



Review article

Intrathoracic goitre: A literatura review

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ARTICLE INFORMATION

Article history:

Received August 26, 2009

Accepted March 16, 2010

Keywords:

Intrathoracic goitre

Substernal

Mediastinal

Palabras clave:

Bocio intratorácico

Subesternal

Mediastinal

A B S T R A C T

Intrathoracic goitre is defined as that goitre which is partially or totally found in the anterior or posterior mediastinum, and its incidence is associated with multinodular goitre. The diagnosis is relatively easy, mainly with imaging tests, and treatment varies, but of choice, surgical treatment is recommended. This article attempts to present a review of the literature on the diagnosis and management of this disease.

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Bocio intratorácico. Revisión de la literatura médica

R E S U M E N

El bocio intratorácico se define como aquel bocio que se encuentra parcial o totalmente en el mediastino y que puede localizarse en el mediastino anterior o posterior; su incidencia está en relación con el bocio multinodular. El diagnóstico se realiza con cierta facilidad, en su mayoría con pruebas de imagen, y el tratamiento es variado, pero de elección se recomienda la cirugía. Con este artículo se pretende realizar una revisión de la literatura médica tanto del diagnóstico como del tratamiento de esta enfermedad.

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Haller described intrathoracic goitre in 1749 and, from that time, it has been given several different names: retrosternal, substernal, subclavicular, mediastinal, among others.^{1,2} The main problem is the fact that there is no uniformity in the definition of this clinical and pathological entity, with no unanimous opinion as to the amount of the thyroid gland that should be located in the thorax or how low it must

descend, to be considered as intrathoracic goitre.^{2,3} Dahan et al define it as goitre that is totally or partially located in the mediastinum and that, when the neck is in an over-extended position, has its lower edge at least 3 cm below the manubrium of the sternum.^{2,4} Other authors define it as a thyroid gland that has grown to double its normal size and has 50% of its tissue below the suprasternal notch.^{1,5,6}

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Incidence

In general, the incidence of intrathoracic goitre is related to the incidence of multinodular goitre; therefore, it is more frequent in areas with endemic iodine deficiency.^{2,6,7} Currently, due to iodization programmes, the incidence rate has fallen. Another factor contributing to the different incidence rates of this disease is the lack of uniformity of the criteria for its definition, as was detailed above. However, authors such as Newman have described a prevalence in the US population of between 0.02%-0.5%.^{1,8} The medical literature reports an incidence for this type of goitre in patients who have undergone thyroid surgery of between 1%-20%,^{3,9-11} which represents between 3%-12% of all mediastinal masses.^{5,8} In Spain there is no data on the prevalence of this disease, and some series, such as the one described by Ríos, report that about 37% of multinodular goitres have an intrathoracic component.^{2,7}

Anatomical classification

Although intrathoracic goitre is frequently located in the upper mediastinum, it can be in the anterior mediastinum, which is the most common location, or in the posterior mediastinum, posterior to the trachea (10%-15%) or oesophagus (less frequent).^{3,10,12}

It is usually classified as primary and secondary. Primary mediastinal goitre is truly intrathoracic. It represents 1% of all intrathoracic goitres. It is not related to the cervical thyroid gland and is congenital; furthermore, in most cases their blood supply comes from the intrathoracic vessels. It originates from embryonic ectopic thyroid cells that descend to the thorax via the aortic arch.^{3,10,12}

The second group, acquired retrosternal goitre, is much more common. It arises in the cervical thyroid gland, due to the growth and descent of this gland into the thorax. Lahey and Swinton described some anatomical factors that lead to this descent, such as the fact that the gland is bound by rigid structures (vertebrae, cervical fascia, thyroid and cricoid cartilages, etc.), except on its lower border. Therefore, there is less resistance in this direction. Furthermore, this is aggravated by negative intrathoracic pressure, descending traction caused by swallowing and the weight of the gland.^{2,3,8,10}

Pathogenesis

The main cause is multinodular goitre (51%), followed by follicular adenoma and chronic autoimmune thyroiditis. The incidence of malignancy varies between 3%-16% and its incidence increases with patient age.^{2,4,8,9,13} Some of the factors that contribute to gland growth are lack of iodine and a defect in thyroxine synthesis that increases TSH levels and accelerates the growth of follicular cells.

Clinical symptoms

According to most authors, this condition is asymptomatic in 20%-30% of cases. It is seen more frequently in women, with a ratio of 3:1, and, generally, between the 5th-6th decade of life. Patients experience symptoms after many years of evolution, due to the slow growth of this lesion. The most commonly reported symptom is a cervical mass (in between 40%-50% of cases). There may be secondary symptoms caused by compression of intrathoracic structures, among which are dyspnoea, stridor, shortness of breath, all caused by airway compression. In some series it is reported that 50% of patients with these goitres had these symptoms. Other compressive symptoms are dysphagia, seen in about 30%-40% of cases (more frequent in posterior mediastinal goitre), rhonchus in 13%, and other more infrequent symptoms caused by vascular compression that cause oesophageal varices with digestive haemorrhages, superior vena cava syndrome, transient ischemic events and cerebral oedema. In 80%-90% of these patients a palpable cervical mass, among other symptoms, is observed. Pemberton's sign and tracheal deviation, amongst others, can also be observed.^{2,3,7,11,12,14,15}

Diagnosis

A simple chest X-ray shows a mediastinal mass or a thickening of the upper mediastinum, as well as tracheal deviation from the midline and different degrees of compression. Computed tomography (CT) provides precise information on the relationships between intrathoracic organs and goitre, and is very useful to the surgeon when planning the type of approach to use. There is less experience with magnetic resonance, but, as with CT, it provides important information. Ultrasound in the thorax is limited by the interference caused by the rib cage and the sternum. Thyroid scintigraphy can be useful for differential diagnosis of goitre from other mediastinal masses; however, it may result in false negatives in the case of cold nodules. And, lastly, fine-needle puncture is very important to diagnose cervical goitre, but is not indicated in mediastinal goitre, due to the risk of haemorrhage, as it causes acute compression of the airways.^{2,8,12}

Treatment

There are different forms of treatment; below we briefly describe each of them.

Pharmacotherapy

Patients with elevated TSH or defects of thyroxine synthesis are candidates for suppressive treatment. Some physicians have used thyroxine hoping to reduce the size and volume of the goitre, but medical literature refutes this, since only 20%-30% of patients respond to this treatment after one year and when the treatment is suspended, the gland increases in

size again. Moreover, goitres with cystic and haemorrhagic changes do not respond to this treatment.

Treatment with radioactive iodine

This has been used in toxic goitre. However, it has been used recently in patients with non-toxic thyroid goitres in the USA, and has been shown in some studies to reduce goitre by 40%, although 1 in 3 patients has no improvement of compressive symptoms. Furthermore, its complications are thyroiditis, transient increase of thyroid volume with the risk of acute compression of the airways, neck pain and dysphagia, among others. It is therefore only used in those patients who have a high surgical risk or in those who cannot undergo surgery.^{7,12,16}

Surgery

The most effective, and most recommended treatment, is surgery. The extent of surgery is a subject of controversy. Partial resection is recommended since it minimises surgical risk of damage to the parathyroid glands and recurrent nerves; however, a significant number of patients suffer a relapse of the lesion 20-30 years later (12%-20%).^{2,17}

Therefore, most authors consider that the technique of choice is total thyroidectomy, especially in those patients with a life expectancy greater than 10 years.^{2,3,12,14,18}

Different surgical approaches can be used. The most commonly used is a surgical approach with a collar incision, since more than 90% of all goitres with an intrathoracic component can be extracted using this technique.¹⁵

In a certain number of patients it is necessary to use a cervicothoracic approach, especially in those patients with primary goitres and those located in the posterior mediastinum. Partial or total median sternotomy, thoracotomy and thoracoscopy are different routes used to resect certain intrathoracic goitres. The choice depends on the location, size and relationship of the mass to vital mediastinal structures. This information is obtained from the imaging tests.^{2,3,12,19}

Median sternotomy is the preferred route and is indicated in^{2,3,12} very large intrathoracic goitres with a blood flow derived from intrathoracic vessels, in goitre in the posterior mediastinum that displaces or compresses the aortic arch associated with superior vena cava syndrome, recurrent goitre and malignant substernal goitre with lymph node metastasis.

Thoracotomy may be appropriate in intrathoracic goitre located in the middle or posterior mediastinum.^{2,12}

Some authors have reported resection using thoracoscopy, but this is probably only appropriate in very small goitres, since it is a very difficult technique.^{12,19} However, Al-Mufarrej et al described in 2008 two cases of resection of posterior mediastinal goitres with minimally invasive surgery using the Da Vinci robot, but this technique needs to be evaluated with further studies.²⁰

Among the complications that can arise are^{2,6,12,18} hypocalcaemia due to damage to the parathyroid glands, damage of the recurrent nerves, haematomas and haemorrhages, surgical wound infections, complications

affecting the airways, such as post-operative tracheal collapse or prolongation of mechanical ventilation due to tracheomalacia, and respiratory tract infection, among others.

These complications are very rare, Ríos Zambudio et al, in a prospective study of 301 thyroidectomies for multinodular goitre found 1% of definitive complications. Furthermore they identified hyperthyroidism and goitre size as risk factors for these complications. On the other hand, Pieracci et al, and the majority of authors, report that thyroidectomy morbidity and mortality for mediastinal goitre is slightly greater than that of cervical goitre, but this is not statistically significant and both are scarce,^{6,12,21} although some authors such as Sancho et al have found that the intrathoracic goitres that reach the carina have a greater risk of sternotomy, post-operative complications, repeat interventions and death.²¹

Conclusions

Intrathoracic goitre has a poor response to thyroxine treatment. Therefore, surgery is the treatment of choice, and preferably total gland resection if it can be removed by a cervical approach, although a thoracic approach may be used in posterior ectopic goitres. Iodine treatment is possible in high risk patients.

Conflict of interests

The authors declare they do not have any conflict of interests.

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