Case report - Cardiac general

Struma cordis in a Jamaican woman

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Abstract

A case of ectopic thyroid within the right ventricle (struma cordis) occurring in a middle-aged Jamaican woman is described. This rare and fascinating condition is curable utilizing standard open-heart surgical techniques. Investigations to rule out primary thyroid or ovarian malignancy, as well as the presence of remaining normal thyroid tissue or other ectopic thyroid rests are important in the management of these patients.

Keywords: Struma cordis; Cardiac tumours; Ectopic thyroid

1. Case report

A 46-year-old woman presented to the University Hospital of the West Indies with a complaint of chronic left lower quadrant abdominal pain and menorrhagia. She also complained of a recent onset of dyspnoea and palpitations which were worse on recumbency. She was a hypertensive who had been previously diagnosed with a left adnexal mass. She was considered for gynecological surgery; however on examination a cardiac murmur was detected.

An electrocardiogram demonstrated right ventricular hypertrophy with left bundle branch block and a chest radiography demonstrated cardiomegaly with normal pulmonary vascular distribution. Echocardiography demonstrated a large mass within the right ventricle obstructing the outflow tract in the manner of a ball-valve (Fig. 1A). The mass appeared to be attached to the apical posterior portion of the inter-ventricular septum. All other chambers and valves appeared normal. Provisional diagnoses included a right-sided cardiac myxoma, a rhabdomyosarcoma cordis or a metastatic deposit from an ovarian malignancy.

Cardiac surgery to remove the mass was embarked upon via a median sternotomy and bicaval cannulation for cardiopulmonary bypass. After cardioplegic arrest, the right atrium was opened and the tricuspid valve retracted, revealing a 6 cm x 5 cm x 3.5 cm white oval mass attached via a broad pedicle to the ventricular septum 2 to 3 cm from the pulmonary valve and extending to the adjacent anterior free right ventricular wall including the moderator band. The tumour was excised with a rim of adjacent septal muscle. The mass possessed a thick fibrous capsule and its cut surface revealed soft yellow tissue (Fig. 1B).

A frozen section analysis of the tumour was performed, followed by formal histological processing. Histologically it was composed of colloid filled follicles of variable size, lined by cuboidal to flattened follicular epithelial cells. Areas of recent and old haemorrhage, cystic degeneration and fibrosis were present throughout. The tissue was superficially infiltrating the adjacent normal cardiac muscle (Fig. 2). The cardiac muscle resection margin was free.

Her post-operative stay in the hospital was uneventful and she was discharged seven days later.

The diagnostic possibilities of a metastatic well-differentiated follicular carcinoma of the thyroid gland or metastatic struma ovarii were thought likely. Intra-cardiac
ectopic thyroid (struma cordis) was considered to be a diagnosis of exclusion.

Post-operatively, thyroid function tests remained normal and a thyroid scan demonstrated normal cervical thyroid tissue and no other ectopic thyroid sites. Three months later she underwent a total abdominal hysterectomy and bilateral salpinooophorectomy. Examination of the specimen revealed left hydrosalpinx with normal ovaries, resulting in a final diagnosis of struma cordis.

Recovery was again uneventful and 2 years after surgery she was euthyroid with excellent cardiac function.

2. Comment

Ectopic thyroid in the heart (struma cordis) is a rare finding without a percentile incidence and only 20 cases reported prior to this one [1]. The ages at presentation range from 25 to 67 years. The vast majority (>85%) of patients have been female (M:F 1:6).

Histologically normal thyroid tissue has been reported in many ectopic locations. Classically, ectopic thyroid tissue has been described in the so-called Woelfer’s area, formed by an isosceles triangle with the edge of the mandible as its base and the concavity in the aortic arch as its apex [2]. Ectopic thyroid tissue has been described from the foramen caecum of the tongue to the diaphragm and is subject to the same diseases that affect the normal thyroid gland. Primary malignancy has indeed been reported in ectopic thyroid in both non-cardiac and cardiac sites [3,4].

The association with the heart is based on the close relationship between the thyroid anlage and the heart in young embryos. In the 17-day-old, 2 somite embryo, the heart is formed by a pair of endothelial tubes surrounded by a myocardial mantle which is open dorsally. These components are in close contact with the area of proliferating endoderm in the floor of the pharynx which is the primordium of the thyroid gland. In the 7 somite 19-day-old embryo, this primordium of the thyroid gland becomes adherent to the heart. By 20 days old, the endothelial tubes fuse and are enclosed within the epicardial mantle, which is now closed dorsally. The heart then descends, however, the descent of the thyroid is arrested by the association of the rapidly developing lobes with adjacent structures.

At the two extremes, portions of the thyroid anlage may be retained within the heart between the endocardium of the right ventricle and the myocardium of the interventricular septum in the region of the conus, or, when no intimate relationship exists, may fail to descend at all producing a lingual thyroid. Clearly, any location in between may exist and thyroid tissue has been described in the suprathyroid region, the infrathyroid region, within the thyroglossal tract, within the trachea, lower pretracheal region, mediastinum, adjacent to the aortic arch or between the aorta and pulmonary trunk, or within the pericardium [1].

When found in the heart it is usually found in the right ventricle on the inter-ventricular septum. About half of the time when its exact position is mentioned, it is located 1–2 cm below the tricuspid valve and the rest of the time just below the pulmonary valve or otherwise unspecified location on the interventricular septum. Location within the left ventricle has been reported (2 of 20 cases) [5]. In all cases, however, it has presented with ventricular outflow tract obstruction of varying degrees with or without ventricular dysrhythmias [6].

Ectopic thyroid tissue in the heart can usually be excised completely, and safely, utilizing standard cardiopulmonary bypass techniques. Once a histological diagnosis is made it is imperative that the existing thyroid be investigated to rule out a well differentiated follicular thyroid malignancy with metastasis to the heart. thyroid function tests should be performed post operatively to confirm the presence of remaining normal thyroid tissue. The long-term prognosis for this fascinating condition is good.

References