Short reports

Intracardiac ectopic thyroid: a case report and review of published cases

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Abstract

A woman with ectopic intracardiac thyroid tissue showing features of a colloid storage goitre presented with ventricular tachycardias and signs of right ventricular outflow obstruction. As the tumour was benign and removed at necropsy, the results of surgery are likely to have been good, but she died from a cardiac arrest before operation.

Intracardiac ectopic thyroid tissue is rare, only six cases having been reported worldwide. We report the first example recorded in the United Kingdom, and give a brief review of the published reports.

Case report

A 63 year old woman was referred to the regional cardiothoracic centre for surgical treatment of an intracardiac tumour. She had presented elsewhere in 1978 with palpitations, when she was found to be hypertensive with prominent right and left ventricular impulses. A third heart sound was audible, with an ejection click and an ejection systolic murmur in the pulmonary area. Electrocardiography showed sinus rhythm and right bundle branch block. Interestingly, a cardiac murmur had been noted 28 years previously after the birth of her second child.

She was treated with antihypertensive drugs and her palpitations improved. The provisional diagnosis of an acyanotic Fallot's tetrad was made but the patient refused cardiac catheterisation. She remained well until 1981, when she was admitted with further episodes of palpitation, which were found to be due to ventricular tachycardia. She was successfully treated with disopyramide. In 1985 she had further episodes of ventricular tachycardia but was eventually stabilised with amiodarone and disopyramide. Thyroid function tests showed raised thyroxine and normal triiodothyronine concentrations and a low level of thyroid stimulating hormone. These findings were attributed to amiodarone and she was started on carbimazole.

In 1988 she was admitted as an emergency after a cardiac arrest at home. She was investigated with a view to implantation of a defibrillator. Echocardiography showed a large intracardiac tumour in the right ventricle. This was confirmed by ventriculography, which also showed severe tricuspid incompetence and outflow tract obstruction with normal coronary arteries and normal left ventricular function. She was referred to our centre for surgery. It was decided to perform an endomyocardial biopsy first to identify the tumour; this showed only normal myocardium. Cardiac fibroma was considered a likely diagnosis and early surgery arranged. The patient, however, had a cardiac arrest and attempts at resuscitation were unsuccessful.

Necropsy findings The thyroid gland was normal. No nodules or tumour were identified. The important postmortem findings were confined to the heart. A small pericardial effusion was present. The heart weighed 580 g and was grossly enlarged with dilatation of the right atrium and right ventricular hypertrophy and dilatation. The right ventricle contained a rubbery oval mass 7.5 × 4 × 3.5 cm arising by a broad base from the interventricular cavity. The anterior tricuspid valve cusp was stretched around its posterior surface and its upper pole projected into the right ventricular outflow tract, causing almost complete occlusion. The mass was easily dissected off the underlying myocardium and the cut surface showed yellowish brown tissue with areas of fibrous scarring and some cystic change. Histological

Figure 1 Coronal section through the heart showing an ectopic thyroid mass arising from the right ventricular aspect of the interventricular septum and impinging on the outflow tract.

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Figure 2 Mature thyroid follicles bounded on one side by myocardium. (Haematoxylin and eosin.)

examination showed thyroid tissue in which most of the follicles were distended with colloid and lined by flattened epithelium, though a few microfollicular areas were also present. Foci of fibrous scarring, haemorrhage, and oncocyctic change were seen in some areas, but there was no histological evidence of hyperplasia or neoplasia. A thin layer of fibrous tissue was present at the smooth interface with underlying myocardium. The appearances were those of ectopic thyroid tissue showing some features of colloid storage goitre (fig 2).

Discussion

Ectopic thyroid tissue has been reported at many sites, from the tongue to the mediastinum; and cases of lingual thyroid are particularly well documented. By comparison, intracardiac thyroid tissue is a rarity with only six cases worldwide¹⁻⁶ (excluding this case).

The most logical explanation for rests of intracardiac thyroid tissue lies in the close proximity of the thyroid anlage to the heart in early embryological development. The thyroid gland takes its origin from an endodermal downgrowth of the floor of the pharynx. Initially this hollow tube, the thyroglossal duct, extends caudally before becoming solid at about the sixth week of embryonic life. Bilembryonation of the distal end of this tube gives rise to the primitive thyroid gland. Prolonged adherence of the thyroid primordium to the myocardial mantle at this stage apparently

results in subendocardial thyroid ectopia (at the other extreme, failure of contact with the developing heart produces a lingual thyroid²).

Interestingly, in all reported cases the thyroid tissue has arisen from the right ventricular aspect of the interventricular septum, giving rise to signs and symptoms of right ventricular outflow tract obstruction or conduction disturbances, or both, in all but one case. This was found incidentally at necropsy in a patient who died from a cerebellar haemangioblastoma.²

Four of the previous cases of intracardiac thyroid tissue have been successfully treated surgically.³⁻⁶ Removal of the tumour at necropsy in our case suggests that, had the patient come to surgery, the "tumour" could have been removed without disturbing the integrity of the right ventricular wall or the tricuspid valve. Owing to the benign nature of the lesion we believed that the outlook would have been good.

Abnormalities of thyroid function have been recorded in only one previous case.³ In this case histological examination of the "tumour" also showed changes of a colloid storage goitre. As there was no evidence of thyrotoxicosis histologically in our case, it seems likely that the abnormal results of the thyroid function test were correctly attributed to amiodarone.

The possibility that intracardiac thyroid tissue represents part of a teratoma has to be considered. Intracardiac teratomas have been reported,⁷ some of which contain areas of thyroid differentiation; but despite examination of multiple blocks in our case no elements other than thyroid tissue were noted. In only one of the other reported cases has tissue other than thyroid tissue been found and in that case the associated tissue was benign congenital polycystic tumour (mesothelioma of the atrioventricular node).⁵ Another possibility is that the intracardiac thyroid tissue represents a solitary metastasis from a very well differentiated follicular cell carcinoma of the thyroid. Despite macroscopic and microscopic examination of the thyroid gland at necropsy there was no evidence of neoplasia.

In summary, we report a case of ectopic intracardiac thyroid tissue showing features of a colloid storage goitre, in which the patient presented with ventricular tachycardias and signs of right ventricular outflow obstruction. We believe this to be the first such case reported in the United Kingdom.

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