A Rare Case of Stroke in a 76-Year-Old Woman: Left Atrial Papillary Fibroelastoma as the Culprit

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Patient: Female, 76-year-old
Final Diagnosis: Left atrial papillary fibroelastoma
Symptoms: Right side weakness and dysarthria
Clinical Procedure: Tumor excision
Specialty: Cardiology

Objective: Rare disease
Background: Papillary fibroelastoma is the most common type of benign primary cardiac tumor and is usually asymptomatic. However, tumor fragments or surface thrombus can embolize and cause transient ischemic attacks, strokes, or myocardial infarction. This report describes a 76-year-old woman who presented with dysarthria and right-sided weakness due to a stroke associated with a left atrial papillary fibroelastoma.

Case Report: A 76-year-old woman visited the Emergency Department because she had right-sided weakness and dysarthria from 12 h ago. Brain magnetic resonance image was done at the Emergency Department, showing multiple small embolic, acute infarction in left basal ganglia and fronto-temporo-parietal lobes. Transthoracic and transesophageal echocardiogram showed a hypermobile echogenic mass (0.8×1.5 cm) with villous surface on the orifice of left atrial appendage. Twenty-four-hour Holter monitoring was performed to evaluate the cause of cerebral infarction, and there was no paroxysmal atrial fibrillation. Thoracic computed tomography angiography also showed a sea anemone-shaped mass around the left atrial appendage. Cardiac tumor excision was done via a lower partial sternotomy. Histopathologic analysis showed multiple delicate fronds, and the avascular fibroelastic cores were lined by a single layer of CD31-positive endothelial cells. Histopathologic findings were consistent with papillary fibroelastoma. The patient was discharged without any other complications on day 30 of hospitalization.

Conclusions: This case highlights the importance of cardiac imaging in patients with acute stroke, including transthoracic and transesophageal echocardiography, which can show the typical imaging features of papillary fibroelastoma and other intracardiac sources of embolus.

Keywords: Cardiac Papillary Fibroelastoma • Embolic Stroke • Echocardiography, Transesophageal

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### Introduction

Primary cardiac tumors are extremely rare diseases, with a very low incidence of less than 0.03% in the general population [1]. Most primary cardiac tumors are benign (>90%), and include myxomas, papillary fibroelastoma (PFE), rhabdomyomas, hemangiomas, lipoma, and leiomyomas [1,2]. PFE is one of the most common primary cardiac tumors and it constitutes 11.5% of all primary cardiac tumors [1,2]. PFE is most commonly asymptomatic and discovered incidentally [3]. It is usually observed on the heart valves; however, it can be detected in other sites of the heart, such as the left atrium [4,5]. PFE can cause myocardial infarction, stroke, valve dysfunction, and sudden cardiac death, particularly when it is located in the left side of the heart [6,7]. Surgical excision is a recommended treatment, unless there is a contraindication to surgery. If the symptomatic patient is a poor surgical candidate, anticoagulation therapy is reasonable to prevent embolic events [8]. This report describes a 76-year-old woman who presented with dysarthria and right-sided weakness due to a stroke associated with a left atrial PFE.

### Case Report

A 76-year-old woman visited the Emergency Department because she had right-sided weakness and dysarthria from 12 h ago. She had hypertension and angina pectoris and was taking aspirin 100 mg, rosuvastatin 10 mg, amlodipine 10 mg, valsartan 160 mg, and hydrochlorothiazide 25 mg. Her blood pressure was 130/85 mmHg, and she had a bradycardia, with heart rate of 45 beats per min. She had no fever, with a body temperature of 36.3°C. Right-sided motor weakness was recovered when she was examined in the Emergency Department. There was no heart murmur except bradycardia. Electrocardiogram showed junctional bradycardia, without ischemic change. There were no abnormal laboratory findings, such as hyperkalemia. Brain magnetic resonance was done for evaluation of the cause of right-sided weakness and dysarthria, and the image showed multiple small embolic, acute infarction in left basal ganglia and fronto-temporo-parietal lobes (Figure 1). Anticoagulation with intravenous heparin was administered. Transthoracic echocardiogram was done for detecting a source of cardioembolism, and there was a mobile mass located in left atrium. Then, transesophageal echocardiogram was done, showing a hypermobile echogenic mass (0.8×1.5 cm) with a villous surface on the orifice of left atrial appendage, and there was no thrombus in the left atrial appendage (Figure 2). Twenty-four-hour Holter monitoring was performed to evaluate the cause of cerebral infarction. It showed intermittent complete atrioventricular block, and there was no paroxysmal atrial fibrillation.

Cardiac tumor excision was planned, and thoracic computed tomography angiography was done, which showed a sea anemone-shaped mass around the left atrial appendage (Figure 3).

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**Figure 1.** Brain magnetic resonance images shows multiple small embolic, acute infarction in left basal ganglia (A, arrow) and left fronto-temporo-parietal lobes (B, arrows).
Cardiac tumor excision was done to prevent further embolic events. After a lower partial sternotomy starting from the right second intercostal space, the patient was on a standard cardiopulmonary bypass, with right femoral artery and femoral vein cannulation, with cooling to 32°C. Cardiac arrest was induced with del Nido cardioplegic solution through an antegrade cannula. A standard right-sided left atriotomy was made parallel to the interatrial groove. The tumor was pedunculated and it was attached to the orifice of the left atrial appendage. Its surface was papillary and villous. Thereafter, cardiac tumor excision was performed. The left atrium was then closed with 4-0 prolene continuous sutures.

Histopathologic analysis showed multiple delicate fronds (Figure 4A, 4B). Elastic fiber and Masson’s trichrome stain highlighted the fibroelastic nature of arborizing fronds arising from a central stalk (Figure 4C, 4D). The avascular fibroelastic cores were lined by a single layer of CD31-positive endothelial cells (Figure 4E). Histopathologic findings were consistent with PFE.

Intermittent complete atrioventricular block was repeated, and a permanent pacemaker was inserted on day 3 after surgery. The patient was discharged without any other complications on hospitalization day 30.

**Discussion**

This case shows that even though primary cardiac tumor is rare, and left atrial PFE is rarer than valvular PFE, and it could be a source of stroke [8]. Therefore, multimodality images should be considered to detect a source of cardioembolism, as much as possible [7]. While cardiac tumors are relatively rare, PFEs are among the more common tumors encountered [5]. Left atrial PFE is rarer than PFEs, which are located on heart valves, and it can be a cause of cardioembolic stroke [5,9]. PFEs are usually located on the downstream side of the aortic and mitral valves and are found on the tricuspid valves and...
endocardium less commonly [7]. PFEs are usually a small size (less than 1.5 cm) and have stalk attachment to the endocardium [10]. In the present case, the PFE was noted to be originating from the left atrium; therefore, histopathology was important in establishing the etiology, given the atypical location.

It is important to differentiate other diagnosis of cardiac mass, such as Lambl excrescence, cardiac myxoma, vegetations, and thrombi. In particular, Lambl excrescences are frequently encountered, and at times it becomes challenging to differentiate them from PFEs. However, these entities are just degenerative changes at valvular coaptation sites and are not associated with embolization [11].

Transesophageal echocardiography is more sensitive to detect PFE than is transthoracic echocardiography, because PFE is small and hypermobile [3]. Cardiac computed tomography and cardiac magnetic resonance image cannot usually identify them, because of the small size and mobility; however, we could detect a typical sea anemone-shaped mass on the orifice of the left atrial appendage via thoracic computed tomography angiography [12].

Undiagnosed paroxysmal atrial fibrillation is frequently the most common cause of a stroke in the elderly when a definitive source is not established [13]. Even though there was no paroxysmal atrial fibrillation in 24-h Holter monitoring in this patient, longer duration of Holter monitoring would have been better to increase the rate of diagnosis of paroxysmal atrial fibrillation.

Surgical excision can be considered in specific scenarios: complications, such as stroke, ischemia due to coronary ostial occlusion by the tumor, highly mobile tumors, or large tumors measuring 1 cm or larger [6,14]. This patient underwent surgical excision because she had stroke and she had a large and hypermobile mass in the left side of the heart.

Yandrapalli et al reported 2 cases of PFE that were located on the aortic valve [15]. One case showed acute myocardial infarction due to obstruction of the right coronary artery with PFE, which was found after autopsy. If PFE could have been found earlier, excision of the tumor would have been helpful to treat the patient. Another case had transient ischemic attack due to PFE on aortic valve. The second case was treated by excision of the tumor. Our case showed embolic infarction caused by PFE, which was located on the orifice of
the left atrial appendage. Transesophageal echocardiography showed a 3-dimensional structure of PFE, and thoracic computed tomography angiography clearly showed a sea anemone-shaped mass. Multimodality images would be helpful to detect a source of cardioembolism in cases of acute myocardial infarction and stroke.

Gowda et al analyzed 725 cases of PFE and reported that tumor mobility was the only independent predictors of death or nonfatal embolization [8]. This case also showed a similar result, that hypermobile PFE on the left atrial appendage was found in a patient with embolic cerebral infarction. Therefore, surgical excision should be considered in symptomatic patients who have hypermobile PFE.

Asymptomatic patients with small, left-sided, nonmobile-type PFEs can be cautiously observed and treated with antithrombotic therapy [14]. Tamin et al reported that cerebrovascular accident risk in patients with PFE suspected by echocardiography who did not undergo surgical excision at 1 and 5 years was 6% and 13%, respectively. It was lower than expected, compared with age- and sex-matched control subjects; however, it was higher than the cerebrovascular accident risk of patients who underwent surgical excision [3]. They recommended that patients who have low surgical risks with left-sided PFE, regardless of size, mobility, or location, consider surgical excision. If surgical risk is high or patients refuse surgery, antiplatelet therapy or anticoagulation therapy is reasonable to reduce the risk of stroke [3,14].

Conclusions

This case highlights the importance of cardiac imaging in patients with acute stroke, including transthoracic and transesophageal echocardiography, which can show the typical imaging features of papillary fibroelastoma and other intra-cardiac sources of embolus. PFE is one of the most common types of primary cardiac tumor and can cause embolic cerebral infarction. Surgical excision can be considered if PFE causes a stroke, is located on the left side of the heart, or is large or hypermobile.

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