Ischemic stroke in a young woman with aortic papillary fibroelastoma: echocardiographic diagnosis and surgical excision

Giorgio Locci, Andrea Pili*

*Cardiology Unit, Businco Hospital, Cagliari, *Cardiology Unit, S. Barbara Hospital, Iglesias (CA), Italy*

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Papillary fibroelastoma is a benign cardiac tumor typically attached to the cardiac valves. The tumor is recognized during life often in patients evaluated for embolic events of unclear etiology, but sometimes it is recognized in totally asymptomatic patients. We describe the case of a papillary fibroelastoma involving the aortic valve in a 27-year-old woman who presented ischemic stroke. The diagnosis was obtained at two-dimensional echocardiography and confirmed at transesophageal echocardiography. The tumor was located in the non-coronary aortic cusp, without aortic regurgitation. No other embolic source was identified. The tumor was surgically removed. The resection was curative by complete surgical excision, without damage of the aortic valve. The biopsy confirmed the diagnosis. The clinical course was uneventful. This case is an example of a primary intracavitary tumor recognized in a young woman with unclear embolic event. Two-dimensional echocardiography proved to be the exam of choice for the early diagnosis of cardiac tumors.

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Address:

Dr. Giorgio Locci

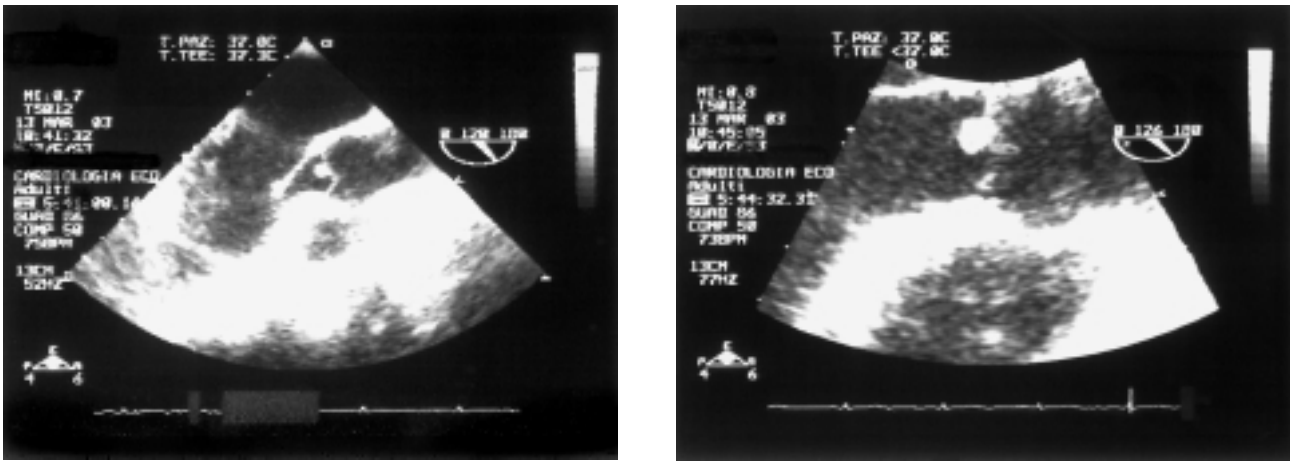
Servizio di Cardiologia
Ospedale Businco
Via Jenner
09121 Cagliari
E-mail:
cardiologia.businco@tiscali.it

Introduction

Papillary fibroelastoma is a rare small benign cardiac tumor with multiple papillary fronds resembling a sea anemone and usually attached by a short pedicle, either single or multiple, ranging from 0.1 to 4 cm in size. This tumor is common in older subjects and accounts for about 7% of primary cardiac tumors. Papillary fibroelastoma may occur typically on cardiac valves, or on the papillary muscle, chordae tendineae, or endocardium. Physical findings are frequently absent with these tumors. Clinical features of cardiac papillary fibroelastoma are variable and related to the involved cardiac component. They include arterial and pulmonary embolism, valvular heart disease, syncope, congestive heart failure, immunological disease, and sudden death[1-3]. Because of their potential for cerebral and coronary embolization, even small papillary fibroelastomas should be excised in the absence of major surgical contraindications. In this report we describe the clinical and echocardiographic findings of a young woman with aortic papillary fibroelastoma who presented ischemic stroke.

Case report

A 27-year-old female patient was diagnosed to have an ischemic stroke in June 2002. The patient reported a medical history of miscarriage. Serological exams, urinalysis, chest X-ray, and ECG were normal. Cultures were negative. Hematological analysis revealed by ELISA a slight elevated titer of anticardiolipin antibodies of IgM isotype; lupus anticoagulant test was negative. A two-dimensional echocardiography was performed which showed suspected endocarditis vegetations in the aortic and mitral valves. Transesophageal echocardiography showed a single mobile pedunculated mass attached to the non-coronary aortic cusp found on the ventricular side of the cusp (4 × 3 mm in diameter) (Fig. 1). Color flow Doppler study of the aortic valve was normal. Transesophageal echocardiographic examination of the mitral valve was normal. The suspected endocarditis vegetation at two-dimensional echocardiographic examination was interpreted as an artifact. Thus, in view of the previous history of ischemic stroke, the presence of antiphospholipid antibodies, and the presence of suspected valvular vegetation, the diagnosis of suspected endocarditis associated with antiphospholipid syndrome and ischemic stroke was made. The patient was discharged from the hospital and received oral anticoagulant therapy. Afterwards the diagnosis of antiphospholipid syndrome was not



A B

Figure 1. A: *transesophageal echocardiography performed before surgery showing, in the non-coronary aortic cusp on the ventricular side of the cusp, a solid mass attached by a short pedicle.* B: *detail of the aortic mass at transesophageal echocardiography.*

confirmed by additional immunohistochemical studies. In fact after 8 weeks, the lupus anticoagulant test was negative, anti-beta$_2$ glycoprotein I antibody and anticardiolipin antibody measurements were negative. Then transesophageal echocardiography was repeated and showed no changes in the aortic valve lesion. This mass was interpreted as a papillary fibroelastoma and the patient continued oral anticoagulants, and excision of the cardiac lesion was planned. On May 31, 2003, the patient underwent heart surgery for removal of the aortic mass. The histologic exam resulted in papillary fibroelastoma. The postoperative course was uneventful. The patient was discharged from hospital without oral anticoagulants. After 18 months the clinical course is uneventful and the patient is in good clinical conditions. Two-dimensional echocardiography is normal without tumor recurrence (Fig. 2).

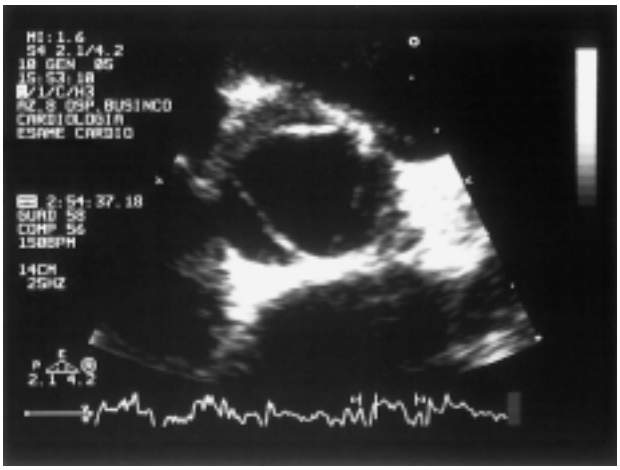


Figure 2. *Two-dimensional echocardiography performed 18 months after the resection of the aortic mass showing a normal aspect of the aortic valve without tumor recurrence.*

Discussion

Papillary fibroelastoma is a rare small benign cardiac tumor with multiple papillary fronds resembling a sea anemone and usually attached by a short pedicle. This tumor is common in older subjects (mean age 60 ± 16 years) and accounts for about 7% of primary cardiac tumors[1,2]. Papillary fibroelastoma may occur on cardiac valves, typically on the aortic and mitral valves (approximately 90%), or on papillary muscle, chordae tendineae, or endocardium[1,3]. Single (91%) or multiple (9%) lesions have been identified ranging from 0.1 to 4 cm in size. Most are < 1 cm in diameter[1-3]. Some papillary fibroelastomas are congenital[4]. Histologically the tumor consists of fibrous tissue covered by an elastic membrane, which in turn is covered by the endocardium[2]. Clinical features of cardiac tumors are variable and related to the involved cardiac component. Fibroelastomas of the left side of the heart have been associated more frequently with severe symptoms. Common presentations include angina or sudden death or neurologic symptoms, such as transient ischemic attacks and cerebrovascular accidents[5-8]. Cerebrovascular symptoms have been described frequently[8-10]. Tumor embolization may be responsible for some of these cerebrovascular events, but several authors also have suggested that these tumors may lead to subsequent embolization of a platelet-fibrin clot[7,10,11]. In the case of coronary ischemic syndromes, etiologic mechanisms have included prolapse of pedunculated left or right coronary cusp tumors into their respective coronary ostia or direct embolization of tumor material from the cusp lesion to the coronary artery[2,6]. When located on the aortic valve, papillary fibroelastomas are usually found on the ventricular side of the cusp in the more central portions[1-3]. Papillary fibroelastomas may occur as the only cardiac pathology, but they also have been described in association with congenital and

valvular disease[4]. In the literature, cases of pulmonary embolism and tricuspid valve fibroelastoma were described[12]. This case shows an example of a papillary fibroelastoma located on the aortic valve that caused cerebral arterial embolism. The etiologic mechanism of cerebral embolism included direct embolization of tumor material from the cusp lesion or embolization of a platelet-fibrin clot. Initially, the aortic mass associated with suspected antiphospholipid syndrome was interpreted as an endocarditis-like vegetation. The antiphospholipid syndrome is a disorder characterized by a variety of clinical phenomena, including arterial and venous thrombosis, thrombocytopenia, and obstetric complications, in conjunction with the presence of antiphospholipid antibodies. Classification criteria for antiphospholipid syndrome were first suggested by Harris et al.[13]. These criteria include: thrombosis (venous or arterial), miscarriage (at least two), lupus anticoagulant test or anticardiolipin antibodies identified on two occasions more than 8 weeks apart. Alarcon-Segovia et al.[14] proposed more detailed criteria. In our case after 8 weeks, the lupus anticoagulant test was negative, anti-beta$_2$ glycoprotein I antibody and anticardiolipin antibody measurements were negative. The diagnosis of antiphospholipid syndrome was not confirmed[15,16]. Differentiation of papillary fibroelastoma from vegetations is important[2,17]. In our case initially diagnostic confusion existed, but considering the clinical setting of the illness and due to transesophageal echocardiography features, the correct diagnosis of papillary fibroelastoma was made. This lesion detected during evaluation of an unexplained cerebral ischemic stroke should certainly be removed, because there is strong circumstantial evidence of a causative relationship. It is possible that there is a large pool of undetected asymptomatic cases, and those cases seen during evaluation of various cardiac or cerebral symptoms represent only the small proportion of patients who become symptomatic. The size of the tumor does not correlate with the development of symptoms. Because of their potential for cerebral and coronary embolization, even small papillary fibroelastomas should be excised in the absence of major surgical contraindications[1,7,18], but there is a lack of consensus on the need for surgery when the mass is in the right heart[2,19]. Given the thrombotic potential of such lesions, preoperative anticoagulation therapy is advisable. However this precaution does not eliminate the risk of embolism[2,7,18]. In our case the patient received preoperative oral anticoagulant therapy, the clinical course was uneventful, and the resection was curative with simple excision from the free edge of the aortic valve noncoronary cusp. After 18 months the clinical course is uneventful, without tumor recurrence. In fact these lesions have not been reported to recur[2]. This case illustrates the decisive contribution of multiplane transesophageal echocardiography in the preoperative investigation. Thus, two-dimensional echocardiography proved to be the best exam for the early diagnosis of cardiac tumors, and transesophageal echocardiography has superior specificity and sensitivity for diagnosing cardiac tumors[20,21].

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