Management of retrosternal goitres: results of early surgical intervention to prevent airway morbidity, and a review of the english literature

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Abstract

We present our experience and also review the world literature on the management of retrosternal goitres (RSGs). There is now irrefutable evidence that almost all RSGs will continue to grow and eventually cause airway compression. We describe the diagnosis, investigation and surgical approach to the management of this condition.

Key words: Goitre, Substernal; Airway Obstruction; Thyroidectomy

Method

We reviewed the medical records of our most recent 19 patients treated for retrosternal goitre (RSG). A thorough search of the english literature on retrosternal goitres was performed on the Medline database using the key words 'substernal goitre', 'retrosternal goitre', 'surgical technique' and 'airway obstruction'. All relevant articles published since 1934 were retrieved and were read by all the authors. In this paper, we review the management of RSG described in the english literature and also present our clinical experience in the management of this condition.

Introduction

Goitre is defined as enlargement of the thyroid gland to twice normal size. If the greater part[1] (over 50 per cent)[2] of a goitre's mass occupies the area below the sternal notch, it is termed retrosternal. Retrosternal goitre is classified as either primary (i.e. truly intrathoracic), arising from aberrant thyroid tissue within the mediastinum, or secondary (i.e. acquired retrosternal), which represents downward growth from a normally located thyroid gland.[3]

The vast majority of RSGs are benign, with malignancy affecting only a small proportion, ranging from 3–16 per cent,[4,5] depending on the series.

An enlarging RSG can cause a variety of symptoms, including respiratory distress, dysphagia and vascular compression, and may even present as a life-threatening emergency if there is a sudden enlargement of the goitre secondary to haemorrhage, cystic degeneration or malignant change. The larynx and trachea are displaced and narrowed by the enlarged mass, which makes either endotracheal intubation or emergency tracheostomy very difficult procedures. It has been estimated that 1–3 per cent of patients with untreated RSG die of respiratory obstruction.[4] Usually, however, RSG in the early stages remains asymptomatic.

The presence of an RSG in a patient is, in general, an indication for thyroidectomy, even in the absence of symptoms, unless the patient is unfit for or unwilling to undergo surgery. A review of the literature failed to find any convincing results for either suppression therapy or treatment with radioactive iodine.[1,4,6–8] Katlic *et al.*[1] pointed out that longstanding multinodular goitres generally respond poorly to thyroid suppression; in their experience, symptoms and signs remained the same or even progressed despite documented suppression of TSH by the administration of levothyroxine. Similarly, Shai *et al.*[9] found little evidence that radiotherapy, radioactive iodine, propylthiouracil and levothyroxine were effective in relieving mechanical symptoms.

Lahey and Swinton[10] reported that a delay in diagnosis or treatment changed the grading of the retrosternal mass. They divided their cases into two grades: grade I, those extending nearly to the arch of the aorta; and grade II, those extending to the arch of the aorta or beyond. Their data showed that the average duration of symptoms for grade I goitres was 13.04 years and that for grade II goitres was 15.10 years. They also reported that only

6.8 per cent of patients with a grade I goitre had severe obstructive symptoms, as compared with 23.5 per cent for grade II goitres.

The majority of RSGs can be removed via a cervical incision. Primary intrathoracic goitres, recurrent goitres and those with malignancy will need either division of the manubrium or full median sternotomy to enable complete excision.[2] It may not be possible to remove the more unusual posterior mediastinal goitre through a cervical incision, in which case a postero-lateral thoracotomy through the third or fourth intercostal space will facilitate delivery of the mass.

Diagnosis

Symptomatic presentation

Retrosternal goitres grow very slowly and most commonly present in patients in their fifth decade.[1] However, there can be a wide age range of presentation: 15–80 years in one study;[11] 22–90 years[12] in another. In our series, the age of presentation varied from 51 to 95 years (Table I) and there was a female preponderance with a ratio of three to one.

There may be a familial history of goitre; in one series,[13] this was noted in 30 per cent of patients. Whilst some patients with RSG will present themselves with a palpable neck mass as their main symptom, the most common presentation of RSG is with respiratory symptoms of varying degree;[14] these may range from a simple cough or hoarseness to significant shortness of breath or stridor. In our series, 63 per cent of patients presented with respiratory symptoms. Less frequently, acute and severe airway compression may present as an emergency. Aside from respiratory symptoms, RSG may present with a range of other features (Table II).

Some patients with RSG, detected perhaps incidentally on radiological examination, will remain apparently asymptomatic, but direct questioning will frequently elicit often quite subtle symptoms.[15] For example, tracheal narrowing progresses slowly and the patient becomes progressively adjusted to the diminished airway and associated stridor; the noisy breathing is therefore often not immediately appreciated.

Physical examination

Clinical examination of the patient with RSG may be unrewarding, and the goitre may be impalpable. However, if the lower pole of a cervical goitre is not palpable, RSG should be suspected. In our experience, the majority of RSG patients had short necks, and it was not at all easy to palpate the goitre cervically. Occasionally, in patients with a small RSG, hyperextending the neck whilst swallowing and raising the arm may make the substernal component palpable in the neck. During this manoeuvre, there is sometimes flushing of the skin, dilation of the external jugular veins or evidence of airway compression, a positive 'Pemberton sign'. Other findings may include tracheal deviation, prominence of the external jugular veins and, occasionally, Horner's syndrome.

Laboratory and radiological investigations

Thyroid function tests should be performed to document thyroid status. The majority of RSG patients will be euthyroid, although a study by Michael and Bradpiece[17] reported that 44 per cent of their RSG patients were hyperthyroid.

A plain chest X-ray is not particularly sensitive in demonstrating RSG,[17] and other radiological assessments are more useful. A computed tomography (CT) scan will demonstrate not only the thyroid origin but also the relationship of the RSG to the trachea, oesophagus, great vessels and mediastinum (Figure 1). Nodal involvement may also be evident in malignant thyroid disease. Delineation of a clear plane between the RSG and surrounding tissues may be helpful in anticipating intra-operative dissection. Cervical ultrasound scanning is not particularly effective in the assessment of goitres in the substernal position. Michael and Bradpiece[17] compared the sensitivity of a variety of pre-operative radiological tests; chest X-rays had a sensitivity of 59 per cent, CT scans a sensitivity of 100 per cent, and ultrasound scans of the thyroid 77%.

Other investigations, including contrast swallow, venography and digital subtraction angiography,[1] have been reported as possible modalities but are not generally necessary.

Radionuclide scans can be misleading and are not generally helpful.

Fine needle biopsy, because of the relative inaccessibility of the RSG, can be very difficult and potentially dangerous.

Surgical management

Indications for surgery

We present our experience (Table I) and that available from a review of the english literature(Table III).

Intubation

In our experience, standard endotracheal intubation by the anaesthetists was carried out by direct visualization of the larynx using a fibre-optic laryngoscope. An endotracheal tube reinforced with steel wire was used to prevent tracheal collapse. Although there may be tracheal compression from the goitre, in almost all cases, passing the endotracheal tube through this narrow area was uneventful. We did not encounter tracheomalacia giving rise to tracheal compression and airway obstruction in the post-operative period.

Surgical technique

The authors (an otolaryngologist and thoracic surgeon working jointly) carried out all RSG resections as elective procedures.

In our experience of patients with RSG, we consider the following as criteria for partial or complete sternotomy: RSG reaching below the aortic arch; RSG the greatest diameter of which was below the thoracic inlet; CT evidence of possible adherence of the RSG to the surrounding tissues, or distortion of

TABLE I

DETAILS OF RSG PATIENTS SELECTED FOR SURGICAL MANAGEMENT

No	Age (years)	Sex	Main symptoms	Other symptoms	Histology	Sternotomy	Vocal fold function	
							Pre-op	Post-op
1	64	F	Shortness of breath, late-onset asthma	Hoarseness & stridor for few months	Colloid hyperplasia	No	Normal	Normal
2	70	F	Dysphagia, dyspnoea for few months	Hearing loss (RSG for 30 years)	Nodular goitre	No	Normal	Normal
3	85	F	Shortness of breath (worse lying flat)	Change in voice quality, dysphagia	Benign multinodular	No	Normal	Normal
4	73	M	RSG noted by urologist, present for many years.	Episodes of choking for few months	Benign multinodular	No	Normal	Normal
5	88	F	Swelling in lower part of neck	Nil	Colloid goitre	No	Normal	Normal
6	80	F	Irritating cough, lump in front of neck	Nil	Multinodular goitre	No	Normal	Normal
7	68	F	Shortness of breath and asthma for few years	Nil	Nodular goitre	No	Normal	Right vocal fold palsy
8	77	M	Respiratory obstruction, increasing dyspnoea	Nil	Follicular adenoma	No	Normal	Normal
9	72	F	Dyspnoea, dysphagia	Inability to sleep comfortably	Follicular Ca	Yes	Normal	Right vocal fold palsy
10	82	M	Dysphonia, dysphagia & stridor	Dyspnoea on lying supine	Colloid goitre	Yes	Normal	Normal
11	95	F	Intermittent hoarseness	Dyspnoea on lying supine	Colloid goitre	No	Normal	Normal
12	73	F	Dyspnoea, hoarse voice	Mild dyspnoea	Papillary Ca	No	Normal	Normal
13	51	M	Incidental finding during investigation for carcinoid tumour	Asymptomatic	Follicular adenoma	No	Normal	Normal
14	78	F	Hoarseness	Intermittent dysphagia with dyspnoea on lying flat	Multinodular goitre	No	Right vocal fold palsy	Right vocal fold palsy
15	63	F	Progressive stridor	Asthma	Benign nodular goitre	Yes	Normal	Normal
16	91	M	Asthma	Stridor, signs of SVC obstruction	Anaplastic Ca	Yes	Normal	Normal
17	82	M	Lump in the neck	Dysphonia	Medullary Ca	Yes	Right RLN sacrificed due to malignancy	Right vocal fold palsy
18	80	M	Hoarseness	Relatively small goitre, not palpable in neck	Benign nodular goitre	No	Left vocal fold palsy	Left vocal fold palsy
19	75	M	Dysphonia	Mild dyspnoea, evidence of SVC obstruction	Not available	Yes	Normal	Normal

RSG = retrosternal goitre; No = number; Pre-op = pre-operative; Post-op = post-operative; F = female; M = male; Ca = carcinoma; SVC = superior vena cava; RLN = recurrent laryngeal nerve

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TABLE II
RETROSTERNAL GOITRE SYMPTOMS[2,14–17,19,23–25]

System	Symptoms
Respiratory	Dyspnoea
	Wheezing
	Choking
	Stridor
Gastro-intestinal	Dysphagia
	'Lump in throat'
Neurological	Vocal fold paralysis/paresis
	Hoarseness
	Horner's syndrome (compression of sympathetic chain)[2]
	Thyroid steal syndrome (hoarseness, choking and dizziness due to vascular compression)[2]
Vascular	Superior vena cava syndrome[2]
	Transient ischaemic attack[2]
	Cerebral oedema [15]
	Gastro-intestinal bleeding[2]
Metabolic	Thyrotoxicosis and weight loss[2]
Infection	Abscess formation[2]

TABLE III
PREVIOUS CITED INDICATIONS FOR SURGICAL REMOVAL OF RETROSTERNAL GOITRES

Study	Indications
Allo & Thompson[4]	Longstanding multinodular goitre
	Elderly patients with underlying thyrotoxicosis
	Suspicion of malignancy
	Evidence of tracheal and oesophageal compression
	Low morbidity in surgical treatment
Newman et al.[17]	Respiratory obstruction (seen in 25% of series)
	(50 per cent tracheal compression may be asymptomatic but any further compression can be catastrophic)
Alfonso et al.[18]	Radiographic signs of tracheal deviation (seen in 33% of series*)

*273 patients

the major vessels and/or contour of the bony thoracic inlet; primary RSG; bilateral RSG; and malignancy within an RSG.

Almost all anterior mediastinal and the vast majority of posterior mediastinal and intrathoracic goitres can be removed by a cervical incision.[18]

Incision and approach

A cervical collar incision was made about 3 cm below the usual thyroidectomy collar incision. In cases in which wider access was required, a further vertical incision downward towards the sternal notch, thus obtaining a 'T-shaped' incision, was sometimes required to improve access, as suggested by Torre et al.[19]
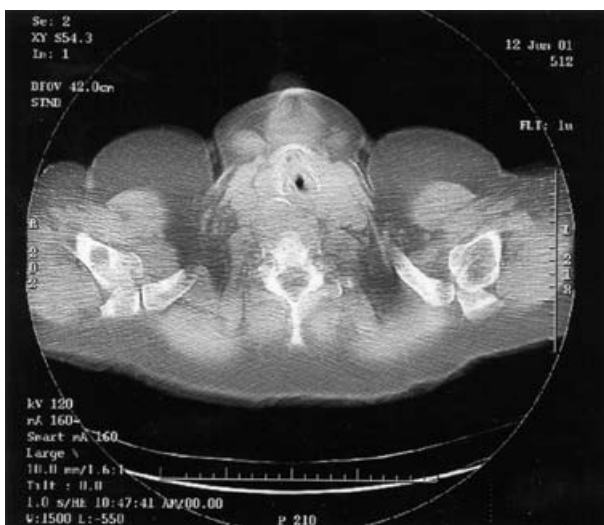

FIG. 1
Computed tomography scan demonstrating the relationship of the retrosternal goitre to the trachea, oesophagus, great vessels and mediastinum.

We divided the strap muscles and exposed the goitre by extra-capsular finger dissection. When the goitre was beyond the reach of the finger, two 'tablespoons' could be used to dissect the goitre from the surrounding structures. Further attempts were then made to extract the goitre through the cervical incision. If this proved impossible or appeared unsafe, a partial or complete sternotomy was performed. In cases in which dissection became unduly difficult owing to apparent adhesions, a frozen section was undertaken to exclude malignancy. In the presence of malignancy, either partial (manubrial) or complete sternotomy was performed, and the goitre was removed en masse.

Thyroid dissection

Using a tablespoon to separate the goitre from surrounding tissues was very effective and significantly less traumatic than using conventional retractors or even finger dissection. The use of sharp retractors can cause damage to the goitre surface, leading to significant haemorrhage. As reported by Katlic et al.,[1] Kocher constructed a special forceps and used a spoon to deliver those masses for which digital extraction failed. Landreneau et al.[23] reported success using a sterile spoon when mobilization by fingers was not possible. Once the thoracic part of the goitre was separated from mediastinal tissue, it was easy to deliver the whole mass using either digital dissection or spoons.

Recurrent laryngeal nerve identification

In our series, identification of the recurrent laryngeal nerve (RLN) was performed by mobilization of the goitre to expose the tracheoesophageal groove. Early, definitive identification of the RLN intra-operatively is of course vital, but can be difficult in cases of RSG, due to the bulk of the goitre and the resultant distortion of the tissue planes and anatomy.[24] The modern nerve stimulator is easy to use and reliable and is therefore widely used.[25] Although useful, the nerve stimulator is not a

substitute for surgical experience. We used the Neurosign® nerve stimulator (The Magstim Company Ltd. Whitland, Wales, UK) for definitive identification of the RLN and also to demonstrate the integrity of the nerve at the completion of RSG resection. If the patient developed an apparent vocal fold palsy in the post-operative period, they could be reassured that the RLN was intact and that recovery was to be anticipated since the likely injury was a neuropraxia.

Vascular and other tissue dissection

In our experience, all but one of our RSG cases was unilateral and therefore the isthmus was divided, the inferior thyroid vessels ligated and the RLN followed to its entrance to the larynx, after which the goitre was removed by ligating and dividing the superior pedicle.

Wax and Briant[15] reported that they did not mobilize the contralateral lobe of the gland if there was no evidence of a malignancy; however, if there was, they performed a total thyroidectomy.

Cho *et al.*[20] stated that, in their experience, the inferior thyroid artery frequently could not be exposed until after delivery of the goitre, and blind ligation of the inferior thyroid artery may endanger the RLN near the cricothyroid joint. They preferred to deliver the goitre from the substernal area first, identify the nerve and then ligate the blood vessels.

In our series, almost all patients had unilateral RSG. We left the normal lobe intact, avoiding enucleation of the parathyroid glands; therefore, the patients did not develop the signs or symptoms of hypoparathyroidism. In cases of bilateral RSG, preservation of the parathyroid glands is particularly important. We normally did not give special attention to identifying the superior laryngeal nerve because the anatomical distortion of the area (due to the large goitre) was significant. Nevertheless, when we ligated the superior pedicle, we made sure that the ligature was applied only to the superior thyroid artery, avoiding ligation of the superior laryngeal nerve.

For larger RSGs, Lahey and Swinton suggested that, instead of sternotomy, it is often possible to decrease the diameter of the goitre by thyroid 'morcellation'.[10] This manoeuvre involves fragmenting or grinding up a large mass in order to facilitate easy delivery of the goitre. Morcellation is not indicated if malignancy is suspected.

Excision of the RSG frequently results in a large and significant 'dead space', and we pack this space with a layer of Surgicel® (Ethicon Inc. NJ, USA). Haematoma formation is always a possibility, and to limit this we place two suction drains, one in the dead space in the mediastinum and the other in the cervical part of the wound.

Conclusion

In the presence of low morbidity and almost zero mortality, early surgical resection of RSG is undoubtedly the most appropriate treatment, even if the patient is asymptomatic. There is no reliable medical treatment available to prevent an enlarging goitre causing acute airway compression or even permanent damage to the function of the RLN. We therefore advocate early surgical removal of RSG by an experienced team comprising both otolaryngological and thoracic surgeons, using the technique described.

- **This paper describes the authors' experiences in managing 19 cases with retrosternal goitre and also reviews the literature on this topic**
- **Surgical excision of retrosternal goitre is shown to be a procedure with very low morbidity and mortality**
- **The authors advocate early surgical excision, even in asymptomatic cases, by an experienced team comprising both otolaryngological and thoracic surgeons**

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