Retrosternal goitre is most commonly defined as one that either descends below the thoracic inlet, or has more than 50% of its volume below this level. A number of other definitions and classifications have, however, been proposed, leading to difficulties in comparing series in the literature. This fact, combined with the poor quality of many studies, means that the evidence base is poor to guide management of those retrosternal goitres without absolute indications for surgery (compressive symptoms, malignancy, thyrotoxicosis). Although surgery is the only effective treatment for retrosternal goitres, in most cases, suppressive therapy with thyroxine is ineffective in reducing the size of multinodular goitres; radioiodine therapy is both generally ineffective in large goitres and may induce acute inflammation and swelling of the gland with the potential for airway obstruction.

Natural history of retrosternal goitres

The natural history of retrosternal goitre is slow, progressive growth, commonly leading to presentation in the fifth or sixth decade of life. Advancing age is associated with increased medical co-morbidity, implying that operation at an earlier stage of goitre development may be associated with reduced complications related to co-existing disease.

The case for surgery due to symptoms associated with retrosternal goitre

Compressive symptoms resulting from a retrosternal goitre, such as cough, dyspnoea, stridor, choking symptoms and dysphagia are regarded as absolute indications for surgery. In addition, radiological indicators of compression (e.g. tracheal deviation) would be seen as an indication for surgery. A proportion of patients with retrosternal goitre are regarded...
as asymptomatic: direct questioning, however, will elicit symptoms which may improve after thyroidectomy, in a large number of such ‘asymptomatic’ patients. Furthermore, the correlation may be poor between symptoms and presence of tracheal deviation, size of goitre or extent of substernal extension as assessed by computed tomography. A policy of using radiological features as indicators for surgery may not, therefore, be appropriate in patients with subtle compressive symptoms. In addition to chronic compressive symptoms, the potential for acute airway obstruction needs to be considered. This may arise from haemorrhage within the thyroid gland or secondary to prolonged mechanical pressure with acute laryngeal oedema and congestion. Although generally thought to be an uncommon complication, in some series of patients with retrosternal goitres acute problems occur with an incidence of between 5–11%. Clearly, development of this complication can have catastrophic consequences, providing a clear rationale for thyroidectomy in patients with retrosternal goitre.

Risk of malignancy in retrosternal goitre

The majority of multinodular goitres, both limited to the neck and including a retrosternal component, are benign in nature. Cervical goitres are amenable to careful clinical and ultrasonographic examination and needle biopsy of suspicious areas, with cytological determination of malignant nodules, leading to patient selection for surgery. Retrosternal components of goitres are not easily imaged by ultrasound due to artefact generated by bony structures. Similarly, intrathoracic nodules are inaccessible to needle biopsy in most instances. This makes the exclusion of malignancy fraught with difficulty in retrosternal goitres. Prospective studies document the incidence of carcinoma development in goitres at 1.3–5.7 new cases per 1000 patients. A recent review of evidence-based management of substernal goitres concluded the incidence of malignant transformation is equivalent in retrosternal goitres to those residing entirely in the neck. Most of these cancers will represent differentiated thyroid carcinomas, with good prognosis, but the possibility should be remembered of development of a more aggressive carcinoma (e.g. anaplastic carcinoma). Certain well-documented risk factors for thyroid carcinoma exist, namely, history of neck irradiation and family history of thyroid cancer. Most cancers encountered do not have obvious predisposing features and, therefore, lack of risk factors alone cannot ensure the benign nature of the goitre. This inability to rule out malignancy in retrosternal goitres provides a further rationale for performing thyroidectomy in these cases.

Operative approach and complications of surgery for retrosternal goitre

Only around 2% of patients undergoing thyroidectomy for retrosternal goitre will require surgical access other than a standard collar incision (either manubriotomy, sternotomy or thoracotomy). Review of the literature regarding the complications of recurrent laryngeal nerve injury, hypoparathyroidism and tracheomalacia after retrosternal goitre excision reveals conflicting results as to whether these specific complications are increased by comparison to cases of excision of simple cervical goitre. Thyroidectomy for retrosternal goitres should be performed by surgeons with a specific interest and experience in thyroid surgery to ensure that complications are minimised and a cervical approach is successful in the maximum number of cases.

Conclusions

Thyroidectomy should be performed in all patients with retrosternal goitre who do not have medical co-morbidity excluding them from surgery. This recommendation is based on the risks of airway obstruction and malignancy, the presence of symptoms in the majority of patients on direct questioning and the tendency for the goitre to demonstrate time-dependent progressive growth. Surgery is associated with low morbidity when performed by experienced thyroid surgeons.

References

The treatment of asymptomatic retrosternal goitre is one of several ongoing controversies in the surgical management of thyroid disease. The lack of high-level evidence to guide the management of retrosternal goitre has been clearly illustrated in a recent comprehensive review. However, the review failed to address the need for surgery in patients with an ‘asymptomatic retrosternal goitre’. Although the majority of patients in published series are symptomatic, the prevalence of thyroid incidentalomas in the neck and mediastinum has risen with the increasingly prolific use of imaging modalities such as ultrasound, computed tomography and PET scanning for conditions unrelated to the thyroid gland. The uncertainty surrounding treatment of this condition is exacerbated by the ambiguity surrounding what constitutes a retrosternal goitre. Although a small proportion are purely intrathoracic, the majority of retrosternal goitres are an extension of thyroid tissue from the neck. Several definitions have been proposed to clarify the meaning of a retrosternal goitre, some of which include a goitre: (i) that descends below the plane of the thoracic inlet; (ii) with more than 50% of the mass lying below the plane of the thoracic inlet; (iii) with major intrathoracic extension requiring reaching into the mediastinum for dissection; (iv) growing into the anterior-superior mediastinum to a depth of >2 cm; or (v) reaching the level of the fourth thoracic vertebra.

This article assumes that most clinicians subscribe to the first definition and suspect/identify a retrosternal goitre on clinical examination, which may then be confirmed by imaging or at surgery.

A non-operative approach is considered to be the standard of care in the treatment of an asymptomatic cervical goitre in the absence of clinical risk factors or suspicious/malignant cytology. In contrast, many authors have traditionally recommended surgery in asymptomatic retrosternal goitre as a prophylaxis against airway and venous compression (due to bleeding or rapid growth), hyperthyroidism and cancer. The recommendations are based on uncontrolled, observational case series that include only a small proportion of truly ‘asymptomatic’ patients. In a recent series of 19 patients with retrosternal goitre in which only one patient was asymptomatic, the authors advocated ‘early excision, even in asymptomatic cases’. The findings of malignancy and airway compromise in selected series of patients with predominantly symptomatic retrosternal goitre do not support an argument for surgery in the wider population of asymptomatic retrosternal goitre, as the proportion of patients with retrosternal goitre who would develop symptoms is largely unknown.

Differences in the recommendations for the treatment of cervical goitre and of retrosternal goitre seem paradoxical as the latter is not a biologically distinct entity but simply a variant (of nodular goitre) by virtue of its anatomy. There is no evidence that the natural history of retrosternal goitre is different to that of a cervical goitre. A study of 672 patients with multinodular goitre showed no evidence of an increased risk of cancer in patients with a retrosternal goitre. Histological examination of multinodular goitres shows the presence of cancer in up to 14% of patients. One argument for early surgical intervention in patients with asymptomatic retrosternal goitre is the inability to obtain a tissue sample to exclude cancer. Although the incidence of malignancy in ‘asymptomatic’ goitres has not been quantified, it would be expected to be lower than in symptomatic goitres.

Although several studies from single centres demonstrate low morbidity and mortality following surgery for retrosternal goitre, a large observational study that included 52,777 thyroidectomies (of which 1153 were on retrosternal goitre) from multiple centres within the state of New York has convincingly demonstrated that retrosternal thyroidectomy (in comparison with cervical thyroidectomy) was associated not only with an increased risk of complications – such as recurrent laryngeal nerve damage (2.1% versus 0.6%, respectively), hypoparathyroidism (5.5% versus 3.5%, respectively) and postoperative bleeding (2.2% and 0.9%, respectively) – but also with increased mortality (1.4% versus 0.1%, respectively).

The term ‘asymptomatic’ may carry different connotations for clinicians and patients and has to be considered in the context of the patient’s general health. For example, surgery may well be indicated in a patient with retrosternal goitre who has a long standing (albeit mistaken) diagnosis of ‘stable asthma’, but is otherwise ‘asymptomatic’. In such cases, findings from flow volume loop measurements in addition to conventional radiology may provide further evidence of upper airway obstruction, which may aid in decision making. We agree that radiological evidence of significant tracheal narrowing and potential airway obstruction...
may be an indication for surgery in a clinically asymptomatic patient. This, however, should not be extrapolated to warrant a recommendation for surgery in all patients with asymptomatic retrosternal goitre.

Conclusions

We do not believe a case has been made for routine surgery in ‘asymptomatic retrosternal goitre’. The argument to support surgery in this condition would seem untenable in iodine-deficient regions, where ultrasound screening of individuals without thyroid disease has documented retrosternal thyroid tissue in 25% of patients above 70 years of age. Well-conducted, prospective studies on the natural history of this condition will be invaluable in providing data to guide management and possibly to predict the need for surgery to prevent complications in selected patients. Until this is available, a policy of observation is justified in a patient with asymptomatic retrosternal extension of a nodular goitre, provided there is neither a suspicion of malignancy nor any evidence of progressive enlargement.

References