TRICUSPID PAPILLARY FIBROELASTOMA CAUSING SYNCOPAL EPISODES

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Papillary fibroelastomas are rare, accounting for fewer than 10% of all primary cardiac tumors.¹ Most often they are asymptomatic, and detection is usually incidental during echocardiography, cardiac catheterization, cardiac operations, or at autopsy. Symptoms, if reported, are usually due to involvement of the left-sided valves; fibroelastomas of the right side of the heart are largely asymptomatic. We report here a case of acute syncopal episodes diagnosed to be due to papillary fibroelastoma of the tricuspid valve.

Case report. An 81-year-old woman had had two sudden, brief episodes of syncope while walking. Both were followed by complete, spontaneous recovery. Her medical history was noncontributory, and results of a physical examination and routine investigations including a chest x-ray film and an electrocardiogram were unremarkable. Two-dimensional echocardiography and transesophageal echocardiography (TEE) revealed normal-sized heart chambers with mild fibrocalcification of the aortic and mitral valves. A solitary round mass, 2 by 2 cm, was attached by a short stalk to an area beneath the anterior leaflet of the tricuspid valve, protruding into the right ventricle (Fig. 1). The mass was mobile from side to side. Tricuspid regurgitation was not present and right ventricular contractility was within normal limits.

Preoperative angiography revealed no coronary or carotid artery disease. At operation, performed with cardiopulmonary bypass and cold crystalloid cardioplegia, a single 2 by 2 cm polypoid tumor was found enmeshed in the chordae tendineae of the anterior leaflet of the tricuspid valve. The tumor had a furry, grayish white surface. There was no thrombus. With its stalk still attached, it extended to the midfundibular chamber. The whole mass along with the involved chordae was excised. Remaining chordae were reattached. No other mass was found in either the right atrium or ventricle, nor was any other abnormality seen; of particular note, there was no associated atrial septal defect or patent foramen ovale. TEE after bypass did not reveal any residual tricuspid regurgitation, and the postoperative period was uneventful.

Histopathologic examination of the mass showed villous papillary tissue consistent with an endocardial papillary fibroelastoma (Fig. 2). The patient had a smooth recov-

ery and did not have any further episodes of syncope during 10 months of follow-up after the operation. A second echocardiogram confirmed the absence of any intracardiac lesion and satisfactory tricuspid valvular function.

Discussion. Cardiac papillary fibroelastomas are uncommon, comprising only 8% of all heart tumors and occurring less frequently than myxomas and lipomas;² however, they are the most common primary tumor of the heart valves.³ These tumors are usually solitary, stalked, and small (less than 1 cm in diameter), affecting adults older than 50 years of age.¹ They may occur alone or in association with other forms of cardiac disease.

Most often these tumors do not cause symptoms or produce physical findings. Those symptoms that do occur are seen with fibroelastomas of the left side of the heart and include angina, myocardial infarction, transient ischemic attacks, varying degrees of stroke, and even sudden cardiac death. Embolization of tumor fragments or of platelet fibrin clots forming on the surface of the tumor may be responsible for these events.¹ In contrast, fibroelastomas of the right side of the heart, of which tricuspid tumors are the commonest, are less clearly related to symptoms. There have been far fewer reports of echocardiographic diagnosis in these,⁴ even though the right-sided heart valve lesions are probably as common as those on the left.² It may be that the usually asymptomatic right-sided lesions do not necessitate medical attention and are thereby underreported.

Only a few reports have been published in which clinical signs and symptoms could actually be attributed to tricuspid fibroelastomas. These have included a tumor "plop" on auscultation,⁵ intermittent right ventricular outflow tract obstruction with cyanotic episodes,⁶ pulmonary embolization,⁷ and congestive heart failure.^{3,8}

So far, however, there is no report in the literature of a tricuspid valve fibroelastoma in an otherwise healthy adult coming to a clinician's attention solely for the symptoms of syncope. Frumin and associates⁵ reported on a patient with near syncopal symptoms in whom a tricuspid tumor was discovered incidentally before bypass grafting; the authors could not attribute her syncope to the tumor. In our patient, however, inasmuch as there were no other cardiovascular or other lesions to explain the symptoms and there was no recurrence of syncope after the operation, the tricuspid tumor appears to be the culprit lesion. When evaluated during the operation, the tumor seemed capable of reaching into the right ventricular outflow tract, and this could have caused intermittent obstruction to the pulmonary blood flow and episodes of syncope. The possibility of the tumor causing paradoxical embolism to the brain was ruled out during the operation.

A diagnosis of tricuspid papillary fibroelastoma can be effectively made by two-dimensional echocardiography.

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Fig. 1. Two-dimensional echocardiogram showing the tumor arising from the tricuspid valve and protruding into the right ventricle.

Fig. 2. Histologic section of the tumor showing villosapillary tissue lined by a flattened layer of cells and a fibrocollagenous core.

Surgical excision is the treatment of choice in all cases, even for small and asymptomatic tumors, because (1) the operation carries a low risk and offers a chance for lasting relief inasmuch as these lesions have not been known to recur, (2) the threat of embolism is ever present, and (3) a preoperative echocardiogram may not reliably distinguish a papillary fibroelastoma from other intracardiac lesions such as myxomas or a vege-

tations. Patients should be given anticoagulants and antiplatelet drugs to reduce the risk of thrombus forming on the tumor surface. TEE can be useful intraoperatively in planning the operation, especially with regard to the adequacy of valvular repair. At operation, if the valvular involvement by the tumor is extensive, repair or even replacement of the valve may be necessary.

We conclude that clinicians should be aware of syncope as a possible symptom in the relatively uncommon condition of papillary fibroelastoma of the tricuspid valve. Diagnosis by echocardiography followed by surgical excision of the tumor should be advised in all cases.

REFERENCES

1. Shahian DM, Labib SB, Chang G. Cardiac papillary fibroelastoma. *Ann Thorac Surg* 1995;59:538-41.
2. McAllister H, Fenoglio J Jr. Tumors of the cardiovascular system. Atlas of tumor pathology, 2nd series, Washington, DC: Armed Forces Institute of Pathology, 1978;15:20-5.
3. Edwards FH, Hale D, Cohen A, Thompson L, Pezzella T, Virmani R. Primary cardiac valve tumors. *Ann Thorac Surg* 1991;52:1127-31.
4. Wolfe JT III, Finck SJ, Safford RE, Persellin ST. Tricuspid valve papillary fibroelastoma: echocardiographic characterization. *Ann Thorac Surg* 1991;51:116-8.
5. Frumin H, O'Donnell L, Kerin NZ, Levine F, Nathan LE Jr, Klein SP. Two-dimensional echocardiographic detection and diagnostic features of tricuspid papillary fibroelastoma. *J Am Coll Cardiol* 1983;2:1016-8.
6. Anderson KR, Fiddler FI, Lie JR. Congenital papillary tumor of the tricuspid valve: an unusual cause of right ventricular outflow obstruction in a neonate with trisomy E. *Mayo Clin Proc* 1977;52:665-9.
7. Neerukonda SK, Jantz RD, Vijay NK, Narrod JA, Schoonmaker FW. Pulmonary embolization of papillary fibroelastoma arising from the tricuspid valve. *Tex Heart Inst J* 1991;18:132-5.
8. Mohan JC, Goel PK, Gambhir DS, Khanna SK, Arora R. Calcified mobile papillary fibroelastoma of the tricuspid valve: a case report. *Indian Heart J* 1987;39:237-9.

MULTIPLE SMALL FENESTRATIONS CREATED DURING CONSTRUCTION OF THE TOTAL CAVOPULMONARY CIRCULATION: SUBSEQUENT COURSE AND SPONTANEOUS CLOSURE

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Fenestration of the baffle between the systemic and pulmonary venous atria at the time of construction of the total cavopulmonary circulation is reported to lower morbidity and risk of takedown of the connection and decrease mortality.¹⁻³ Cardiac output is maintained by right-to-left shunt across the fenestration and right atrial pressure is lowered, but with consequent persisting systemic desaturation.¹⁻³ A single fenestration has usually been created.¹⁻⁵ Fenestration size has most frequently been 3.5² to 4 mm,^{1,3,4} with a corresponding cross-sectional area of 9.6 to 12.6 mm², but has varied from 2.7 mm³ to 9 mm⁴ with cross-sectional areas of 5.7 to 63.9 mm². Transcatheter device closure¹⁻⁴ or surgical closure with a snare placed around the fenestration⁵ has been performed at variable times after the initial operation. Spontaneous closure of fenestrations has been noted,^{1,2} more commonly in the smaller fenestrations.³ We placed multiple small fenestrations with a total combined cross-sectional area similar to that resulting from a single 3.5 to

4 mm fenestration to achieve a similar oximetric and hemodynamic outcome, but in the hope that early spontaneous closure would occur, avoiding the need for further interventions.

Five patients aged 2.8 to 5.9 years had multiple small fenestrations created in the baffle at the time of total cavopulmonary connection. All operations were performed by the same surgeon. Fenestrations were created in the polytetrafluoroethylene membrane used for the right atrial lateral tunnel with the use of a 2.5 mm punch (Scanlan) or cut with a blade, approximately 3 mm apart, before the patch was incorporated in the right atrial closure. Postoperative mean right atrial pressures 24 to 48 hours after the operation and saturations at hospital discharge were recorded. The hospital stays were 7 to 19 days. All patients were evaluated electively 5 to 10 months after the procedure to establish the status of the multiple small fenestrations. Four patients were studied with transesophageal echocardiography and one patient with transthoracic echocardiography. Saturations with the patients breathing air were determined by transcutaneous pulse oximetry at follow-up.

Results are presented in Table I and include diagnosis and previous operation, fenestration size and number, postoperative right atrial pressures and saturations, and the status of the fenestrations and saturations at follow-up. Transesophageal echocardiography provided excellent imaging of the baffle wall permitting delineation of each of the small fenestrations when present. In patient 1 the

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