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Ischemic Stroke Revealing Valvular Fibroelastoma: A Case Report

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Abstract

Papillary fibroelastoma is a rare, benign primary cardiac tumor of unknown etiology. It most commonly arises from the valvular endocardium. It affects all age groups from the neonatal period to the tenth decade of life. Clinically, patients with papillary fibroelastoma are asymptomatic in one-third of the cases and their discovery is fortuitous, but it can be the cause of serious embolic complications such as ischemic cerebral accidents, acute coronary syndromes, and sudden death. Currently, the diagnosis is easily evoked in echocardiography. Surgical excision of this tumor is a safe and effective treatment to prevent the risk of embolic recurrence. We report the case of a 59-year-old patient who was hospitalized for a left MCA (middle cerebral artery) ischemic stroke on FLAIR sequence confirmed on cerebral MRI. Transthoracic and transesophageal echocardiography revealed an aortic valve tumor. Histological examination of the surgical specimens confirmed the diagnosis of fibroelastoma. Simple excision of the tumor protected the patient from an embolic recurrence.

Keywords: Fibroelastoma; Ischemic stroke; Aortic; TEE; Surgery

Introduction

Papillary fibroelastoma is a very rare benign cardiac tumor, with a preferential valve location. Currently, it is easily diagnosed by echocardiography. Fibroelastoma is often asymptomatic, as it can be the cause of serious embolic events. Surgical excision of this tumor is the safest and most effective treatment to prevent the risk of embolic recurrence. We report the case of a patient who suffered a stroke. Transthoracic and transesophageal echocardiography revealed an aortic valve tumor. Histological examination of the surgical specimens confirmed the diagnosis of fibroelastoma. Simple excision of the tumor protected the patient from embolic resurgence.

Case Report

We report the case of a 59-year-old patient, who was a smoker and has been in end-stage renal failure for 3 years, on hemodialysis 3 times a week. He presented right hemiplegia, aphasia, and non-quantified fever. The cerebral MRI revealed a left MCA (middle cerebral artery) ischemic stroke on FLAIR sequence (Figure 7). On the ECG, there was a regular sinus rhythm with diffuse negative T waves, and without conduction abnormalities (Figure 1).

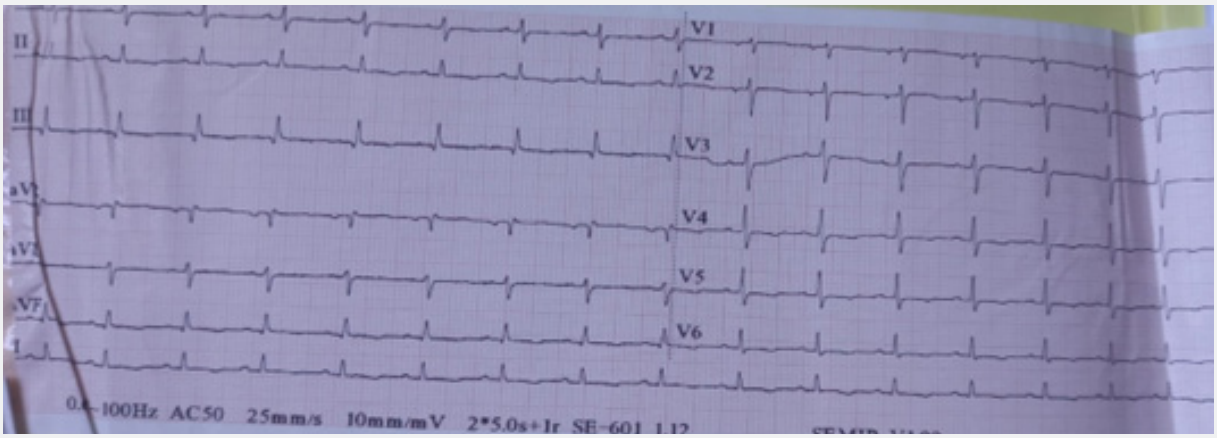

Figure 1: ECG showing regular sinus rhythm, heart rate at 95 BPM with diffuse negative T waves, and fixed PR.

On his biological assessment, there was a slight increase in white blood cells at 12800e/mm, the C reactive protein was elevated at 96 mg/l, and blood cultures were negative. No further biological abnormalities were noted.

The trans-thoracic echography revealed a calcified tricuspid aortic valve, of a rounded hyperechoic image on the ventricular side of the aortic valve, measuring (13x17mm) (Figure 2-3), the mean gradient on the aortic valve was measured at 41mmgh with an aortic valve area of 0.8 cm^2, associated with moderate aortic regurgitation.

The clinical and imaging presentation was suggestive of two possible diagnoses: infective endocarditis and a tumoral mass in particular a fibroelastoma. Therefore, the need to conduct transesophageal echocardiography.

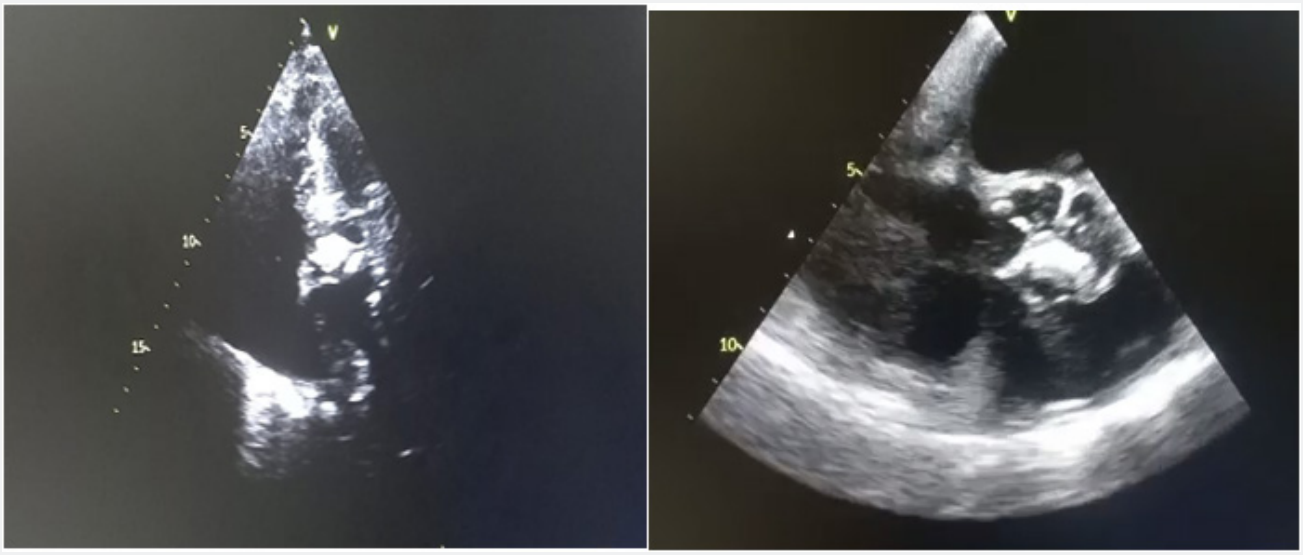

Figure 2: a 5 chamber view showing a circular hyperechoic mass on the ventricular aortic side measuring 13x17mm.
Figure 3: a short axis sternal view showing the aortic valve with a hyperechoic mass inserted on the right coronary cusp.

TEE unveiled a sessile 8x15mm mass, located on the ventricular surface of the right coronary cusp, associated with moderate aortic insufficiency (Figures 4, 5, and 6).

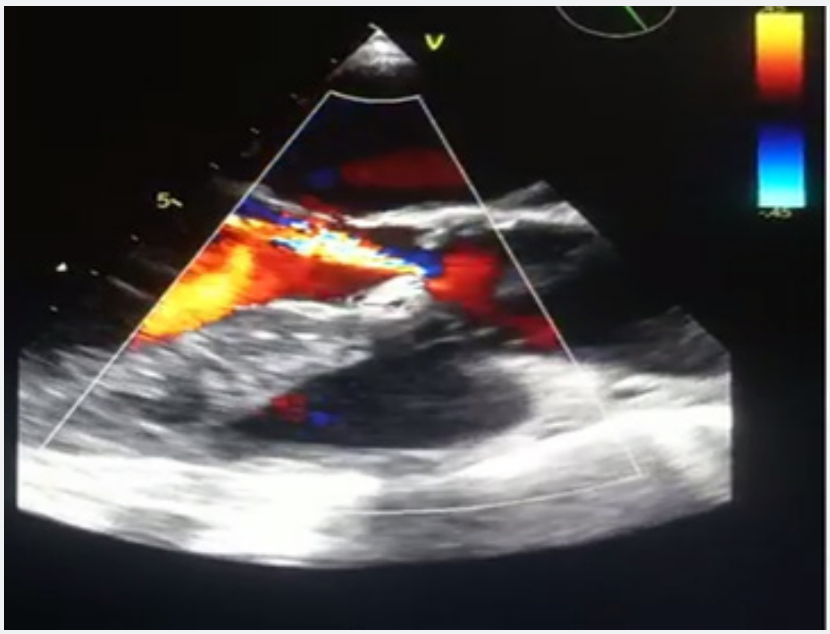

Figure 4: TEE showing moderate aortic regurgitation.

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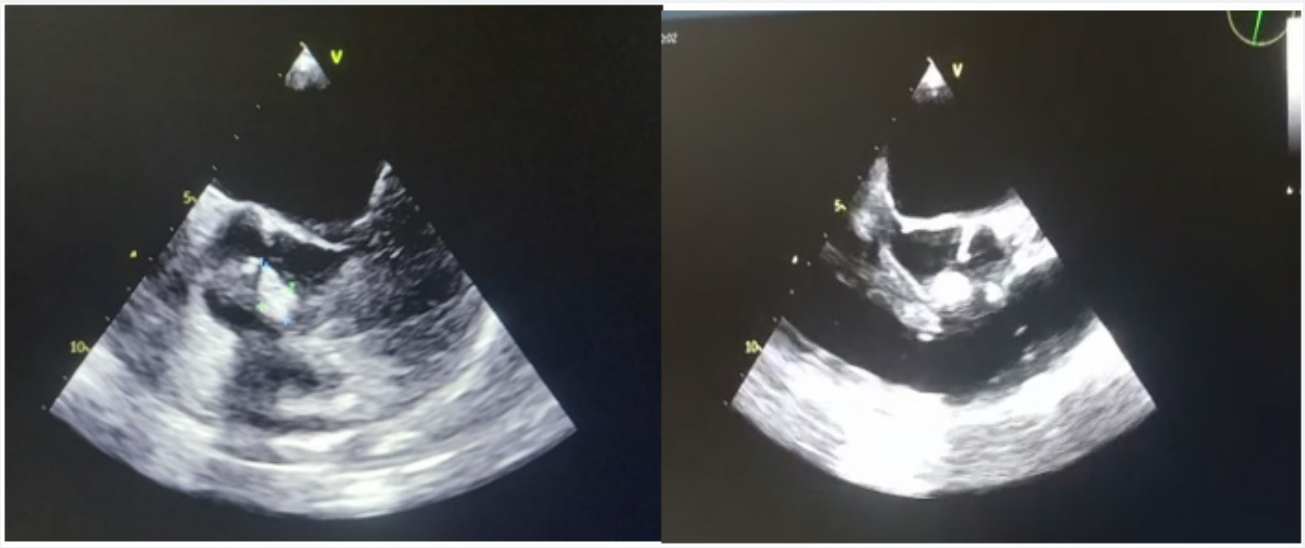

Figure 5 and 6: TEE showing a sessile mass on the ventricular side of the right coronary cusp measuring 13x8mm.

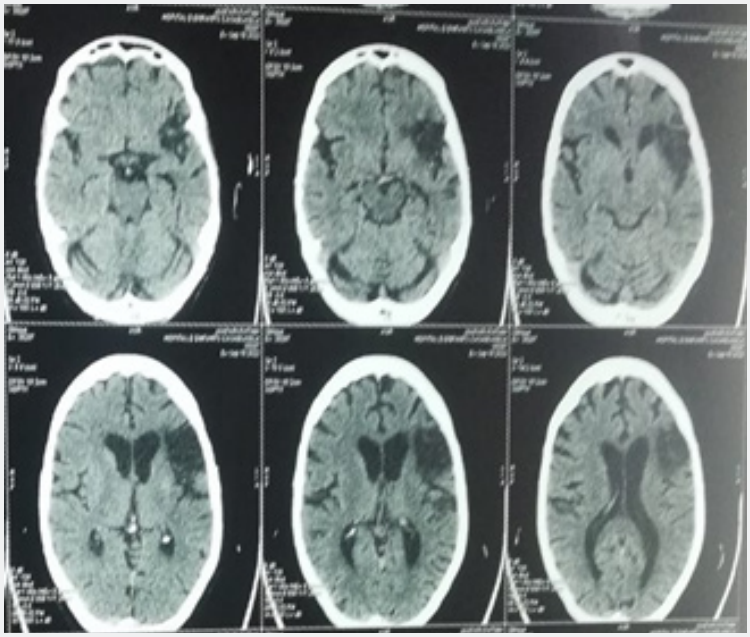

Figure 7: Cerebral MRI, a left MCA (middle cerebral artery) ischemic stroke on FLAIR sequence.

Given this context and since the rest of the etiological assessment was negative and given the absence of ultrasound change at one month, the tumor hypothesis was reinforced. Surgical intervention under extracorporeal circulation allowed the tumor excision with the replacement of the aortic valve by a mechanical valve. Histological examination proved that it was a fibroelastoma. The patient was able to leave the hospital on the tenth postoperative day.

Discussion

The heart is very rarely the site of tumors. In the general population, their incidence is 0.02%. Cardiac papillary fibroelastomas (CPF) represent about 10% of primary cardiac tumors. They are the most frequent benign intracardiac formations after myxomas and lipomas. CPFs are a recognized source of ischemic strokes of cardiac origin, easily detected by echocardiography with a clear advantage for transesophageal imaging [1,2].

The aortic valve is the most affected (30% of cases) followed by the mitral valve (20-25% of cases) [1]. Macroscopically, it looks like a sea anemone. Its implantation base is pedunculated. Its body forms many folds. Its size can vary from two millimeters to seven centimeters [2]. Histologically, it is lined with a monolayer

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of endothelial cells. The underlying connective tissue is rich in collagen fibers, elastic fibers, glycosaminoglycans, and smooth muscle cells. No neoplastic cells are described in the literature. However, complex cytogenetic abnormalities have been identified, giving them characteristics of local malignancy [3]. The possible locations of fibroelastomas are numerous [2,4,5]: the aortic valve most often and exceptionally non-valvular: the left ventricle wall, pulmonary vein, and right ventricle. They can be single or, in less than 10% of cases, multiple [5].

The diagnosis of papillary fibroelastoma was most often on autopsies [6] until the advent of TEE. The spiral CT scan has shown good performance [7]. Magnetic resonance imaging can also help with diagnosis. It shows a solid mass characterized by a moderate signal hyperintensity in T1 and T2 weighted MRI sequences without injection of gadolinium [8].

Currently, they are often detected incidentally, during a cardiac ultrasound, and most often remain asymptomatic because they are only exceptionally responsible for valvular dysfunction [4].

Nevertheless, papillary fibroelastoma can be a source of emboli in the coronary arteries [5] with a risk of sudden cardiac death [8], the arteries of the lower limbs [4], the central retinal artery, or the cerebral arteries. These ischemic strokes can occur at any age. In half of the cases described in the literature, the discovery of papillary fibroelastoma is preceded by transient [4] or constituted ischemic attacks, all of which often affect the left Sylvian territory.

Ischemic strokes due to papillary fibroelastoma are embolic due to a detachment and migration of a fragment of Lambl's excrescence or a thrombus formed on the papillary fibroelastoma [9]. This embolic character could be demonstrated on transcranial Doppler, by the detection of HITS (high-intensity transient signals) which disappeared after surgical excision of this malformation.

The differential diagnoses of fibroelastoma are intracardiac thrombus, other tumors, benign or more rarely malignant, and vegetations in the context of infective endocarditis as the case of our observation. The clinical context, evolution under anticoagulant and/or antibiotic treatment and imaging data are necessary to confirm the diagnosis [10].

Indeed, because of the embolic risk and ischemic recurrence, the treatment consists of tumor excision. As in the case of our patient, valvular repair, or even replacement, is sometimes necessary depending on the size, location, and associated lesions [4,11].

Gowda et al. [2] proposed an alternative treatment based on ultrasound monitoring under the protection of anticoagulant treatment. The indication for surgery was based, in asymptomatic patients, on the mobile nature of the tumor. However, no study has validated this strategy. By protecting the patient from a serious cardiovascular complication and in the absence of tumor reoccurrence described in the literature, we believe, like many authors [1,4-6], that surgical treatment must be systematic. It allows complete healing, with low operative risk.

Conclusion

Papillary fibroelastoma is a rare but dangerous cardiac tumor, easily identified on echocardiography. Surgical excision remains the reference treatment.

Declarations

Ethics approval and consent to participate: IT is not applicable.

Consent for Publication

In accordance with international and academic standards, written consent for publication was obtained from the patient and retained by the authors.

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