

## Radical and Harmless Shave Resection of Atypical Papillary Fibroelastomas of the Cardiac Valves

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### ABSTRACT

**Background:** Papillary fibroelastomas (PFEs) are rare cardiac tumors usually treated by shave resection. Up to 20% of the patients are actually denied such a conservative surgical approach because atypical morphology is thought to preclude radical and effective outcomes.

**Methods/Results:** Surgical tricks and tips of shave resection are elucidated, and a case series presentation of the morphology of atypical PFEs treated by shave resection is described.

**Conclusion:** In experienced hands surgical shave resection is the gold standard for the treatment of PFE even in atypical presentations.

### INTRODUCTION

Papillary fibroelastoma (PFE) is a benign tumor with multiple papillary fronds resembling a sea anemone most commonly found at the endocardium of the cardiac valves; it accounts for 7% of cardiac tumors [Howard 1999].

Almost like pathogenesis, epidemiology remains largely uncertain. Cardiac fibroelastomas occur sporadically and exhibit a wide distribution of age with predominance between the fourth and eighth decades of life [Parthenakis 2009], though in some cases they may develop in infants [Hindupur 2005]. Their true incidence is largely unknown, and sex distribution is controversial [Sun 2001].

Although a PFE is benign in nature, it mandates surgical treatment because of both its friability and valve location. Indeed, it has a definite potential for embolization into the coronary or cerebral arteries resulting in devastating complications such as myocardial infarction or stroke. Current indications for surgical treatment are presence of cardiac or cerebrovascular symptoms or larger dimensions in asymptomatic cases.

Shave surgical resection is usually the standard treatment with excellent long-term results because recurrence has been

seldom reported [Sun 2001; Hynes 2002]. Up to 20% of PFEs seem not to be suitable for shave resection and undergo more extensive procedures [Westhof 2007; Gopaldas 2009].

Here we describe 3 cases of atypical PFEs surgically treated with shave resection. Mid-term follow-up results demonstrate that this procedure is both radical and harmless in these cases.

### CASE REPORT

#### *Surgical Technique*

Preoperative evaluation was always achieved with combined transthoracic and transesophageal echocardiographic approaches for a more comprehensive and accurate assessment to define localization and relationships with adjacent structures. Shave resection was performed through a median sternotomy on normothermic cardiopulmonary bypass, with standard aortic cannulation and right atrial or bicaval venous drainage under warm blood cardioplegic arrest. Optical magnification  $\times 3.5$  and surgical tools for microsurgery (ie, sharp point blade) were mandatory equipment requirements. Maximal accuracy was indeed devoted to discriminate between the tumoral outer surface and the endocardial layer in order to achieve both radical PFE resection and optimal preservation of the structural integrity of the valvular apparatus. Intraoperative transesophageal echocardiography was always performed in order to evaluate the surgical results soon after cardiopulmonary bypass weaning.

#### *Case 1*

A 69-year-old hypertensive man was referred for recent onset of exertion angina and a positive stress test. Transthoracic echocardiographic evaluation disclosed a round, mobile, echodense pedunculated mass of nearly 0.5 cm  $\times$  0.6 cm adherent to the ventricular side of the left coronary aortic cusp; the aortic valve was competent, and left ventricular function was globally and regionally preserved. Transesophageal echocardiography confirmed such a diagnosis. Coronary angiography disclosed total occlusion of the circumflex at the ostium and absence of other significant stenoses. On surgical inspection, the aortic valve proved normal on its vessel side, but on the ventricular aspect there was a long, mobile pedunculated mass firmly adherent through a large stalk to the midportion of the left coronary

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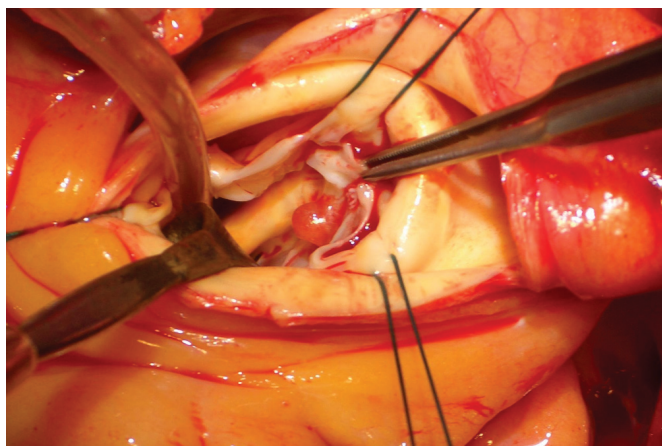


Figure 1. Intraoperative view in Case 1.

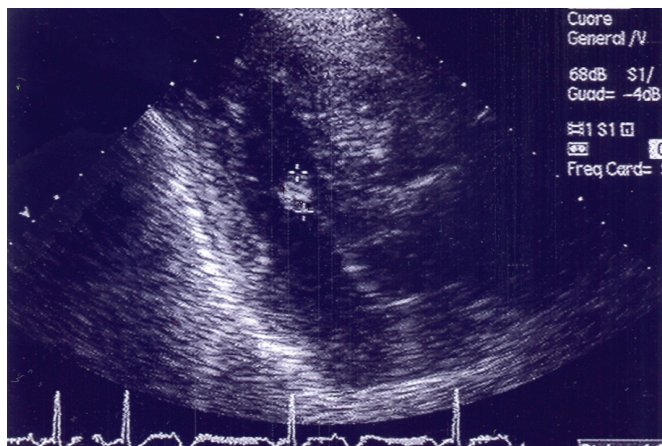


Figure 3. Echocardiographic view in Case 2.

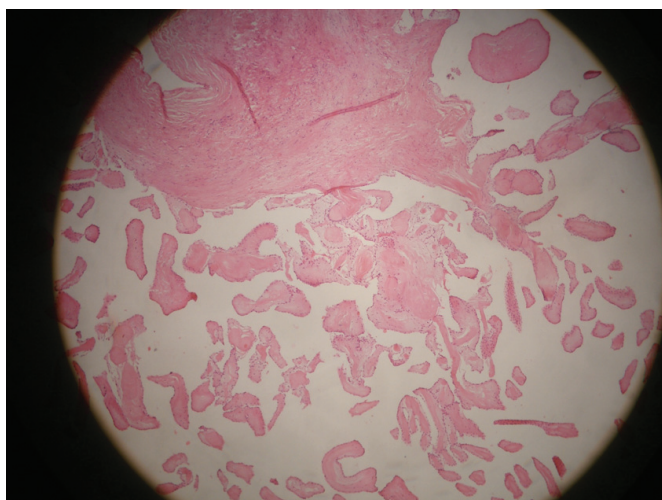


Figure 2. Pathology specimen in Case 1.

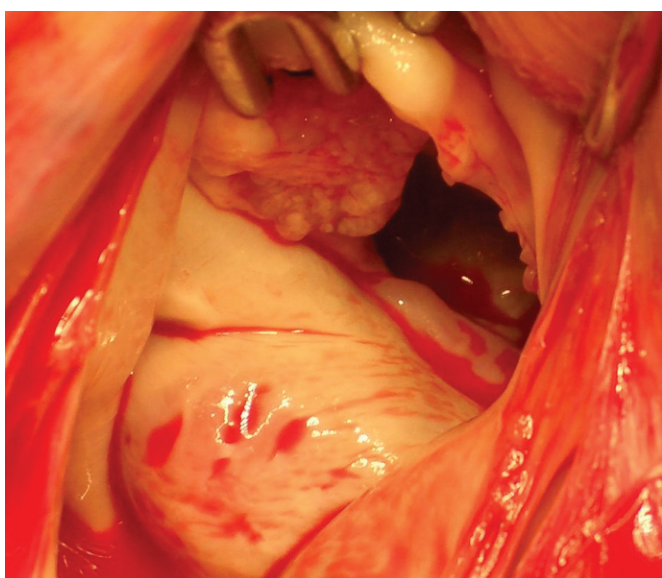


Figure 4. Intraoperative view in Case 2.

cuspid (Figure 1). Shave resection was performed, and great care was employed in order to prevent the highly friable mass from fragmentation and embolization. A vein graft was also placed on a first obtuse marginal branch. Aortic valve competence was first tested with a hydrodynamic test and then confirmed by transesophageal echocardiography after cardiopulmonary bypass weaning. The postoperative course was uneventful, and the patient was discharged on the sixth postoperative day. The pathology examination of the surgical specimen confirmed the diagnosis of PFE (Figure 2). At 12-month follow-up, the patient was asymptomatic with an active lifestyle, and echocardiography showed no recurrence and normal aortic valve function.

### Case 2

A 49-year-old hypertensive woman presented with an occasional echocardiographic finding of an ovoid mass in the right atrium. A subsequent nuclear magnetic resonance spectroscopy, prescribed by her family physician, disclosed a peduncolated mass adherent to the ventricular aspect of the anterior leaflet of the mitral valve of nearly 1 cm × 1 cm. Transesophageal

echocardiography at admission confirmed the diagnosis of PFE, the mitral and aortic valves were competent, and left atrial and ventricular dimensions and function were normal (Figures 3 and 4). Shave resection was performed taking care not to damage the chordae because the papillary projections of the tumor were firmly adherent. After nearly 20 minutes of aortic cross-clamping, a radical excision was achieved and, as demonstrated by intraoperative transesophageal echocardiography, the integrity of the mitral valve apparatus was preserved. The patient was discharged on the sixth postoperative day, and at 12-month follow-up she was free from recurrence with no trace of mitral regurgitation. Gross pathologic examination and microscopy confirmed the diagnosis of PFE.

### Case 3

A 59-year-old woman was referred for diastolic aortic murmur during the follow-up period after surgical treatment



Figure 5. Gross pathology in Case 3.

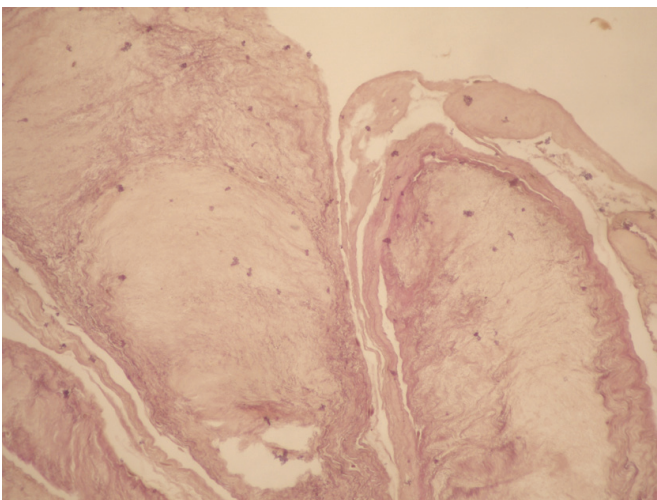


Figure 6. Microscopic pathology in Case 3.

of thyroid carcinoma. The patient also presented with chronic hepatitis B, arterial hypertension, and chronic obstructive pulmonary disease. Echocardiographic assessment revealed an echo-dense peduncolated round formation 12 mm in diameter located on the aortic side of the right coronary cusp. Left and right ventricular function was normal. A mild to moderate degree of aortic regurgitation was disclosed. Coronary angiography was negative. At surgical inspection, the tumoral mass was firmly adherent to the middle aspect of the free edge of the right coronary cusp with a large stalk (nearly 8 mm). Such a mass caused the involved aortic cusp to prolapse into the left ventricular outflow tract. Shave resection was performed within 10 minutes of aortic cross-clamping. Aortic regurgitation proved less than trivial at intraoperative echocardiographic assessment. The patient was discharged on the sixth postoperative day and, at 6-month follow-up, there is neither recurrence nor aortic regurgitation. Pathology confirmed the diagnosis of PFE (Figures 5 and 6).

## DISCUSSION

The rationale for surgical treatment of PFEs by means of shave resection is based on their peculiar pathology [Ahuja 2008]. They indeed display a sea anemone morphology with a superficial endothelial layer covering a fibrovascular core rich in fibroblasts with a variable amount of macrophages and dendritic cells. Such tumors are usually full fluttering within cardiac chambers because they are attached to the valvular endocardium only by a small based thin pedicle. Shave resection is deemed not applicable in those atypical cases in which sessile tumor or larger dimensions are present and/or the papillae are firmly stuck to the cusps or subvalvular endocardium. In these cases, more demanding procedures have been reported ranging from partial valve-sparing procedures with or without pericardial patching to prosthetic replacement [Ngaage 2005; Westhof 2007]. The case series reported here argue in favor of shave resection even in these more complicated settings. Severe aortic adherence through a large stalk, cusp prolapse due to “mass effect,” and extensive mitral chordate involvement were indeed effectively treated with this conservative approach. Meticulous radical resection with microsurgical tools under visual magnification helped preservation and even restoration of valve function as demonstrated by echocardiographic findings. Additionally, follow-up data confirmed good long-term performance of treated valves and disclosed also absence of recurrence. In view of such data, we conclude that shave resection, in experienced hands, is a safe and effective approach also in atypical PFEs.

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