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Papillary Fibroelastoma of the Right Coronary Cusp

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ABSTRACT

Cardiac papillary fibroelastoma (CPF) is a pathologically benign cardiac tumor. This tumor usually arises from cardiac valves, and it is the most common cardiac valvular tumor. This usually single and pedunculated tumor should be surgically treated when the mass is mobile and the patient has experienced a prior stroke, even if the stroke has manifested as a transient ischemic attack. Surgical treatment is definitive, and no recurrences have been reported in the literature. We describe a patient who had a diagnosis of CPF while undergoing investigation for a cryptogenic stroke.

INTRODUCTION

Cardiac papillary fibroelastoma (CPF) is a rare, benign cardiac tumor that usually arises from cardiac valve leaflets. Such tumors are the third most common primary tumors of the heart after myxomas, which are the most common, and lipomas [Granger 2005; Hino 2007]. CPF is a histologically characteristic lesion with a hitherto unknown histogenesis. CPF has become more frequently diagnosed as cardiac-imaging techniques and their availability have advanced. CPF constitutes approximately 10% of all cardiac tumors and has an estimated incidence of approximately 0.021% to 0.33% in autopsy series [Al-Mohammad 1998; Granger 2005; Biçer 2009]. Because these tumors are mostly asymptomatic, they are usually diagnosed accidentally when a patient is undergoing investigations for a separate pathology. These seemingly benign tumors have a tendency to cause stroke via secondary thrombus emboli or infarction due to coronary malperfusion via obstruction of coronary ostia when located on the left side of the heart, which is the usual location [Al-Mohammad 1998; Granger 2005; Hsu 2006; Sydow 2008].

Sometimes the presentation is unusual, such as a sudden loss of vision because of occlusion of a branch of the retinal artery, splenic infarction due to an embolus from an aortic valve lesion, or hemodynamic compromise [Crestanello 2002; Ergun 2007; Gilbert 2009].

In our report, we describe a patient who received a diagnosis of CPF while undergoing investigation for a cryptogenic stroke.

CASE REPORT

A 71-year-old female patient was referred to our clinic in April 2009 as she was being investigated for the cause of a cerebrovascular accident causing left hemiparesia. The symptoms had receded, leaving only a residual speech difficulty and slight loss of motor force in the left extremities. The cerebrovascular event had remained incomplete. Her magnetic resonance imaging results were unremarkable except for cerebral and cerebellar atrophy, which was evaluated as concordant with her age. The patient's vascular magnetic resonance angiogram was normal; however, a spiral computed tomography scan showed areas of ischemic change in bilateral

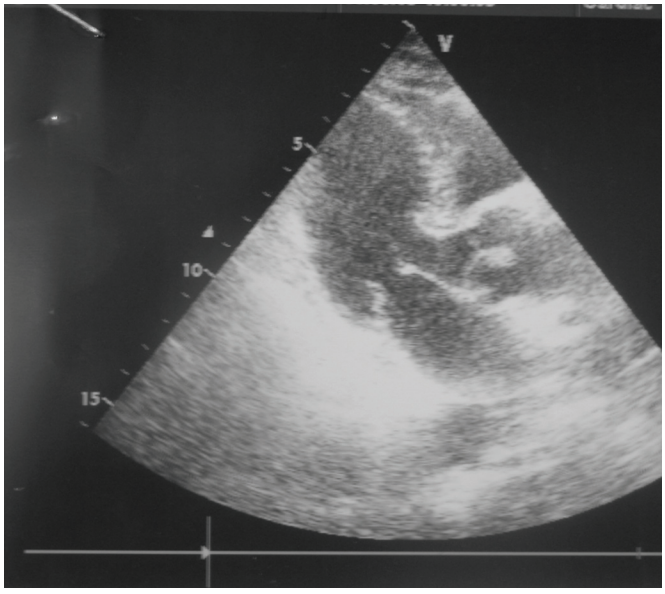

Figure 1. Echocardiographic image of the fibroelastoma.

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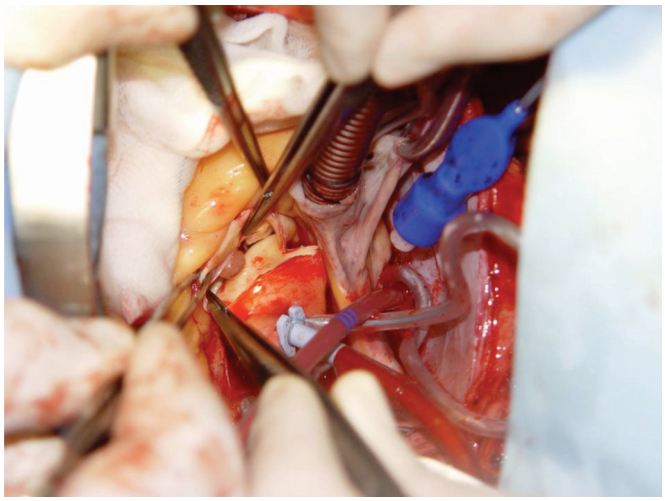

Figure 2. Surgically exposed right coronary cusp papilloblastoma.

periventricular white matter. The electroencephalogram showed signs of lateralization.

The unspecific findings led to a thorough cardiovascular evaluation. Her transthoracic echocardiogram revealed a mass on the aortic valve. Her preoperative echocardiogram had not revealed any regurgitation. The mass was round, mobile, and 0.5 cm in diameter. It was located centrally on the coapting edges of the aortic valve (Figure 1). Because of the possibility of concomitant coronary artery disease, the patient underwent coronary angiography. Her coronary arteries appeared normal.

The possibility of a renewed stroke because of the mobility of the valvular mass was a strong indication for surgical intervention. The patient underwent open heart surgery with a standard midline sternotomy and cardiopulmonary bypass (CPB) with single 2-stage cannulation and mild systemic hypothermia (33°C). The aortic cross-clamp time was 19 minutes, and the total CPB time was 36 minutes.

Upon entering the aorta, we observed a mass of 1 × 1 cm and gelatinous consistency that was located on the free edge of the right coronary leaflet (Figure 2). The mass was shave-excised, thereby sparing the integrity of the valve. The valve was examined with respect to the presence of aortic regurgitation, and the valve leaflets were observed to be competent.

After an uneventful recovery, the patient was discharged on the fourth postoperative day with a prescription of 100 mg/day of acetylsalicylic acid. Her histopathologic specimen was diagnosed as papillary fibroelastoma of the aortic cusp. No further therapy was recommended. The patient was placed on outpatient follow-up.

DISCUSSION

Cardiac valve tumors constitute only approximately 8% of all cardiac tumors [Sankar 1999]. CPF is the most common cardiac valve tumor. This tumor is an entity that all cardiac surgeons should know about.

Two peculiarities make CPF tumors interesting. First, these tumors are often asymptomatic, but although benign, they may cause fatal complications when undiagnosed. Second, when they become symptomatic they can mimic many diseases [Al-Mohammad 1998] because they cause end-organ malperfusion, with stroke, myocardial infarction, and intestinal ischemia being leading causes of morbidity and mortality [Granger 2005; Hsu 2006; Sydow 2008]. Unusual presentations also exist. They may cause occlusion of retinal artery branches leading to vision loss [Ergun 2007], splenic infarction [Gilbert 2009], acute valvular dysfunction, or sudden death.

These tumors are small, usually <1 cm in diameter; they are often solitary [Sankar 1999]. They usually arise on the valvular endocardium and are usually located on the left heart, mainly on aortic or mitral valves [Yerebakan 2009]. Although morphologically specific, they are histogenetically indistinct. The histologic characteristics consist of multiple papillary fronds, which distinguish this tumor from myxoma [Al-Mohammad 1998]. Its macroscopic features resemble those of sea anemones, especially when the tumor is immersed in saline [Sun 2001].

The importance of surgery is derived from 2 facts. The first is the importance of the presence of an index of suspicion in diagnosis, and the second is the fact that when diagnosed, these tumors usually present with stroke. Therefore, a diagnosis of CPF should always be in mind in the differential diagnosis of cryptogenic stroke.

Our patient is a good example of patients who present with a cerebral infarct and who undergo operation with valve-preserving shave excision to enable total preservation of valve function. Another point to emphasize concerns the surgical procedure. In some cases, a second lesion is present; therefore, a complete inspection during the operation is necessary to ensure that a second lesion is not missed.

Surgical treatment therefore should be chosen for all symptomatic patients and all asymptomatic patients with pedunculated lesions or masses >1 cm in diameter [Gopoldas 2009]. As for the transition period to surgery in unstable patients, anticoagulation should be recommended only for patients at high surgical risk [Sastre-Garriga 2000].

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