

An unusual case of papillary fibroelastoma “invading” the mitral valve

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We report the case of a 28-year-old patient who was admitted to an emergency department for left-sided motor deficit. The medical history showed multiple transient ischemic attacks over the past 2 years. Preoperative cerebral magnetic resonance imaging showed right sylvian infarct. Transthoracic echocardiography revealed a round, highly mobile, pedunculated 6- by 5-mm mass attached to the anterior mitral leaflet near the posterior commissure. Transesophageal echocardiogram showed similar images (Figures 1 and 2, arrows). This mass did not alter the mitral valve function. The ejection fraction was normal and no associated valve disease was found.

Clinical Summary

The patient was referred for cardiac surgery to achieve ablation of this clinically symptomatic mass. He was operated on successfully with moderate hypothermic cardiopulmonary bypass (33°C). We used a left atriotomy access to the mitral valve. Tumor was ablated and the mitral valve preserved. The gross appearance of the analyzed tumors showed a papillary fibroelastoma (PFE) that had a characteristic frondlike appearance. The postoperative period was uneventful and the patient was discharged at the 11th day with platelet antiaggregant therapy and regular echocardiographic follow-up. Histopathologic examination diagnosed PFE. Indeed, the histopathologic description of all the samples confirmed the diagnosis of PFE by showing the presence of specific fronds connected to a common pedicle. These latter structures were found to contain three dissociated levels: (1) a superficial endothelial layer surrounding the tumor, (2) an intermediate edematous and myxomatous layer, and (3) a central core with a concentric avascular fibrosis and mesenchymal cells.

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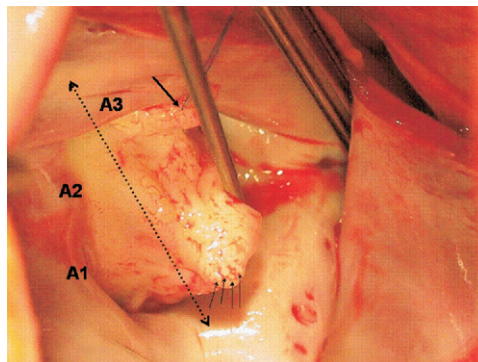


Figure 1. The broken line on the anterior leaflet marks the border between clear (normal) and rough (infiltrated) zones. The mitral valve showed infiltration of the anterior leaflet by multiple PFEs mainly on the free edge (discontinued arrows). A1, A2, and A3, Anterior mitral valve segments.

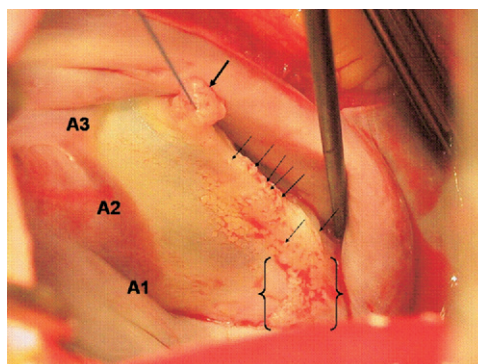


Figure 2. The biggest part of the mass (thick arrow) looked like an “active volcano,” and the small lesions had a “lava flow” shape (thin arrows and between brackets). A1, A2, and A3, Anterior mitral valve segments.

Discussion

Multiple PFEs are extremely uncommon.^{1,2} Indeed, they are usually single, small, and may be pedunculated. In our case, the mitral valve had an infiltrated anterior leaflet mainly on the free edge (Figure 1, discontinued arrow). The invasive characteristic of this unusual observation may be explained by the anatomic characteristic of the anterior leaflet. Indeed, it is divided into two zones, a rough zone and a clear zone. The rough zone is the thickest part; it is irregular, sometimes nodular, and corresponds to the area

where first- and second-order cords are attached to the underside of this area. In our case the rough zone was totally covered by the lesions. Operatively, the biggest part of the mass looked like an “active volcano,” and the small lesions had a “lava flow” shape (Figure 2). Given this unusual and unexpected diagnosis and the young age of the patient, we opted for exclusive tumor ablation without mitral valve replacement. Although the infiltration of valvular tissue by the PFE is impossible to prove inasmuch as the mitral valve was not extracted, the natural history of this tumor is unknown and preserving the native mitral valve could have been the optimal option. To the best of our knowledge this is the first case of invading PFE on the anterior leaflet of the mitral valve

reported in the literature. Clinicians should be aware of this PFE presentation to decide how to manage such patients surgically.

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Total arch replacement through a midsternotomy for a right-sided aortic arch aneurysm with an aberrant left subclavian artery

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The surgical approach for a right-sided aortic arch aneurysm with an aberrant left subclavian artery (ALSA) has to be considered carefully.¹⁻⁴ We have successfully performed total arch replacement for a right-sided aortic arch aneurysm with an ALSA through a midsternotomy alone in 2 consecutive cases. The present article describes the surgical technique we used.

Clinical Summary

Two male patients, aged 55 and 73 years, respectively, were referred to our institution within a period of 1 month for surgical repair of a right-sided aortic arch aneurysm. Both were asymptomatic. They weighed 72 and 65 kg. Multidimensional computed tomography showed a right-sided aortic arch with an ALSA. The aneurysm was located at the base of the ALSA (Kommerell's diverticulum). With regard to the arrangement of the arch vessels, a left common carotid artery arose from the aorta as the first branch, followed by a right subclavian artery, a right carotid artery, and the ALSA (Figure 1). No

concomitant congenital heart disease was identified by means of preoperative evaluation in either of the patients.

Surgical Technique

Through a midsternotomy, cardiopulmonary bypass was achieved with cannulation of the ascending aorta and right atrium. The first 3 arch vessels were easily exposed and taped. After initiation of core cooling, the ALSA was identified by means of palpation and divided from the left side of the anterior mediastinum (Figure 2, A and B). Circulatory arrest was established at a core temperature of 26°C. The aortic arch was incised, and antegrade selective cerebral perfusion was established. Four balloon-tip cannulas were inserted directly into the right subclavian artery, the right common carotid artery, and the left common carotid artery from inside the aortic arch and the transected ALSA from outside the aorta. A 20-mm Hemashield quadrifurcated graft (Boston Scientific, Natick, Mass) was used for reconstruction. While dissecting the distal arch, the right phrenic nerve was identified and taped to prevent injury. The descending aorta was completely transected, and open distal anastomosis was performed in an elephant trunk manner by using a short graft (Figure 2, C and D). The quadrifurcated graft was anastomosed to the short graft pulled out of the descending aorta. Reperfusion and rewarming were started through the femoral artery. Then the proximal end of the quadrifurcated graft was anastomosed to the aortic root, and the aorta was declamped. Finally, the arch vessels were independently reconstructed by using, in order, the branches of the graft, anastomosis of the right common carotid artery, and the right subclavian artery. Then the left common carotid artery and the left subclavian artery were reconstructed with the same branch of the quadrifurcated graft in an end-to-side and end-to-end manner, respectively. The postoperative course was uneventful in both cases.

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