

Aortic Valve Papillary Fibroelastoma

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Cardiac papillary fibroelastoma (CPF) is a rare primary benign cardiac tumor. Before the use of echocardiography, the lesion was identified at autopsy or incidentally during cardiac surgery. CPF is the third most common primary cardiac tumor after atrial myxoma and lipoma, and is the most common tumor of the valvular endothelium. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) permit diagnosis of the tumor in living patients. CPF may be the cause of

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Case report

A 76-year-old woman was referred to the authors' institution for further evaluation of difficulty of speaking and mental confusion with transient right hemiparesis. There was no history of either cardiac or neurological disease.

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On physical examination, the patient's blood pressure was 145/85 mmHg, and hematological values were within normal limits. The electrocardiogram showed left axis type with normal sinus rhythm, and a heart rate of 78 beats per min. Computed tomography of the head showed a hypodense lesion in the left basal ganglia.

Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) revealed the presence of an echogenic mass (ca. 5 × 8 mm) which was attached to the left coronary leaflet in the central area on the aortic side of the valve, without a stalk (Fig. 1). Differentiation of this mass as a tumor, a thrombus, or



Figure 1: Preoperative transesophageal echocardiography of the aortic valve.

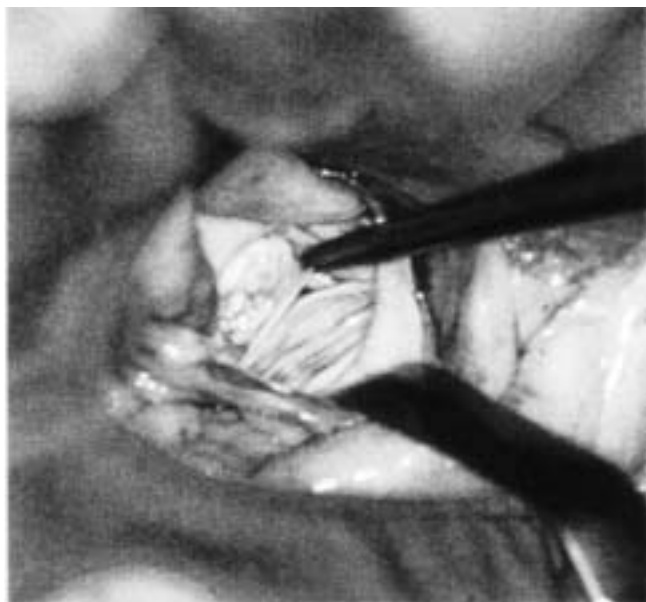


Figure 2: Aortic valve before resection of the fibroelastoma.

vegetation was not possible before surgery. The patient was treated with warfarin without resolution of the mass, and accepted for surgery with the presumptive diagnosis of papillary fibroelastoma. The operation was scheduled to be performed at six weeks after the last neurological event. However, during this waiting period the patient developed a new cerebral embolic ischemia under sufficient anticoagulation therapy; she had difficulty swallowing, and this necessitated the percutaneous implantation of a gastric feeding tube. Thus, urgent surgery was scheduled.

At surgery, a white, friable mass of 8 mm diameter was identified centrally on the aortic surface of the left coronary leaflet (Fig. 2). The mass was resected tangentially to the aortic cusp surface, the area of excision being treated with topical application of 0.65% glutaraldehyde for 5 min. Thereafter, the valve area was rinsed with generous quantities of 0.9% NaCl solution. The aortic leaflet was left intact (Fig. 3). After cardiopulmonary bypass, intraoperative TEE showed no aortic valve regurgitation.

Histological examination of the tumor showed a fibromatous core, surrounded by a dense mesh of elastic fibers and a loose connective tissue matrix. The outer layer consisted of regular endothelial cells. These findings were consistent with the diagnosis of papillary fibroelastoma.

The patient had an uncomplicated postoperative recovery and was discharged after 10 days. TEE showed no recurrent tumor or aortic regurgitation at 30 months postoperatively.

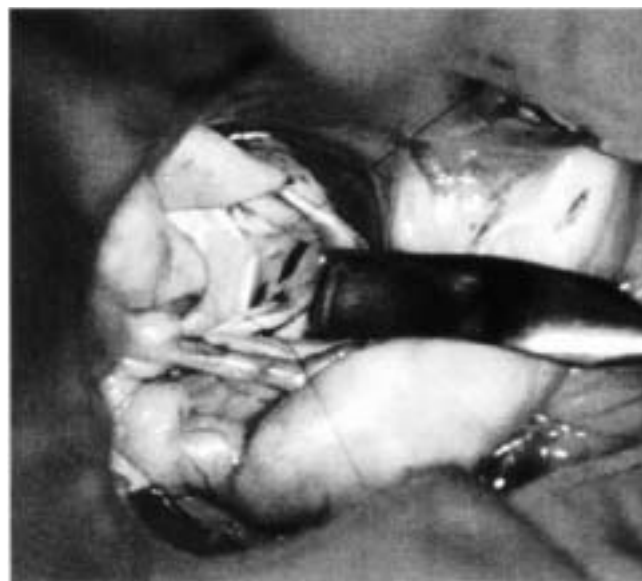


Figure 3: Aortic valve after resection of the fibroelastoma.

Discussion

Cardiac papillary fibroelastoma is a rare benign tumor, the etiology of which is unknown. The lesion has a valvular predilection, and arises most frequently from the valvular endocardium (1-3). It is usually isolated, but there may also be multiple sites (4). Diagnostically, echocardiography has been well proven to be effective in detecting CPFs, and this should therefore be the method of choice to search for these tumors. In this respect, TEE is superior to TTE as it delineates much more detail with regard to the size, shape, surface and mobility of the lesion (5-7). However, it is not possible to differentiate CPF from other lesions (e.g. endocarditis), including malignancies, by using echocardiography alone.

Initially, it was considered that these normally small tumors were not clinically significant, though several recent reports exist of serious symptoms and complications that might be attributed to tumor cells, to thromboembolism, or to myocardial ischemia (6,8). The evidence that CPFs can cause cardiac or neurological symptoms is now well established. Common presentations include angina, sudden death or neurological symptoms, such as transient ischemic attack and cerebrovascular accident. The embolic material may arise either from fragments of the tumor or from thrombus that, not uncommonly, forms around the tumor.

Upon diagnosis, preoperative anticoagulation therapy is prudent in order to reduce the risk of embolization. However, as shown in the present patient, this precaution does not eliminate the risk of emboli, which can still originate from the tumor itself.

The potential for life-threatening complications of left-sided CPFs is an indication for prompt surgical resection of the tumor, regardless of its size or shape. The native valve can often be preserved (1,8,9).

Most commonly, CPFs bear a short and thin stalk attached to a free leaflet edge, and this enables a relatively straightforward valve-sparing resection. CPFs lacking a stalk and situated in the center of a leaflet or cusp (as in the present patient) necessitate tangential resection from the adjacent valve tissue. As the aortic cusps are extremely delicate, the creation of a defect must be avoided, although a subtotal resection would leave residual tumor tissue. Denaturation with glutaraldehyde, which is a widely used surface fixative in biological cardiac implants, should eliminate the risk of tumor recurrence, even in the case of residual tumor cells. Following cardiopulmonary bypass, intraoperative TEE should be used to assess valvular function after tumor excision. Moreover, a pathological examination following surgical excision of the lesion is mandatory.

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