CARDIAC VALVULAR PAPILLARY FIBROELASTOMA: AN USUAL CAUSE OF ACUTE ISCHEMIC STROKE.

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ABSTRACT

Cardiac tumors are uncommon, and valvar papillary fibroelastomas are considered relatively rare. Most of them are asymptomatic, but they are a potential source of systemic emboli, stroke, myocardial infarction and sudden death. We report tow cases of cardiac valvular papillary fibroelastoma diagnosed in the course of acute ischemic strokes. Surgical excisions of the tumors were successfully performed in the tow cases and the postoperative courses were uncomplicated. We will discuss the clinical, histological and therapeutic aspects.

Keywords: Heart neoplasms, Heart valve diseases, Papillary fibroelastoma

INTRODUCTION:

Papillary fibroelastomas (PFE) are endocardial tumors which represent only 16 % of all cardiac tumors [1]. Despite its benign nature, fibroelastomas can take dramatic events because of its valves and embolic complications. We report tow cases of cardiac papillary fibroelastomas collected at the cardiovascular department of Ibn Sina hospital-Rabat; Morocco, between January 2008 and January 2013 and we will review the literature.

CASES REPORTS:

Patient 1: A 36-year-old man without cardiovascular risk factors who presented a capsulolenticular ischemia .Etiological check-up showed a mass of the anterior mitral valve. The patient was referred to our department. The Neurological examination revealed a soft left facial hemiparesis. Cardiovascular examination was unremarkable. The ECG recorded sinus rhythm .Holter –ECG recording did not reveal any rhythm or conduction disorders. The chest X-ray and the biological check-up were normal. The transthoracic echocardiography showed a fluctuant echogenic mass, sitting at the atrial side of the anterior mitral valve, measuring 10 mm (Fig1A).

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Figure 1: (A) Transthoracic echocardiography showing a fluctuant tumor above the anterior mitral valve. (B) Transthoracic echocardiography showing a mass in the ventricular side of right coronary cusp. (C) Operative view showing the mass in the ventricular side of right coronary cusp. (D) Operative specimen less than 10 mm.

The transesophageal echocardiography confirmed the same data of the transthoracic echocardiography and eliminated the possibility of a left atrial thrombus.

**Patient 2:** A 62-year-old man who presented aphasia was transferred to our department for PFE. The transthoracic echocardiography revealed an echogenic mass measuring 6 mm on the ventricular side of the aortic valve (Fig 1B). The ECG recorded sinus rhythm. Holter–ECG recording did not reveal any rhythm or conduction disorders. The chest X-ray and the biological check-up were normal. Preoperative transesophageal echocardiography confirmed the same characteristics of the tumefaction. The two patients were operated by limited sternotomy under a total cardiopulmonary bypass in normothermia. In the first case the tumor was resected with the implantation base and the defect was repaired by simple suture. In the second case the tumor was removed by simple shaving from the ventricular side of the aortic valve (Fig 1C and 1D).

In the two cases the histologic examination of the surgical specimen revealed a papillary fibroelastoma (Fig 2).

**Figure 2:** Histological section, appearance of papillary fibroelastoma: endothelium-coated rich in extracellular matrix, collagen and elastin (hematoxylin-eosin, 10× magnification).

The postoperative course was simple and the patients were discharged after eight days and were doing well 6 months later with a normal echocardiography. The two patients were followed respectively for 5 and 3 years and are without recurrence.

**DISCUSSION**

Primary tumors of the heart are less than 5% of all cardiac tumors [1]. Papillary fibroelastoma is a benign endocardial tumor representing 16% of benign tumors of the heart [2]. It is the second most frequent benign cardiac tumor after myxoma and the first common valve tumor [3]. Its pathogenesis...
is not yet well understood, nevertheless cruric origin (wall thrombus) organized was suspected. [4]

Macroscopically, the tumor presents aspect of fern leaves. Underwater the tumor presents a classical appearance of a sea Anemone [5]. Its diameter does not exceed 1 cm in 83%.

It can sit in any valve. The ventricular side of the aortic valve and the atrial side of the atrioventricular valves are the most commonly affected: 29 % in the aortic valve, 25 % in the mitral valve, tricuspid and pulmonary valve are also reached, respectively in 17 and 13 % of cases [6]. It rarely develops in the papillary muscle, chordae tendineae or endocardium [6].

Histopathology of the tumor shows endothelium overlying the axes of loose connective tissue around a central axis of collagen or elastic fibers [7].

Although asymptomatic in most cases, the fibroelastoma may present general clinical expression as follows: fever, fatigue or takes an alarming event, particularly by valvular dysfunction or systemic embolism [1]. In fact the most fibroelastomas are often diagnosed incidentally [8]. In symptomatic patients. The most common clinical presentation is stroke or transient ischemic attack caused by cerebral emboli, originating from a thrombus on the tumor, or by a piece of the tumor itself [8]. In our tow cases the clinical presentations were an acute ischemic strokes. The transthoracic echocardiography orients the diagnosis but the transesophageal echocardiography can establish the diagnosis by showing an echogenic swelling center (center collagen) eliminating a myxoma, vegetations or thrombus [1, 7]. However, the diagnostic is based on histological examination of the surgical specimen.

Because of the potentially high risk of embolism, several authors think that surgery is indicated even if the patient is asymptomatic and whatever the size of the tumor [1, 7]. Benign nature and the absence of recidivism in the literature encourage the conservative surgery by limited excision. Because of the high potential risk of embolism recurrence our tow patients were successfully operated.

CONCLUSION

Cardiac papillary fibroelastoma is often revealed by alarming clinical symptoms specially stroke or transient ischemic attack. The transeosophageal echocardiography is useful, the positive diagnostic is histological and surgical excision is usually recommended.

REFERENCES


