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Cardiac Papillary Fibroelastoma and Stroke in a Young Man – Etiology and Treatment

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Introduction

Stroke in the young usually requires extensive evaluation because the uncommon causes of cerebrovascular diseases are more often seen in this population. Cardiac papillary fibroelastoma (CPF) is a rare primary cardiac tumor whose clinical importance was obscure until the introduction of echocardiography and magnetic resonance imaging (MRI) [1, 2]. Indeed, CPF used to be described as an incidental finding in autopsy studies [3–6]. In spite of its benign nature, CPF is nowadays associated with embolism causing myocardial infarction, sudden death and cerebral ischemia [4, 6–10]. However, most CPF are still found incidentally at echocardiography examination, cardiac angiography, cardiac surgery or autopsy. Cardiac symptoms are not common [2, 8, 11].

We report the case of a 32-year-old man who presented with an ischemic stroke associated with CPF. Contrary to literature recommendations [5, 10, 11], our patient refused surgical excision of the tumor. Instead, he received long-term anticoagulation therapy as alternative treatment.

Case Report

A 32-year-old man presented with 3-day right hemiparesis and aphasia of sudden onset. He had no cardiological symptoms, his past medical history was unremarkable and he was taking no medication. Blood pressure on admission was 130/80 mm Hg, and his rhythmic heart rate was 76 beats/min. Cardiorespiratory examination was normal. Neurological examination was significant for a right hemiparesis and mild global aphasia. No mental status disturbance was noted.

A computed tomography (CT) scan revealed a left middle cerebral artery territory infarct (fig. 1). High density in the proximal middle cerebral artery, as a sign of thrombosis, was not noted. Carotid duplex sonography was normal. There was no abrupt contrast stop in any vessel on the cerebral arteriography, as a sign of spontaneous left middle cerebral artery recanalization. Except for a 15 times elevated C-reactive-protein level, an ultrasensitive, complete laboratory workup including coagulation tests, fibrin degradation products, antithrombin III protein, C and S protein

activities, rheumatoid factor, antinuclear antibody, anticardiolipin antibody and serum VDRL was negative. Chest radiography and electrocardiogram showed no remarkable findings.

A transthoracic echocardiogram revealed a mobile mass attached to the chordae of the mitral valve. The transesophageal echocardiogram confirmed the presence of a 2 × 2 mm pedunculated mobile round mass, attached to the chordae of the posterior leaflet of the mitral valve, the appearance of which was consistent with CPF (fig. 2). Heart MRI was then obtained to characterize the tissue tumor. It demonstrated a round tumor with signal intensity similar to the cardiac muscle on T₁- and T₂-weighted sequences consistent with the presumptive diagnosis of CPF rather than a myxoma (fig. 3). There were neither valve abnormality, intracavitary thrombus, septal defects nor mitral regurgitation.

Surgical excision of the tumor was initially proposed as first-choice treatment. However, due to the intimate relationship between tumor and mitral chordae, the possible need of valvar replacement could not be ignored. The patient refused to undergo surgery, and, alternatively, he has received oral anticoagulation therapy with sodium warfarin. Along a 48-month follow-up period, the International Normalized Ratio varied between the therapeutic interval of 2 and 3. Cardiac function remained stable, and neurological deficits improved slowly (current modified Rankin scale and Barthel index scores both 1); the patient has returned to his daily activities. Control brain MRI after 48 months did not reveal any new asymptomatic infarct lesion (fig. 4).

Fig. 1. First CT scan of the brain showing a left middle cerebral artery territory infarct.

Fig. 2. Transesophageal echocardiogram. The lesion exhibited homogeneous hyper-echogenicity and measured roughly 1.1 cm. The echographic characteristics were highly suggestive of CPF. AE = Left atrium; VE = left ventriculus.

Fig. 3. Cardiac MRI. The valve presented a normal aspect, and there were no indications of intracavitary thrombus, mitral regurgitation or evidence of septal defects; however, the tumor intimately adhered to the chordae tendineae.

Fig. 4. MRI of the brain 48 months after the initial neurological event showing no new infarcted area.

Discussion

We report the case of stroke in a young adult associated with a cardiac mass. Although there is no histological confirmation, its radiological appearance is highly consistent with CPF [1, 8, 11, 12]. The thrombophilia screening was not complete, and the possible contribution of a nonevaluated factor (i.e. homocysteine, factor V Leiden, prothrombin gene mutation) cannot be excluded.

The estimated prevalence of primary cardiac tumor is 0.02% [7, 8, 13], and CPF, accounting for only 7–8% of them, is the third most common after atrial myxomas and lipomas [4, 7, 14, 15]. Although possibly found anywhere in the heart, CPF affects predominantly the cardiac valves (70–80%) [8, 12]. Indeed, CPF is the most common valvular tumor [2, 7, 16, 17]. Among them, the mitral valve is the second most common involved (35%) [8, 12], with CPF tumors occurring on anterior or posterior leaflets, the chordae tendineae or the papillary muscles [18]. In a large systematic review [8], CPF size varied between 2 and 70 mm, most being smaller than 10 mm. Multiple tumor occurrences are rare [4, 6].

Multiple soft and friable papillary fronds attached to the endocardium by a short pedicle give CPF a flower-like appearance, sometimes also described as a ‘sea anemone’ [4, 8, 15]. The tumor consists of fibrous tissue surrounded by an elastic membrane, covered by the endothelium [4, 7, 15]. This structure may be obscured by adherent thrombus [4, 8, 15].

The most common clinical manifestation of CPF is related to its tendency to embolize [7]. Indeed, we found in our revision 20 patients either with retinal ischemia, transient ischemic attack or stroke associated with CPF. Two mechanisms could explain such a process: either embolization from the tumor itself or from the thrombus on its surface [2, 5, 6, 8, 10, 15, 17, 19, 20]. However, unlike myxomas, pathology studies have only rarely found evidence of CPF fragments at the site of the arterial occlusion, suggesting that a thrombus may be the source of emboli in most cases [4, 9, 21, 22]. Determination of embolus composition may be decisive for choosing the appropriate treatment [23]. Surgical excision with valve repair is considered the first-choice treatment for CPF,

especially in symptomatic or large mobile tumors [4, 7–12, 19]. Its pedicle makes removal of CPF usually easy. Selected patients, as in our case, may need valve replacement [9–11, 15]. Oral anticoagulation is recommended only for patients who are not surgical candidates or for those whose tumor is asymptomatic and non-mobile [4, 8, 10]. However, no randomized controlled data are available on treatment efficacy. To our knowledge, there is no literature report of a symptomatic patient who has not presented any recurrent ischemic event after long-term oral anticoagulation [4–6, 9–11, 16, 17, 20, 23, 24].

In conclusion, CPF is a potentially treatable cause of embolic ‘stroke in the young’ [15]. Surgery excision is curative and should be the first choice. For those patients who are waiting for surgery or are not surgical candidates, oral anticoagulation therapy may be an alternative treatment. Indeed, the tumor may act as a nidus for thrombus formation rather than embolize its fragments. On the other hand, randomized controlled studies are needed to evaluate the efficacy of treatment choices.

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A Caudal Mesencephalic Infarct Presenting Only with Tetra-Ataxia and Tremor

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Introduction

Midbrain lesions cause various clinical syndromes, including oculomotor disorders, ataxia, hemiplegia, and sensory disturbances [1–5]. Cerebellar ataxia can occur bilaterally with lesions located at the decussation of the superior cerebellar peduncles at the level of the caudal midbrain; cerebellar ataxia is almost always present with other mesencephalic syndromes, such as ophthalmoplegia [1–4]. We describe a patient with a small infarct located