Case Reports

Cardiac Papillary Fibroelastoma Excision Combined with Reconstructive Surgery

Tengis TKEBUCHAVA, MD, Ludwig K. VON SEGESSER,1 MD, Augusto GALLINO,2 MD, Olaf DIRSCH,3 MD, and Marco I. TURINA, MD

SUMMARY

Three cases of cardiac papillary fibroelastomas are described. Two-dimensional echocardiography detected the tumors in the mitral valve, the cordae tendinae and in the apex of the left ventricle - a unique location. The tumor excisions were combined with bypass operation, mitral valve reconstruction, repair of cordae tendinae and Maze-procedure. The three patients are doing well after surgery. (Jpn Heart J 1997; 38: 457-462)

Key words: Papillary fibroelastoma, Left ventricular tumor, Two-dimensional echocardiography

Cardiac papillary fibroelastomas are rare tumors. They occur mainly on a valve1) but may appear in any chamber of the heart. The left ventricle, however, is extremely rarely involved. Usually the course is benign but can become symptomatic in the form of heart ischemia leading to myocardial infarct,2,3) cerebral embolization4,5) or even sudden unexpected death.1,6) That explains why before 1981 these lesions were diagnosed incidentally during autopsies or surgery. Only with the development of invasive investigation techniques and ultrasonography during the last decades has identification on the living patient become possible.

The cases discussed below reflect successful outcomes after excisions of papillary fibroelastomas combined with various reconstructive operations.

CASE REPORTS

Case 1: A 69-year-old man was examined due to bradycardiac atrial fibrillation. During the complete investigation electrocardiography showed atrial fibril-
Figure 1. a: Two-dimensional apical four-chamber view showing the tumor (arrow) in the left ventricle (RV = LV = LA = RA. Calibration on the left side of the fan corresponds to a distance of 1 cm). b: Apical long-axis view showing the tumor (arrow) adherent to the anteropapical wall of the left ventricle.

lation with a rate of 40/minute and monotopic premature ventricular contractions; ergometry revealed inferior ischemia. Two-dimensional transthoracic echocardiography demonstrated a stalked ball-shaped tumor 14 mm in diameter (Figure 1a, b) antero-apical in the apex of left ventricle. No hypokinesia in the area of the tumor connection was noted. The ejection fraction was normal. Coronary angiography detected a 50% stenosis proximal of the left anterior descending coronary artery. The computed tomogram showed a filling defect in the left ventricle. Diabetes mellitus, not insulin dependent, and systemic hypertension were the only recognized risk factors. The physical examination was normal. Since Interleukin-6 was not increased (3.5 pg/ml) a fibromyoma rather than a myxoma was suspected. The possibility of intraventricular thrombus was excluded as well, because hypokinesia was absent. The operation was performed with the aid of extracorporeal circulation and intermittent antegrade blood cardioplegic solution in the aortic root. Via left atrial incision, the tumor was identified on the apex of the left ventricle. The jelly-like mass was gently removed. Finally, a coronary bypass graft mammary artery to the left anterior descending coronary artery was performed. The histological examination showed papillary fronds consisting of a central core of dense connective tissue surrounded by a layer of loose connective tissue. Focally hyperplastic endocardial cells were
on the surface. The postoperative course was uneventful and the patient was discharged in good condition seven days later. One year after the operation the patient's condition is satisfactory.

**Case 2:** A 50-year-old man was investigated due to paroxysmal tachycardia during the last three years. Ambulant check-up revealed an intermittent atrial fibrillation. The physical examination contributed no further diagnostic information but the echocardiogram disclosed a 1.0 cm tumor attached to the mitral valve chordae. Coronary angiography showed intact vessels. After a preliminary
workup, surgery was performed with standard cardiopulmonary bypass, moderate hypothermia, and cold ante- and retrograde cardioplegic arrest of the heart. First, a Maze-procedure was performed as treatment of chronic atrial fibrillation. Thereafter an incision through the left atrium disclosed a myxomata-like tumor 1 cm in diameter (Figure 2). The tumor was excised and chorda tendinae repaired with Prolene 5–0. The mitral valve remained tight. Histological examination confirmed a papillary fibroelastoma (Figure 3). On the second postoperative day the patient developed an absolute arrhythmia which was converted into sinus rhythm by means of antiarrhythmic therapy. During the following days the rhythm remained stable so that no further therapy (amiodarone) was necessary. The five month follow-up was uneventful.

**Case 3**: A 31-year-old woman with unremarkable medical history presented with sudden onset of aphasia and right hand weakness. A cranial computed tomography disclosed a large ischemic area in the left temporoparietal region. An echocardiogram showed a 1.1 cm tumor attached to the anterior leaflet of the mitral valve. There were no cardiac symptoms. Neurologically the patient was adequate but a mild right facial droop, hemiparesis on the right side and aphasia were present.

Additional investigations both cardio-cranial and of other organs revealed no pathologies. She postponed surgical intervention and continued therapy at home with warfarin, as well as speech and physical therapy. Gradually her condition improved although residual neurological symptoms persisted.

She was operated on 11 months later. After a right anterolateral incision in the 5th intercostal space cardiopulmonary bypass was initiated. The left atrium was incised and an approximately 1 cm flat mass of tumor on the free edge of the anterior mitral leaflet was found. The tumor was entirely removed together with adjoining tissue of the valve. The valve defect was then repaired; valvuloplasty of both commissures completed the operation. A pathological examination identified a papillary fibroelastoma. The patient made an uneventful recovery; she was extubated the first postoperative day, and has been doing well for 10 months.

**DISCUSSION**

Papillary fibroelastomas are considered benign tumors since their course is often symptomless. Generally they are discovered in older patients. However, this lesion can be biologically malignant, and it may cause dangerous complications. Papillary fibroelastomas resemble a sea anemone consisting of multiple papillary fronds arranged on a stalk. These fronds consist of a collagen core surrounded by elastic fibers and loose connective tissue and covered by endocardial cells. Because of their papillary configurations and soft, fragile nature, they are a potential
CARDIAL PAPILLARY FIBROELASTOMA 461

source of ostium obturation of coronary arteries leading to ischemia or myocardial infarction.²,³ They also serve as a substrate for fibrin and platelet aggregation with subsequent coronary⁷ or cerebral embolization,⁴,⁵ when located in the left atrium or ventricle. When they originate in the right chambers, they can embolize to a pulmonary artery⁸ or even block the right ventricular outflow tract with cyanotic spells resulting.⁹ In the literature, some cases of sudden unexpected death are mentioned.¹,⁶ It is important to note that although the mass ranges from 0.1 to 4 cm, the tumors can cause complications independent of their size. Some authors state that the lesion represents only 1% of all primary cardiac tumors, while others claim 7%, and yet others contend 10%.⁵,¹⁰⁻¹² The vast majority of the tumors (90%) are located on valves¹ and are rarely located in the left ventricle. Until now, location in the left ventricle has been mentioned only in a few reports.⁴,⁷,¹³

In spite of the high pressure in this chamber, i.e. associated risk of tearing of particles, there was only one event of embolization in these three patients. Moreover, if this lesion has a thrombogenic origin,¹⁴ why is this location of the tumor so rare? It is interesting that Levinsky et al.² discussed the development of multiple papillary fibroelastomas after septal myomectomy. However, in spite of frequent surgical manipulation of the left ventricle, the left ventricle remains an uncommon place for papillary fibroelastomas. It is also striking that most patients with this tumor are over 50 years of age with an absence of coronary disease.¹,¹¹

There is scant information concerning surgical intervention and long-term results. Taking into consideration the unpredictable course of the lesion, we believe that operation is warranted at any age as soon as the diagnosis is established. Two-dimensional echocardiography is a reliable method for discovering this lesion. Additional cardiosurgical intervention, when necessary, is not contraindicated and results in good surgical outcomes.

REFERENCES

7. Mazzucco A, Bortolotti U, Thiene G. Left ventricular papillary fibroelastoma with coronary