Benign Congenital Intracardiac Thyroid and Polycystic Tumor Causing Right Ventricular Outflow Tract Obstruction and Conduction Disturbance

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A patient with a rare intracardiac tumor arising from the right side of the interventricular septum who developed conduction disturbances and symptoms of right ventricular outflow tract obstruction is reported. The patient was successfully treated with insertion of a permanent pacemaker and surgical removal of the tumor.

Primary benign cardiac tumors, apart from myxomas, are uncommon. Ectopic thyroid tissue in the heart is rare, with only three previous cases reported (1–3). Benign congenital polycystic tumors, although also rare, have been reported more commonly, especially as a cause of sudden death (4–7). We describe a patient who presented with dizziness on exertion and progressive conduction defects treated successfully by permanent pacemaker insertion and surgical removal. Study of the intracardiac tumor revealed both ectopic thyroid and benign polycystic tissue.

Case Report

History. A 57 year old woman presented with a history of atypical chest pain for 4 weeks, increasing shortness of breath for 6 months and dizziness for 18 months. A syncopal episode had occurred 3 months previously while the patient was squatting. There was a past history of mild hypertension and depression, and she had known of a systolic murmur for at least 4 years.

Physical examination. The blood pressure was 140/90 mm Hg and the pulse rate was 70/min. The jugular venous pressure was not raised. The pulmonary component of the second heart sound was soft. No parasternal impulse was noted. A grade 3/6 systolic ejection murmur increasing with inspiration and disappearing during the strain phase of the Valsalva maneuver was heard at the left sternal border. The thyroid gland was not enlarged.

Electrocardiogram. On admission it showed sinus rhythm with left axis deviation and T wave inversion in the inferior and lateral leads. Two days later first degree heart block (PR interval 0.24 s) and right bundle branch block developed. Seven days after admission, transient asymptomatic 2:1 atrioventricular (AV) block occurred. The chest X-ray film revealed a normal-sized heart and clear lung fields.

Two-dimensional echocardiography and cardiac catheterization. Echocardiography revealed a large mass in the right ventricular cavity attached to the upper part of the interventricular septum and protruding into the infundibulum (Fig. 1). At cardiac catheterization, a peak systolic gradient of 80 mm Hg was present between the mid-right ventricle and its outflow tract. Right ventricular angiography confirmed the presence of the mass in the right ventricle attached to the interventricular septum (Fig. 2). The left ventriculogram and coronary arteriograms were normal, apart from a small area of abnormal circulation arising from a septal branch of the left anterior descending artery. A dynamic computed tomographic scan also showed a mass in the right ventricular infundibulum, with normal appearance of the right atrium.

Treatment. In view of the progressive conduction defect, a permanent cardiac pacemaker was inserted with an endocardial lead positioned at the right ventricular apex. At
elective cardiotomy, an ovoid cystic tumor (approximately 3 x 2 cm) was found arising from the upper part of the interventricular septum (Fig. 3) below the tricuspid valve and protruding into the right ventricular outflow tract. It was dissected off the right ventricular septum and anterior leaflet of the tricuspid valve. The cut surface revealed dark brown viscous fluid.

Pathology. Microscopically the tumor was not encapsulated, and two distinct areas of change were seen. The wall of a large cyst was composed of colloid-containing follicular tissue with the morphologic features of normal

Figure 1. Two-dimensional echocardiogram in the four chamber subcostal view, showing the tumor (arrow) extending from the interventricular septum into the right ventricular outflow tract. LA = left atrium; RA = right atrium.

Figure 2. Right ventricular angiogram, with pacing wire and catheter in the right ventricle, demonstrating the large filling defect of the tumor (arrow) extending below the tricuspid valve and protruding into the right ventricular outflow tract.

Figure 3. Macroscopic appearance of the tumor showing the ovoid and cystic appearance.

Figure 4. A. Microscopic appearance of the ectopic thyroid tissue showing colloid-containing follicular tissue with the morphologic features of normal thyroid tissue (original magnification x 100, reduced by 34%). B. Microscopic appearance of the benign congenital polycystic tissue showing tubular spaces in fibrous stroma. The spaces are lined with cuboid cells and filled with amorphous periodic acid-Schiff-positive material (original magnification x 200, reduced by 34%).
thyroid tissue; thyroglobulin stain was positive (Fig. 4A).
Several small cysts were composed of various sized tubules lined with multilayered cuboid cells, some with a suggestion of a brush border on the luminal surface. The lumina were filled with amorphous periodic acid-Schiff-positive material and macrophages; the thyroglobulin and epithelial stains were negative (Fig. 4B).

Clinical course. Results of thyroid function tests performed postoperatively were normal. The postoperative course was uncomplicated, and the patient was discharged 7 days later. On review 12 months later the patient was well and had pacing rhythm. Moderate tricuspid regurgitation was noted. A repeat echocardiogram showed no evidence of tumor recurrence, and thyroid function tests remained normal.

Discussion

To our knowledge, this patient represents the first reported case of an intracardiac tumor containing both heterotropic thyroid tissue and benign congenital polycystic tumor. She presented clinically with a right ventricular tumor causing right ventricular outflow obstruction and progressive conduction defects. The diagnosis of intracardiac tumor was made by two-dimensional echocardiography which accurately defined the location, size and point of attachment of the tumor.

Pathology. The pathologic examination of the right ventricular tumor showed areas of both intracardiac ectopic thyroid tissue and benign congenital polycystic tumor. There have been three previous reports of a benign intracardiac thyroid mass, one found at routine autopsy (1) and two, as in this case, removed successfully at surgery (2,3). As in our case, in the three previous cases the tumor originated from the right side of the ventricular septum and caused right ventricular outflow tract obstruction. The presence of nonmalignant heterotropic thyroid tissue in the heart has been explained (1) as an abnormality of morphogenesis occurring at the time when there is a close relation between the thyroid primordium and the heart in the early human embryo.

In previous cases, benign congenital polycystic tumor was situated within or very near the AV node. The more popular name of this tumor is mesothelioma of the AV node. Other names have included benign coelothelioma and lymphangiendothelioma of the AV node (4–7). However, in view of the uncertainty of the cellular origin of the tumor, the descriptive term of James and Galakhov (6) seems more appropriate. Patients previously reported on have presented with long-standing complete heart block with narrow QRS complexes. Our patient presented differently, with progressive conduction defects including right bundle branch block and partial AV block. The differences in presentation may be explained by the different site of the tumor which in our case did not obviously involve the AV node but was attached to the upper part of the interventricular septum immediately below the anterior leaflet of the tricuspid valve. It is possible that one abnormality of morphogenesis could account for the presence of both the ectopic thyroid and benign polycystic tissue in one tumor if both the mesothelial tissue covering the embryonic heart and the primordium of the thyroid gland were trapped together in the interventricular septum as the embryonic heart folded on itself (1,4).

The successful treatment of this case demonstrates the importance of diagnosis of intracardiac tumors because, although rare, they are often surgically curable.

We are indebted to Lyndal Darkin for performing echocardiographic studies and Zora Marko for typing the manuscript.

References