Cardiac Papillary Fibroelastoma as a Cause of Embolic Stroke: Ultrasound and Histopathological Characteristics

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Abstract

Although a cardiac papillary fibroelastoma is a benign cardiac tumor, it can cause severe embolic complications. A 51-year-old man presented with an ischemic stroke in the right middle cerebral artery territory. Transesophageal echocardiography revealed a small mobile tumor on the mitral valve as the only detectable source of emboli to the brain. On histology, the tumor was diagnosed as a papillary fibroelastoma. In this paper, the detailed characteristics of the tumor on ultrasound and histopathology are documented. In patients with cryptogenic stroke, transesophageal echocardiography should be done to rule out such an unusual embolic heart disease.

Key words: papillary fibroelastoma, cardiac tumor, stroke, cerebral infarction, transesophageal echocardiography, ultrasound

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Introduction

Primary cardiac tumors are rare and generally benign; they are found in about 0.02% of autopsy samples (1). After myxoma and lipoma, cardiac papillary fibroelastoma (CPF) is the next most common primary cardiac tumor; it accounts for approximately 8% of all primary cardiac tumors (2). Although CPF is benign, it can cause life-threatening complications, such as stroke, systemic embolism, acute cardiac failure, and sudden death. Thus, prompt diagnosis and appropriate management are needed for this tumor. We report a case of an embolic stroke due to CPF, which was detected on transesophageal echocardiography (TEE) and successfully resected surgically.

Case Report

A 51-year-old man who was a heavy smoker but who did not have any other vascular risk factors suddenly suffered from left hemiparesis and was admitted 30 minutes later. On admission, his blood pressure was 158/78 mmHg, and his pulse was 88 beats per minute and regular. There were no heart murmurs. He was disoriented, his comprehension was impaired, and he had left-sided unilateral spatial neglect, as well as the extinction phenomenon. His eyes were deviated to the right. There was marked left upper and lower limb weakness with decreased pinprick sensation. The deep tendon reflex of the left knee was enhanced. The National Institutes of Health Stroke Scale (NIHSS) score was 10. Within the initial hour after stroke onset, these neurological deficits were rapidly improved, and, except for a mild left hemiparesis, returned to normal, resulting in the NIHSS score of 3.

Blood tests, including hemostatic and autoimmunological parameters, were normal. To be specific, prothrombin time, activated partial thromboplastin time, D-dimer, thrombin-antithrombin III complex, and protein C and S activities were within normal limits, and antinuclear, antiphospholipid, anticardiolipin antibodies and lupus anticoagulant testing were negative.

An emergent computed tomography of the brain showed only an asymptomatic old infarct in the left occipital lobe. On diffusion-weighted brain magnetic resonance imaging
(MRI), a hyperintense lesion was delineated in the right parietal cortex (Fig. 1A). The carotid duplex ultrasound was unremarkable. Three hours after stroke onset, the right carotid arteriogram revealed occlusion of the descending branch from the middle cerebral artery (Fig. 1B, C). The branch occlusion on angiography, together with a territorial cortical infarct on MRI and the spectacular shrinking deficit of neurological symptoms (3), suggested an embolic mechanism as the cause of the stroke.

Chest X-ray was normal. Transthoracic echocardiography (TTE) and long-term electrocardiography showed no embolicigenic disorders including atrial fibrillation. TEE was performed on the third hospital day, and a small mobile mass (5.0×5.0 mm) on the posterior leaflet of the mitral valve on the atrial side was found (Fig. 1D). The mass was oval, well-demarcated, and isoechoic. The valve itself was intact. Other potential sources of emboli, including right-to-left shunt and aortic atheroma, were not detected. There were no physiological or laboratory data suggesting infective endocarditis or malignant diseases causing non-bacterial thrombotic endocarditis. Intravenous heparin was immediately started, followed by oral warfarin therapy; however, the mass did not shrink on the follow-up TEE two weeks later. Thus, the patient was diagnosed as having a cardiac tumor.

Anticoagulation with warfarin was continued to prevent possible embolism due to the superimposed thrombus present on the cardiac mass. Since CPF causes acute coronary syndrome and sudden cardiac death by embolism to the coronary artery (4-8), we performed coronary angiography and ascertained that the coronary arteries were intact. On the 65th hospital day, the patient had a surgical resection of the mass. Macroscopically, a white round mass, 5.0 mm in diameter, was found adhering to the posterior mitral leaflet (Fig. 2A). The mass did not have a peduncle. The surface was covered with an agar-like material (Fig. 2B); no thrombus was noted. The mass consisted of many white papillary fragments with frond-like projections. Microscopically, typical elastic fibers within a central fibrous core, covered by a single layer of endothelial cells that supported the diagnosis of CPF, were found (Fig. 2C, D). The patient had no perioperative complications and no further embolic events.

Discussion

In this paper, the detailed characteristics of the CPF on
ultrasound and histopathology are documented. It was found that a benign CPF, which was identified only on TEE, could cause an embolic stroke.

CPF is usually located on the aortic valve (44%) or the mitral valve (35%), and it is almost always solitary (4, 9, 10). Echocardiographic appearance of CPF include the following findings: (a) the tumor is round, oval, or irregular in appearance, with well-demarcated borders and a homogeneous echogenicity; (b) CPF has a clear surface with a bright echoic core of collagen fibers or fibrous tissue; (c) most CPFs are small (99% were <20 mm in the largest dimension); (d) nearly half of CPFs have small stalks and are highly mobile; (e) CPF exhibits movement on echocardiography similar to the flexion-extension of a finger (1, 9, 11). The myxoma, vegetation, and intracardiac thrombus should be differentiated from the CPF on TEE. The myxoma is larger and more heterogeneous than CPF, and is mostly located in the left atrium. The vegetation is an irregularly shaped, discrete echogenic mass, and shows high-frequency movement independent from that of intrinsic structures. The thrombus shows a central echolucency due to clot lysis, laminated appearance, and irregular or lobulated border, does not have a pedicle, and is typically located in the atrial appendage (1, 2, 12, 13).

CPF is histologically composed of multiple papillary frond-like projections and has a central core of dense acellular collagen covered by a single layer of normal endothelial cells; CPF adheres to the endocardium by a short peduncle (4, 9, 10, 14, 15). Although the mass in the present patient did not have a peduncle, other pathological findings were compatible with those previously reported (4, 9, 10, 14, 15).

As shown in this case, TTE often fails to detect this tumor. In a case-control study, the sensitivity of TTE for CPF was 61.9%, while that of TEE was 76.6% (9). Even when abnormal lesions are seen on TTE, it is often difficult to diagnose them as CPF, partly because of difficulty in identifying the site at which the tumor is attached to the cardiac structure (18). In contrast, TEE helps to differentiate CPF from other intracardiac tumors, in particular myxoma and the vegetation of endocarditis (19). When the CPF is large, TEE can delineate a clear surface with a bright echoic core of collagen fibers or fibrous tissue and a stippled edge with vibration at the tumor-blood interface (11, 15, 20). The present case suggests that TEE is mandatory for patients with embolic stroke who are not found to have an embolicogenic disorder on routine cardiac examinations, even if their past history is non-contributory and their cardiac status is normal.

A fibrin thrombus attached to the circumference of the
CPF (4, 21, 22), as well as the CPF itself (4, 7, 8), can be a source of embolic material. Thus, anticoagulant therapy is recommended for stroke patients with CPF up to the day of the surgical resection. In general, for patients who develop symptomatic embolism, surgical excision is recommended (4, 9). In addition, when the tumor is mobile, pedunculated, or has a diameter >10 mm, patients should be treated surgically even if they are asymptomatic (4, 9, 23). These recommendations are not based on a randomized controlled study, since CPF is rare. However, CPF re-growth following surgical excision has not been reported.

Although no randomized controlled data are available on its efficacy, the symptomatic patients who are not surgical candidates could be treated with long-term oral anticoagulation (4, 5). The previous reports have provided no firm evidence-based guidelines regarding the use and duration of anticoagulation post excision of CPF (4, 16). Some reported cases continued (16, 24) and some discontinued anticoagulation postoperatively (11, 23). All of these patients have remained free of cardiovascular or cerebral events without a recurrence since their operation.

In conclusion, CPF is a rare but potentially treatable cause of embolic stroke. TEE should be done in patients with cryptogenic stroke. In patients with CPF, anticoagulant therapy followed by surgical resection should be considered, even though the tumor is a benign lesion.

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