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Mini-Review Article

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Challenges in the Management of Secondary Substernal Goiters: A Mini-Review

(Running title: secondary substernal goiters review)

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Abstract

The secondary substernal goiter (SSG), originating from the cervical thyroid gland, may cause significant compression effects on neighboring vital structures, in particular compromising the safety of airways. The diagnosis should be clinically oriented and helped by imaging studies. There are still controversies and issues of debate, such as the precise definition, diagnosis, recommendation of indication for surgery, management of specific airway problems at intubation / extubation, or the risk of transformation to malignancy, and the comparison of results. Multidisciplinary approach of SSG in a high-volume center is required to achieve the most optimal outcome. The conservative medical management is reported inadequate. Some controversies surround the rationale of surgery in asymptomatic cases, but this view is recently weakened. Surgical mass removal in all symptomatic and most of the asymptomatic cases, performed electively and via a cervicotomy, is considered as the preferred treatment approach for SSG. Specific anaesthesiologic techniques and use of relative adjuncts to secure and maintain the airways, either in elective or emergent surgery, may be needed. Only rarely are an accessory extracervical approach or an emergency tracheostomy required. In most cases, particularly the elective that are managed by a skilled team, postoperative complications are reported essentially similar to those of standard excisional thyroid surgery. In the vast majority of cases, compressive symptoms attributable to the disease disappear soon after surgery. Conclusively, the surgical removal of the SSG is a challenge but is strongly advocated for patients before or at the first signs of tracheal compression, even if it might involve a higher risk of complications compared to cervical goiters. Herein, the relevant basic knowledge and research has been briefly reviewed.

Keywords: *secondary substernal goiter, retrosternal goiter, mediastinal goiter, diagnosis, thyroidectomy.*

1. Introduction

The secondary substernal goiter (SSG) is the excessively enlarging thyroid gland that originates from the cervical component of the thyroid and progressively develops downward into the mediastinum [1-3]. Variability in proposed definitions in the literature results in problematic distribution of incidence rates (2-19 % of all thyroidectomies), and makes it difficult to recommend indications for surgery and compare results [1,4]. The accumulative incidence of retrosternal goiter extension was 6.28 % in a large systematic review [2] comprising of 22125 thyroid goiter patients. An accepted more precise definition is that, SSG is a goiter with extension below the thoracic inlet with the patient in a soupine position or with hyperextended neck (at computed tomography - CT) [2].

Making the correct timely diagnosis, persuading the reluctant or scared patient to accept the treatment, particularly in endemic undeveloped countries, following a meticulous surgical technique that is always challenging, and managing specific airway problems at intubation and extubation are still principle issues of debate and may raise concerns for the caring physicians [5].

This is a mini-review of the pathophysiology, clinical presentation, diagnosis, treatment options, and perioperative management of the SSGs. A comprehensive search of the electronic databases (PubMed / MEDLINE, Google Scholar) regarding recent progress was accordingly conducted, as was an attempt to address current inquiries.

2. Discussion

2.1. Pathophysiology

The negative intrathoracic pressure, in interactive collaboration with the respiratory movements and the traction muscle forces during swallowing, is predominantly facilitating the continuous downward migration of the insidiously enlarging cervical thyroid gland into the mediastinum [2,6]. With these mechanisms, the goiter swelling impacts on the surrounding tissues, particularly at the level of the thoracic inlet, and, consequently, causes mechanical compression on vital structures of the neck and the thorax, such as the trachea, great vessels (superior vena cava - SVC, carotid artery), esophagus and the laryngeal nerves (specifically, the recurrent - RLN) or the sympathetic chain [4,7,8].

An important aspect to consider, that is also a controversial one, is the potential risk of malignancy of the long - standing SSGs [6].

Based on the Torre ‘s et al [9] analysis, the longer clinical history of a thyroid mass is associated with a higher risk of intramediastinal development and neoplastic transformation. In the literature, the vast majority of surgically treated SSGs are benign (struma), and the malignancy rates vary widely among

reports (3-22 %) [3,4,6]. In a multicenter study [10] comprising of 19662 patients who had undergone total thyroidectomy (TT), malignancy was significantly more frequent in the SSG group, treated either with cervical approach (22.4 %) or manubriotomy (36.2 %), than in the cervical goiter (CG) group (10.4 %).

2.2. Clinical picture and diagnosis

SSGs are initially asymptomatic and impalpable, and may be incidentally discovered [6]. When symptoms occur, gradual respiratory distress and other airway problems predominate and can be exarcerbated and even life-threatening [6,7,11,12]. Relatively, the tracheomalacia condition, usually due to the presence of a large compressing SSG , may cause severe pre - , intra -, or post - operative airway obstruction problems , such as dyspnea , cough , expiratory stridor or wheezing ; the systematic review by Huins et al [2] described an incidence of 1 % , but this was reported to be as high as 10 % in SSGs reaching up to the aortic arch or existing for more than 5 years [1,5,12,13]. Other common symptoms or findings include the dysphagia or bleeding from esophageal varices, voice changes caused by compression of laryngeal nerves, the SVC syndrome, and, very rarely, cerebral edema [5,12]. As of functional aspect, the majority of SSGs are euthyroid [3].

Not all SSG cases are presenting with a palpable neck mass, and the diagnosis may be suspected on incidental chest x-ray in up to 20 – 40 % of them [4,8,14]. The Pemperton's manoeuvre can occasionally contribute to palpate a deep goiter component, as well as to identify some latent cases [15-17]. The widely used thyroid ultrasound is not generally combined with fine needle aspiration/ biopsy of the swelling component, since its puncture is technically difficult and may have severe complications such as endothyroidal or endothoracic arterial bleeding and pneumothorax [14,15,18]. CT scan of neck and upper chest is the most useful imaging method for the mass extent and its compression effects on the neighboring vital structures, as well as for evaluation of possible difficulties in tracheal intubation and planning of surgery (Fig. 1); among cases with CT - measured tracheal lumen reduction of < 15 mm, those with < 5 mm are considered as critically obstructed [3, 17]. Further work - up includes thyroid function tests, baseline serum calcium levels, pulmonary function evaluation, and laryngological / bronchological assessment [4,16]. Spirometry is usually indicative of upper airway obstruction, even in asymptomatic SSG patients [15,16].

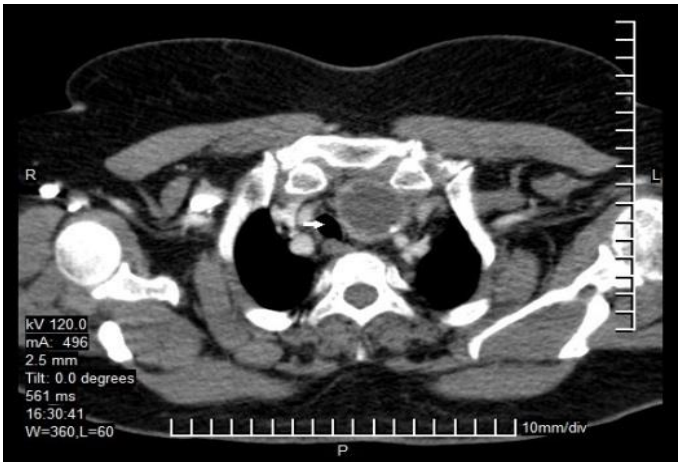

Figure 1: Neck-upper mediastinal CT scan: axial image depicting SSG descending into the mediastinum in a 52-year-old female, compressing and dislocating the trachea (white arrow). The patient had a high preoperative thyroglobulin, but histologically no malignancy was documented.

2.3. Medical treatment and indications for surgery

Medical management of large SSGs with thyroxine supplementation or radioactive iodine (^{131}I) ablation or combination of the two (thyroid hormone is less effective than ^{131}I) is a treatment option for poor surgical candidates, as it is associated with increased risk for severe adverse side effects and radiation - induced malignancies, very rarely being sufficient [14]. The recently employed tracheobroncheal stenting can prevent tracheal obstruction in some cases and delay definite treatment [1,5].

The surgical removal of the SSG mass , usually , a TT via a cervicotomy, in all symptomatic and most of the asymptomatic cases is considered as the gold standard treatment option , predominantly because: (i) progressive compression of neighboring vital structures is strictly associated with the gradual diving of the gland , leading even to acute airway obstruction ; (ii) ~40 % of asymptomatic cases have some degree of airway obstruction on pulmonary functional evaluations , and more SSG cases have hidden malignancy than CG cases ; and (iii) therapeutic ^{131} I is not effective and may be complicated with respiratory distress [10,14,15]. Occasionally, cosmesis can also be an indication for surgery [14].

2.4. Perioperative airway management

For elective surgery, a preliminary preparation of the patient to be euthyroid is essential. The multidisciplinary approach, including experienced professionals in a specialized center, is crucial to overcome relative issues of controversy, offer the appropriate preparation to each surgical candidate, particularly to those who are anticipated to face difficulties in intubation or in maintaining of patent airways or are hyperfunctional, and to proceed to the mass removal achieving the most optimal results [17]. Improved anaesthetical and surgical practices in modern thyroid surgery, and the better prediction and targeted management of complications, need to be widely adopted in order to have low mortality rates (0.16 %) [16,19]. Particularly, the majority of SSG cases with tracheal compression undergoing surgery can be managed with conventional intravenous induction of anaesthesia under muscle relaxant and performing direct laryngoscopy [17]. However, airway compromise and difficult intubation (even when it is preoperatively expected) in cases with large SSGs causing tracheal deviation (2-14 .3 %) [11,13] is often extremely challenging, even for the most experienced anaesthesiologist. There are no specific or precise guidelines from the Difficult Airway Society (United Kingdom) or the American Society of Anaesthesiologists (ASA) for the management of each obstructed airway case [16,20].

Acute complete airway occlusion in cases with large SSGs requiring emergency techniques of securing airways and surgical intervention (life-threatening TT) is caused by intrathyroidal haemorrhage, tracheal compression, or luminal invasion (tumoral) and RLN infiltration, and can occur suddenly, during induction of anaesthesia or surgical resection, or at the time of extubation [16]. Notably, an emergent tracheostomy is technically not allowed by the in situ presence of the thyroidal mass which impedes the access to the trachea, hiding all landmarks; emergent tracheostomy is performed rarely (<1 % in SSGs), mostly for dynamic airway collapse following extubation on postthyroidectomy tracheomalacia (PTTM) [17,21]. Besides, an inhalation induction followed by laryngoscopy and orotracheal or blind nasal intubation, may be

considered dangerous because of complete obstruction following loss of consciousness [21].

The emergency healthcare ABC management plan to secure the airway and ensure an adequate ventilation and oxygenation in the "difficult airway algorithm" of the ASA includes four choices : (i) securing the airway before or after induction of anaesthesia using small - caliber endotracheal tubes ("awake fiberoptic intubation/ AFOI" and "asleep" techniques) ; (ii) maintaining " spontaneous or ablate spontaneous" ventilation ; (iii) using "invasive or non-invasive '' airways ; and (iv) using "conventional direct or indirect" laryngoscopy [16,17,21]. Accordingly, awake techniques can be unpleasant and distressing and their success depends on patient's tolerance, cooperation and operator's (anaesthesiologist) skills [16,17]. Very rarely, airway management techniques include advanced methods such as the cardiopulmonary bypass (CPB) or extracorporeal membrane oxygenation (ECMO) [5,16]. Surgical removal of the mass is the final goal [21].

2.5. Surgical treatment

TT, or near - total, subtotal or ipsilateral "relieve" thyroidectomy represent the treatment of choice for acute airway obstruction caused by compression from the thyroidal mass [4,18,21]. Open cervicotomy remains the most commonly performed surgical approach for the mass removal (Fig. 2), mostly being a TT.

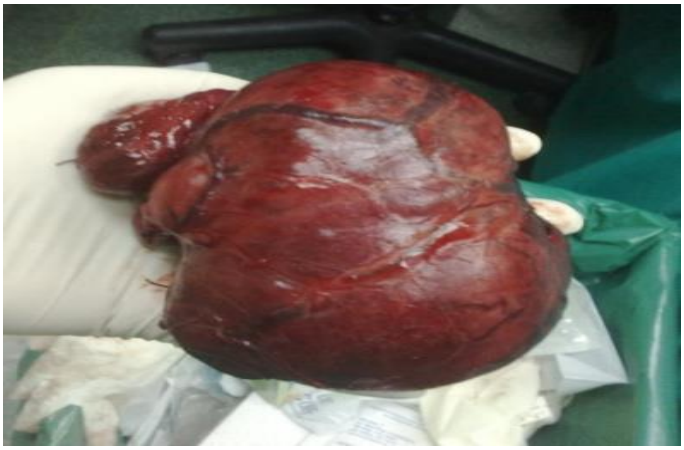

Figure 2: Intraoperative view: extracted mediastinal component of SSG in a 62-year-old female.

The meticulous surgical technique is based on capsular gland dissection in the avascular plane and gentle handling/ preservation of all vital neighboring structures, with particular care given also for haemostasis and management of the residual post-thyroidectomy space (introduction of soft suction drain), as well as for possible problems at extubation ("hidden" tracheomalacia / PTTM?) [3,16,17]. An open combined extracervical (substernal) approach, being definitely decided on the surgical table, may be added in some cases (~2 %) with masses: (i) larger than the thoracic inlet; (ii) suspected for invasive malignancy and associated with lymphadenopathy; and (iii) located posteriorly or having anomalous blood supply [4]. Accessory access could be a sternotomy or an anterolateral thoracotomy. Recently, proposed minimally invasive techniques, such as the endoscopic and robotic, technically demanding, have provided encouraging results [6, 22]. In the vast majority of patients, obstructive airway symptoms, dysphagia, and even neurologic symptoms, attributable to the space - occupying disease, generally disappear completely after surgical removal [14,21].

2.6. Complications

Both intraoperative and postoperative complications of SSG removal can be serious, but are essentially similar to those of standard thyroid surgery, particularly if the procedure was performed in a high-volume center on an elective basis, and a cervicotomic access was used [14,23,24]. However, some investigators reported that, complications and morbidity after TT are higher in SSG cases than in CG cases [10,19]. In most series, mortality is practically zero [15, 17,21].

Acute respiratory distress immediately after extubation means either vocal cord(s) paralysis due to damage of the RLN(s), or tracheal collapse / obstruction due to PTTM (<1 %), or surgical field bleeding / haematoma, and airway edema [14,16]. In most of these cases, emergent re - intubation is the preferred intervention, followed by fiberoptic laryngoscopic evaluation after 24-48 hours, and occasionally a tracheostomy [8,17]. RLN palsy has been reported to be permanent in up to 2.1 % of cases, and transient in up to 5.6 % [8,16,19]. Severe postoperative bleeding (<1 %) [3] requires emergent re-operation to suture the spurting vessel and evacuate blood of extravasation. Hypoparathyroidism, either transient (up to 33 % of cases) [8,19] or permanent (< 5 %) [3,19,21], requires early detection and treatment: I.V. calcium glyconate in acute symptomatic cases, oral calcium and vitamin D in chronic disease. Finally, surgical site infections are reported in up to 3 % of cases, have consequences that can be costly and tolerated badly by the patient, and require close daily care with clean dressings [25]. Substernal thyroidectomy, either via a sternotomy or thoracotomy, is associated with markedly increased morbidity, mostly attributable to the surgical site itself (i.e., wound infection / dehiscence, pain) [23,25].

3. Conclusion

A multidisciplinary team approach allows safe management of the SSG, either compressive or not. The elective surgical SSG mass removal is challenging for anaesthesiologists and surgeons, and is strongly advocated for all symptomatic cases, preferably, at the first sign of tracheal compression, as well as most of the asymptomatic cases, even if it might involve a higher risk of complications compared to the removal of cervical goiters. In the life-threatening case of patient with acute respiratory distress, emergency specific anaesthesiologic techniques for securing the airways and emergency thyroidectomy in a high-volume center are warranted.

Abbreviations

ABC: Airway, breathing, circulation

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Conflict of interest

The author has no conflict of interest to declare.

Ethical Approval/ Informed Consent

Written informed consents were obtained from the patients whose images are used.

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