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2530-0180/

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The first reported case of *struma cordis* in Spain[☆]



El primer caso descrito de *struma cordis* en España

Introduction

Struma cordis, or cardiac ectopic thyroid tissue, is a rare condition that affects 1 in every 100,000–300,000 people, and is mostly diagnosed in middle-aged women.¹

The thyroid primordium starts between days 24 and 32 of pregnancy from a thickening of the endodermal epithelium located at the base of the embryonic midline. In turn, the base of the tongue forms and is commonly referred to as the foramen or blind orifice. The appearance of ectopic thyroid tissue is the result of aberrant embryogenesis during migration of the thyroid primordium in the fourth week of pregnancy. Such migration normally takes place from the foramen to the typical thyroid gland location anterior to the second-fourth tracheal cartilage.^{1,2}

According to necropsy studies, the prevalence of ectopic thyroid tissue ranges between 7 and 10%, and the diagnosis is usually established in the first three decades of life. The most common site of appearance is the base of the tongue (in up to 90% of cases), though thyroid remnants may be found over the entire trajectory of the thyroglossal duct, and even in the mediastinum.² The first case of intrapericardial ectopic thyroid tissue was reported following necropsy in 1941.³ In most reported cases, the thyroid tissue is located in the interventricular septum or right ventricle, with obstruction of the right ventricular outflow tract.^{2,4}

Case report

We present the first case of *struma cordis* described in Spain. The patient was a 28-year-old woman with no relevant dis-

ease history or toxic habits, referred to Cardiology for the evaluation of a heart murmur of recent onset.

She had no dyspnea, chest pain or other evidence of heart failure. The physical examination revealed little more than a mild tricuspid systolic murmur at auscultation.

The blood tests and electrocardiographic findings were within normal limits. Transthoracic (TTE) and transesophageal echocardiography (TEE) revealed a 3.5 cm³ broad-pediced and scantily mobile tumor lesion in the intra-ventricular septum of the right ventricle, close to the tricuspid valve (Fig. 1).

A midline sternotomy was performed, with a right atriotomy and removal of the well delimited mass supported by the interventricular septum. The pathology study revealed the presence of ectopic thyroid tissue with focal fibrosis and dystrophic calcifications. There were no signs of malignancy.

Following the histological findings, a neck ultrasound study was carried out to verify the presence of eutopic thyroid tissue, and thyroid function testing after surgery showed TSH 1.3 mIU/l (reference range: 0.25–5). Thyroid scintigraphy revealed a single focus of radiopharmaceutical uptake in the anterocervical region.

Discussion

Although the molecular mechanisms underlying thyroid dysgenesis are not fully understood, mutations of genes *TITF-1* (*Nkx2-1*), *Foxe1* (*TITF-2*) and *PAX-8* have a crucial impact upon thyroid morphogenesis and differentiation, and may cause alterations in thyroid migration, as well as conditioning the appearance of ectopic thyroid tissue remnants. The literature on this subject is scarce, though cases with familial aggregation have been reported.¹

Most cases of thyroid ectopia are located on the midline of the neck along the trajectory of the thyroglossal duct from the base of the tongue to the diaphragm, and coexist with normal thyroid tissue. The clinical manifestations of thyroid ectopia range from the absence of symptoms to altered thyroid gland function. The most common manifestation of thyroid dysfunction is the appearance of hormonal hypofunction, which in turn is related to the amount of ectopic thyroid tissue. However, symptoms of hyperthyroidism have been reported, even with histological features similar to those of Graves' disease. Depending

[☆] Please cite this article as: Díaz-Perdigones CM, Cano-Niego J, Hernández-García C, López-Medina JA, González-González S. El primer caso descrito de *struma cordis* en España. *Endocrinol Diabetes Nutr.* 2018;65:472–473.



Figure 1 Transthoracic echocardiography showing *struma cordis* at right ventricle level.

on the location and size of the mass, some patients may experience dyspnea, stridor, central chest pressure sensation, palpitations and ventricular arrhythmias, particularly in the case of mediastinal or intracardiac presentations, though such lesions usually constitute incidental findings.^{4,5}

If the transthoracic or transesophageal echocardiography findings suggest the presence of ectopic thyroid tissue, a neck ultrasound exploration should be performed on a complementary basis. This will allow us to either confirm or discard the presence of a thyroid gland, with a view to determining that *struma cordis* is not the only existing thyroid tissue. Radionuclide I-123, I-131 or Tc-99 scintigraphy may be useful for determining the existence of other ectopic thyroid tissue remnants, though the identification of two concomitant ectopic sites is very uncommon.⁴⁻⁶

Another suitable option for anatomical and functional characterization in the event of suspected mediastinal thyroid tissue is SPECT-CT. The typical CT image of ectopic thyroid tissue is a well delimited homogeneous mass with an attenuation index of 70 ± 10 HU, which suggests a benign nature. In addition, magnetic resonance imaging (MRI) is used, in particular for diagnosing a lingual thyroid gland when there are difficulties in differentiating between thyroid tissue and tongue muscle.⁴⁻⁶

The histological study provides the definitive diagnosis of *struma cordis*. The differential diagnosis is established mainly with metastasis of follicular thyroid carcinoma or primary cardiac tumors such as myxoma.⁷

Surgery is the treatment of choice, with resection of the ectopic tissue using cardiopulmonary bypass techniques, but should be limited to symptomatic patients in view of the high surgical risk involved. Treatment with I-131 may be considered in patients who are not eligible for surgery, or even as an alternative management option in patients who refuse surgery.⁷ In other ectopic thyroid tissues, treatment with levothyroxine may reduce the size of the mass. This has not been demonstrated in the case of *struma cordis*, however.

Thyroid function should be checked after surgery. The recommended timing for this is between the third and sixth week after the operation. In the absence of a eutopic thyroid gland, it is advisable to start levothyroxine at doses of $1.2\text{--}1.8 \mu\text{g/kg/day}$ after surgery.⁸ The long term prognosis is good if there have been no complications secondary to surgery.⁷⁻⁹

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2530-0180/

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