Right ventricular outflow tract tumour: an unsuspected intracardiac ectopic thyroid mass

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Abstract

Ectopic thyroid is a rare embryogenic anomaly that occurs during its migration from foramen caecum to its pretracheal position. An intracardiac ectopic location is even rarer and found most commonly in the right ventricular outflow tract in sporadic reports. While surgery in symptomatic patients seems appropriate, resection of non-neoplastic ectopic tissue remains a clinical equipoise. Its occurrence is often unsuspected by clinicians, but its possibility should be considered due to its typical location in the right ventricular outflow tract. Unlike true neoplastic intracardiac tumour which mandates surgical resection, both surgical and non-surgical approach may be considered for an intracardiac ectopic thyroid mass.

Keywords: Cardiac • Tumour • Surgery • Thyroid

INTRODUCTION

With a prevalence of ~1 per 100 000–300 000 people, ectopic thyroid tissue is a rare developmental abnormality as a result of aberrant embryogenesis when it descends from the floor of the primitive foregut to its final pretracheal position [1]. Lingual thyroid at the base of tongue is the most form of ectopic thyroid, while intracardiac form is extremely rare [1]. Its occurrence is often unsuspected by clinicians and rarely considered during their work-up. We presented a case report and discussed the management approach of this rare non-neoplastic intracardiac tumour.

CASE REPORT

A 46-year old lady presented with non-specific chest pain. Transthoracic echocardiography showed a 2.6 cm right ventricular outflow tract (RVOT) mass attached to the interventricular septum (Fig. 1). Computed tomography scan did not show evidence of primary tumour elsewhere. Preoperative angiography showed normal coronaries and a large feeding vessel to the tumour.

In view of its potential to obstruct the outflow tract, surgery was performed. On normothermic bypass and cardioplegic arrest, the tumour was exposed via a transannular incision across RVOT preserving the pulmonary valve leaflets. A broad-based, globular tumour was found within the septum and completely resected. Histology confirmed an unencapsulated but well-defined nodule of thyroid tissue, composed of follicles of varying size, with a uniform texture of colloid and no architectural or nuclear features of papillary carcinoma of the thyroid consistent with ectopic thyroid tissue.

Neck ultrasound showed normal thyroid parenchyma with tiny colloid cysts but no solid lesion to suggest neoplasm. A normal thyroid function test was also confirmed postoperatively. Following these investigations, she was discharged home 2 weeks following surgery.

DISCUSSION

Since its first necropsy description in 1941, most intrapericardial ectopic thyroid cases were reported after 1980 with ~10 cases every decade (Table 1). Three-quarter of intracardiac ectopic thyroid was found within the RVOT, and usually originated from the interventricular septum. In a single case report, two synchronous masses were found in the left ventricular outflow tract and interventricular septum in the same patient [2].
Excluding necropsy findings, the mass was found incidentally in almost a third of patients, usually after discovery of a heart murmur (Table 1). Dyspnoea and palpitation are two common symptoms, presumably secondary to outflow tract obstruction and irritation of conduction pathways. Death due to cardiac arrest with recurrent ventricular tachycardia and severe RVOT obstruction has been reported with a large ectopic thyroid mass. Typically, thyroid function tests and neck USS are normal.

Although secondary tumours are far more common in the heart, intracardiac ectopic thyroid is usually solitary and presents more like a primary cardiac neoplasm. Secondary tumour without dissemination elsewhere is very rare. Cardiac myxoma of RVOT origin has been reported but, unlike broad-based ectopic thyroid mass, myxoma is usually pedunculated [3].

Most patients underwent surgical excision, and this should be undertaken with the aim of completely excising the mass, with a low threshold to repair the tricuspid valve or patch any septal defect if necessary. Favourable clinical outcome had been reported post-resection (median follow-up 3 years, maximum 13 years; 1 died early postoperatively due to renal failure) and resolution of fatal arrhythmia had been described post-operatively [4].

Although an intracardiac ectopic thyroid is extremely rare, its location is typical and the detection of a RVOT in middle-age female patients should raise the suspicion of an ectopic thyroid mass and be further confirmed by thyroid scintigraphy [1]. Surgical resection should be offered to symptomatic patients with right ventricular outflow obstruction, where recurrent arrhythmia is a problem or when diagnosis is unclear. Management for asymptomatic patients is debatable. By definition, an ectopic thyroid consists of normal thyroid tissue occurring out of its usual location and is therefore not neoplastic. It would have been present since birth, but does not portend a problem until much later in life. If this diagnosis were confirmed preoperatively, a small incidental mass without any haemodynamic effect in an otherwise asymptomatic patient may not always require surgical resection. In at least four reports, the mass was either biopsied or only partially resected to avoid damaging intracardiac structures [2]. In these cases, maximum follow-up to 5 years, no further growth of the mass has been reported, but there was progressive tricuspid regurgitation in one patient [2]. In non-cardiac ectopic thyroid, the role of suppressive hormonal therapy using levothyroxine or radioiodine ablation to reduce the size of thyroid mass has been described [1], but its role in intracardiac ectopic thyroid is unknown. If a conservative approach is chosen, two important considerations must be taken into account (i) potential growth of the mass and (ii) potential malignant transformation, as postulated in a single report, mandating a close follow-up in these patients [5].

In summary, an ectopic thyroid is an extremely rare intracardiac tumour that is often unsuspected by clinicians. However, this rare possibility should be considered due to its typical location in the RVOT and its common occurrence in female patients. This tumour is readily confirmed by thyroid scintigraphy, and unlike true neoplastic intracardiac tumour which mandates surgical resection, both surgical and non-surgical approaches may be considered for an intracardiac ectopic thyroid.

Figure 1: (A) Transthoracic echocardiography demonstrating a right ventricular outflow tract mass (circle) arising from the septum in parasternal short-axis view; (B): Preoperative angiogram demonstrating a feeding vessel (white arrow) from the left anterior descending artery supplying the tumour; (C) resected mass weighing 6 g and measuring 2.4 × 1.9 × 1.8 cm with a cross-section showing a yellow, focally myxoid appearance.
Table 1: Summary of literature of reported intrapericardial ectopic thyroid

<table>
<thead>
<tr>
<th>Years reported</th>
<th>n = 36</th>
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<tbody>
<tr>
<td>Year &lt;1980</td>
<td>2</td>
</tr>
<tr>
<td>1980–1989</td>
<td>11</td>
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Age (median, range) 55.0 (25.0–74.0) year
Gender 30 females 83.3%

Presentation
Incidental 10 27.8%
Symptomatic 22 61.1%
Necropsy 4 11.1%

Symptoms (n = 22 patients)
Dyspnoea 10 45.5%
Palpitation 7 31.8%
Dizzy/presyncope 4 18.2%
Syncope 3 13.6%
Chest pain 1 4.5%

Location
Intraventricular septum 25 (RVOT-23, LVOT-1, IVS-1) 69.4% (RVOT-64%, LVOT-2.8%, IVS-2.8%)
RV outflow tract 4 11.1%
RV free wall 1 2.8%
Right atrium 1 2.8%
Aorta** 4 11.1%
LVOT and septum* 1 2.8%

Surgery (n = 30 patients)*
Excision 22 71.0%
Excision and tricuspid surgery 4 12.9%
Excision and septal patch 3 9.7%
Partial resection 1 3.2%

*One patient with two synchronous masses—one was excised and one was left alone.
**Three ascending aorta, 1 aortic root (attached to adventitia, not intraluminal).

RVOT-RV: right ventricular outflow tract; LVOT-LV: left ventricular outflow tract; IVS: intraventricular septum.

Conflict of interest: none declared.

REFERENCES


