Case report
Cardiac papillary fibroelastoma of the mitral valve chordae

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Abstract
Papillary fibroelastomas are rare, benign, primary cardiac tumors, usually single and small. The neoplasm consists of a leafy, soft excrescence typically located on the cardiac valves. Although papillary fibroelastomas are usually an asymptomatic incidental finding at autopsy, or during cardiac operation, they are occasionally associated with embolic coronary or cerebral symptoms. A case of a patient is reported with papillary fibroelastoma of the mitral valve chordae, who presented several transitory ischemic attacks characterized by loss of conscience, visual bilateral deficit and right emiparesis. Because of their potential systemic embolization, we believe that these lesions should be always excised. © 1998 Elsevier Science B.V. All rights reserved.

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1. Introduction
Papillary fibroelastomas (PFE) are rare benign cardiac tumors that constitute about 7% of all primary cardiac tumors, but they are the most common primary tumors of heart valves [1,2]. In the past they had been occasionally identified at autopsy or as incidental findings during cardiac surgery. Nowadays, with the advent of echocardiography, the diagnosis of this lesion in asymptomatic patients is possible, allowing an aggressive surgical treatment to prevent cerebral or coronary embolization.

2. Case report
The patient is a 72-year-old woman with an unremarkable medical history, except for a hyperthyroidism treated with radioactive iodine. She has been suffering sporadic episodes of atrial fibrillation for 20 years. At 66 years of age she presented with the first transitory ischemic attack (TIA) characterized by tempo-spatial confusion and bilateral loss of the visus, that disappeared in few hours. A new TIA occurred 1 year later. At the duplex scan the carotid arteries appeared normal. The electrocardiogram showed chronic atrial fibrillation. A cranial computed tomography (CT) scan showed diffuse cortical cerebral atrophy with occipital and cerebellar bilateral ischemic lesions which were more evident on the left side. The transthoracic (TT) and transesophageal (TE) echocardiogram assessed the presence of a small (2 cm in diameter) and mobile left-endoventricular echodensity, attached to the septum and anterior wall; moderate mitral regurgitation;
mild aortic insufficiency; mild tricuspid regurgitation and biatrial dilatation. At 71 years of age she suffered from two other TIAs, the latter with right emiparesis. A cardiac magnetic resonance (MR) confirmed the presence of a round mass 2 cm in diameter with smooth borders and a signal intensity similar to smooth muscle (Fig. 1). Because of these clinical and laboratory findings the neoplasm was surgically excised. During cardiopulmonary bypass and aortic crossclamping a left ventriculotomy parallel to the left anterior descending artery was performed. The mass was attached with a thin stalk to two second order chordae of the anterior mitral valve leaflet. The tumor was excised, together with few second order chordae, with no damage to the mitral valve apparatus. The patient had an uneventful postoperative course and was discharged on the 12th postoperative day. Grossly, the neoplasm, measuring 4 × 3 cm, was gelatinous, white-yellowish and with many fronds. It was entirely sampled and the specimens, after fixing in 10% buffered formalin and embedding in paraffin, were cut and stained with haematoxylin-eosin, Masson’s trichrome, elastic Van Gieson and Alcian blue Pas. Immunohistochemical (IIC) stainings were performed by the streptavidin-biotin-peroxidase-complex technique. Microscopically, the tumor was composed of multiple avascular papillae covered by a layer of endocardial cells, focally hyperplastic. Each papilla had a central hyalinized collagenous core, surrounded by a loose, mixomatous and acid-mucopolysaccharide rich matrix. In the central cores, collagen and elastic tissues were present in variable proportion and few intermixed mononuclear cells were detected (Fig. 2). At IIC characterization, the lining cells showed a widespread positivity for vimentin and neuron-specific enolase, a focal staining with factor VIII related antigen. At follow up, 12 months later, the patient had no further neurologic events and a TT echocardiography assessed good left ventricular function without images of recurring endoventricular mass.

Fig. 1. Magnetic resonance illustrating a tumor attached to the ventricular septum and involving the chordae of the anterior mitral leaflet; both atrial chambers were dilated.

Fig. 2. Elongated and branching avascular papillary fronds with central hyalinized collagenous core, lined by a single layer of endothelial cells (hematoxylin and eosin stain, original magnification × 10).

3. Discussion

PFE is a rare and benign cardiac tumor (about 7% of all primary cardiac tumor) representing the most common primary tumor of the heart valves [1,2]. The majority (about 90%) of these benign lesions occur on the valves’ leaflets particularly of the left heart, but they may be located on all intracardiac structures [1–5]. These neoplasms are usually small, less than 1 cm in diameter, and more than 90% are solitary, although multiple localization has been reported.

The incidence is not clear because most patients are asymptomatic. PFEs usually occur in elderly patients (> 65 years) even if some PFEs have been described in newborn babies with cardiac congenital abnormalities [6]. Hystogenesis is unclear and there are several theories: hamartomatous or neoplastic, congenital and thrombotic [1]. The well-organized papillary structure and the distribution of papillary tumors, where thrombus are rare, have been cited as evidence for the hamartomatous theory [7]. The coexistence with congenital cardiac abnormalities and the young age of few patients support the congenital basis for the lesion. Many authors accept the view of Pomerance [8] that these are giant Lambl excrescences, formed by organization of successive layers of fibrin deposits on the endothelium. The primary event leading to the proliferation of the cells of the endothelial surface and to the formation of the papillae remains unknown. The chronic endocardium trauma appears important in the pathogenesis: turbulent blood flow and mechanical trauma may contribute to the endothelial cell hyperplasia and to the degenerative changes in the connective tissue of the stalk and in the cores of the papillae [7]. It is our opinion that the neoplastic origin is the most reliable one because the lesion has the same histogenetical
characteristic of true benign neoplasm. Supporting this theory we did not find fibrin thrombus on the fibroelastoma. Clinical symptoms are different and due to significant potential embolization, such as transient ischemic attacks or strokes, myocardial infarction or angina, peripheral acute ischemia, pulmonary recurring embolism or sudden death [1–5, 8–10]. In the literature, a PFE causing intermittent right ventricular outflow tract obstruction with cyanotic spells has been reported [6]. Embolization is due both to the great brittleness and to the tendency for easy platelet aggregation on the tumor surface [1]. Two-dimensional echocardiography (transsthoracic and transesophageal) is the most direct, reliable and the less invasive method of establishing the presence of intracardiac tumors, those with small dimension too [1, 4, 9]. To date the cardiac catheterization has not been useful and in patients with coronary artery disease a coronary angiography is indicated. Once the diagnosis of intracardiac tumor is established, MR may add more useful information. Today, operative resection of the tumor should proceed on an urgent basis because of the potential cerebral and cardiac embolization [1, 2, 9, 10]. The most problematic point is to establish the excision via (transatrial, transventricular, transaortic, combined), which depends on the exact location of the implanted stalk. Tumors may be simply excised with or without valve repair or with valve replacement depending on the extension of the valve apparatus involved with the tumor. Mitral valve reconstruction appears more easily feasible than aortic valve [1, 5, 9, 10]. In our case, the tumor was excised through a left ventriculotomy which provided an optimal surgical vision and a radical excision of the mass with no damage to the mitral valve apparatus.

References