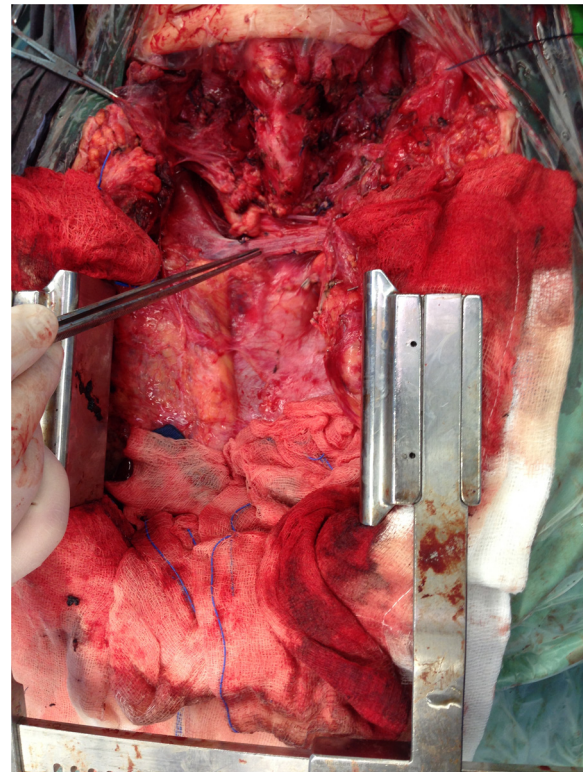


**Fig. 1.** CT scan demonstrating severe stenosis and disposition of the trachea.



**Fig. 2.** Partly retracted goiter revealing a fibrous, cord-like brachiocephalic vein.

the enlarged thyroid gland was exposed. Manipulations were very strenuous due to the size of the thyroid mass, while the isthmus and lower left lobe of the gland were strictly adhered to the brachiocephalic vein, leading to the execution of a midline sternotomy, for complete exposure of the thyroid gland and the great vessels. After manipulation, the isthmus and left lower lobe of the substernal goiter were carefully dissected from a fibrous and cord-like brachiocephalic vein (Figs. 2 and 3). Both recurrent laryngeal nerves



**Fig. 3.** Complete resection of the goiter showing the hard compressed brachiocephalic vein, responsible for upper left extremity symptoms.

and two parathyroid glands were located and preserved. Total excision of the enlarged goiter was performed, along with a nodule of thymus gland. Pathology report was significant for a 325gr multi-lobular goiter, with a maximum diameter of 15.3 cm (Fig. 4). After surgery patient was taken to the ICU for surveillance.

Post-surgical period was uneventful. Patient was taken to the ward on the 2nd postoperative day, while he was discharged on



**Fig. 4.** Excised substernal goiter, weighting 325 gr. Below is a nodule of excised thymus gland.



**Fig. 5.** 6 month follow up demonstrating good closure of the sternal and cervical wounds.

the 7th. At 6 month follow up, he was doing fine, while he reported no further symptoms from his upper left extremity (Fig. 5).

### 3. Discussion

Substernal goiters are generally defined those where more than 50% of the thyroid volume is distally protruding through the thoracic inlet [3]. Most substernal goiters are located in the anterior mediastinum in approximately 90% of cases [4]. The diversity in the definition of substernal goiters given by various centers causes their reported rates to vary greatly between 5% and 22% [5–8]. In our center we refer to a goiter as substernal when part of the thyroid mass is descending towards the mediastinum and below the level of the thoracic inlet, independently of the thyroid volume included. We prefer to use the term ‘descending goiter’. In a retrospective analysis of unpublished raw data from our center during the past 5 years, approximately 15% of surgically removed goiters were referred to as ‘descending’ or substernal.

Substernal goiters may account for a number of symptoms apart from thyroid disease related ones, depending on the intrathoracic extent of the thyroid mass and its relation to visceral or vascular tissues. The most commonly encountered symptoms include dyspnea, cough and dysphagia, due to the disposition and stenosis of the trachea or the esophagus. Patients may also exhibit dysphonia due to recurrent laryngeal nerve compression, or superior vena cava syndrome [7–9]. In most cases, development of symptoms is slow and progressive, following enlargement of the goiter, hence the late identification of the mass when its size is overly extended [10].

A number of imaging modalities can identify the presence of a large substernal goiter. Chest radiograph may be significant for trachea displacement. Usually, large goiters extend to one side of the mediastinum, causing a lateral displacement or stenosis of the trachea on the contralateral side (7). CT scan is invaluable for preoperative evaluation of the location and extent of the enlarged thyroid gland inside the thoracic cavity, and its anatomic relations to the surrounding tissues [11].

Management of large substernal goiters usually involves their surgical resection, with malignancy being reported in approximately 15% of cases [9]. Most substernal goiter resections can be managed through a cervical Kocher incision, still in certain cases performing a thoracotomy may be required. In two recent large studies, Coskun et al., [7] and Nankee et al., [8] reported the percentage of median sternotomies performed during resection of substernal goiters, while they also suggested indications regarding thoracotomy during substernal goiter resection. Their median sternotomy rates were recorded at 9.5% and 5.5% respectively, while both authors indicated that patients with chest pressure, goiter extension to the aortic arch and ectopic thyroid tissue in the mediastinum may require a median sternotomy [7,8]. In another study, Sand et al., [12], suggested progressive enlargement of the substernal thyroid mass as an indication for the application of thoracotomy to achieve effective resection of the pathologic gland. He also reported that the left lobe of substernal goiters was more likely (70%) to descend inside the thoracic cavity than the right one, such as in our case.

Execution of a thoracotomy is associated with increased blood loss intraoperatively, while postoperatively it accounts for incidents of transient hypocalcemia and recurrent laryngeal nerve paralysis, and longer hospital stay [7,8]. Despite those adverse effects, thoracotomy is considered a safe procedure when performed by experienced surgeons, demonstrating no difference in mortality and morbidity rates and having very good long-term postoperative results [12,13].

In our case, the rare presentation of the massive gland with extremity swelling and heaviness reported by the patient, pointed us towards surgical resection of the substernal goiter. The decision to perform a median sternotomy was taken intraoperatively, due to the size and strict adhesion of the isthmus and left lower lobe of

the thyroid gland to the brachiocephalic vein, causing compression and a fibrous, cord-like 'reshape' of the vein wall, responsible for compromised venous return and, subsequently, upper extremity symptoms.

#### 4. Conclusion

In conclusion, surgical management of substernal goiters is usually performed through a cervical incision, although in certain cases a midline sternotomy may also be required. In our experience, the lack of a uniform definition and the diversity of indications regarding the execution of a median sternotomy for their resection, lead to a patient tailored surgical approach. Still, despite the intraoperative adverse effects and its possible postoperative complications, thoracotomy is considered a safe option in the hands of an experienced surgeon.

#### Conflict of interest

All authors declare that they have no conflict of interest.

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#### Ethical approval

None.

#### Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Author contribution

GS wrote the manuscript, assisted in the operation, reviewed the literature, while he also critically revised the submitted manuscript. EC, KK, SS and SD assisted on the operation, and reviewed the literature. GD performed the operation, while he also supervised the manuscript preparation process. All authors read and approved the final version of the submitted manuscript.

#### Guarantor

Georgios Sahsamani and Georgios Dimitrakopoulos.

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The work has been reported in line with the CARE criteria. J.J. Gagnier, G. Kienle, D.G. Altman, D. Moher, H. Sox, D. Riley, et al. The CARE guidelines: consensus-based clinical case report guideline development. *J. Clin. Epidemiol.* 2014, 67 (1): 46–51.

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