








Mitral papillary fibroelastoma as a cause of stroke in a young patient

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ARTICLE INFORMATION

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Competing interests

The authors declare no competing interests.

Figures

Images from complementary tests are shown with patient's consent.

Abbreviation

TEE: transesophageal echocardiography

ABSTRACT

Papillary fibroelastoma is an infrequent tumor, with a prevalence estimated between 1 and 7.9% of all primary cardiac tumors, which—at the same time—have a very low incidence (between 0.001 and 0.28%), because the great majority of them (almost a 90%) are benign. Here is presented the case of a 41-year-old man who had a stroke. A cardiac tumor in the anterior mitral valve suggestive of papillary fibroelastoma was found in the echocardiographic study. The surgical intervention was decided because of the possibility of resulting embolic complications with recurrence of cerebrovascular ischemic events. A triangular resection of the tumor and mitral annuloplasty were performed with good results. The pathological study confirmed the diagnosis.

Keywords: Heart neoplasms, Fibroelastoma, Stroke, Echocardiography

Fibroelastoma papilar mitral como causa de ictus en paciente joven

RESUMEN

El fibroelastoma papilar es un tumor poco frecuente, con una prevalencia estimada entre el 1 y el 7,9% de todos los tumores cardíacos primarios, los que —a su vez— presentan una muy baja incidencia (entre 0,001 y 0,28%), pues la gran mayoría de ellos (casi un 90%) son benignos. Se presenta el caso de un hombre de 41 años de edad que debutó con un accidente cerebrovascular. En el estudio ecocardiográfico se halló un tumor cardíaco en la valva anterior mitral sugerente de fibroelastoma papilar. Se decidió la intervención quirúrgica ante la posibilidad de complicaciones embólicas derivadas, con recurrencia de episodios isquémicos cerebrovasculares. Se realizó resección triangular de dicha tumoración y anuloplastia mitral con buen resultado. El estudio anatomopatológico confirmó el diagnóstico.

Palabras clave: Tumores cardíacos, Fibroelastoma, Accidente cerebrovascular, Ecocardiografía

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INTRODUCTION

Papillary fibroelastoma is an infrequent tumor with an estimated prevalence between 1 and 7.9% of all primary cardiac tumors¹ which—at the

same time— have a very low incidence (between 0.001 and 0.28%). The great majority of papillary fibroelastomas (almost 90%) are benign².

The case of a 41-year-old man with stroke of unknown origin following initial diagnostic tests is presented. Echocardiographic examination revealed a tumor on the mitral valve. The tumor was surgically removed and final diagnosis of mitral papillary fibroelastoma was confirmed by pathological study.

CASE REPORT

We present the case of a 41-year-old man with no known drug allergies, one-pack-a-day ex-smoker and occasional alcohol consumer with no family history of cardiocerebrovascular disease or coagulopathy who presented with ischemic stroke: left frontoparietal cortical cerebral infarction. Transcranial Doppler ultrasound of supra-aortic branches found no disorders. Right-to-left shunt study was negative at baseline and during Valsalva maneuver while the Leiden thrombophilia test yielded normal results for all mutations studied (factor V Leiden mutation and prothrombin G20210A).

The search for embolic source was followed by transthoracic echocardiography showing a homogeneous image over the mitral valve, without mitral regurgitation; hence, transesophageal echocardiography (TEE) and cardiac magnetic resonance were subsequently performed. The latter showed a nodu-

lar image of about 7 mm attached to the anterior leaflet of the mitral valve with poor definition by cardiac magnetic resonance.

The TEE (**Figure 1**) showed a thin leaflet mitral valve with preserved mobility and function, with no regurgitation. On the atrial side of the anterior leaflet (region A1-A2), an 8 × 8 mm rounded mass—well circumscribed, homogeneous, freely mobile, with no junctional pedicle or leaflet infiltration and not interfering with mitral valve kinetics— was visualized. The described mass was consistent with papillary fibroelastoma or, less likely, mitral valve myxoma. The rest of the structures were normal: intact atrial septum, with no masses found at the fossa ovalis; normal left atrial appendage blood flow and size, with no signs of internal occlusion; trivalvular aortic valve with thin leaflets, with proper function, mobility and blood flow.

Nondilated left ventricle with preserved systolic function and normal regional motility. Descending thoracic aortic segments without atheromatous plaques; no pericardial effusion and no myocardial masses.

In short, our case was a young patient with a history of ischemic stroke in whom a mitral valve mass with the previously described features was detected: cardiac tumor (possible fibroelastoma). In view of possible embolic complications and recurrence of cerebrovascular ischemic events, a meeting with the referral Department of Cardiac Surgery was arranged to evaluate the case and consider tumor excision

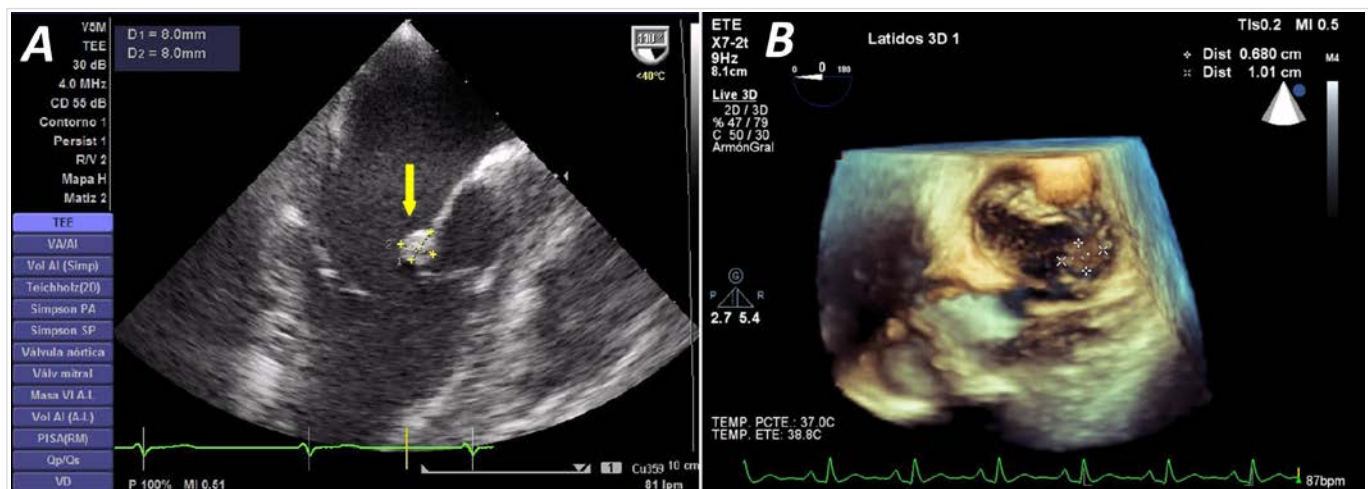


Figure 1. Transesophageal echocardiogram. **A.** Midesophageal long-axis view of the left ventricle. The nearly 8 × 8 mm mass is observed on the anterior mitral leaflet (arrow); it is not pedunculated, nor does it infiltrate the leaflet. **B.** Three-dimensional (3D) echocardiography. Left atrial visualization of the mitral valve was similar to that viewed during surgery. A mass on the atrial side of A1, —measuring approximately 10 × 7 mm— is seen.

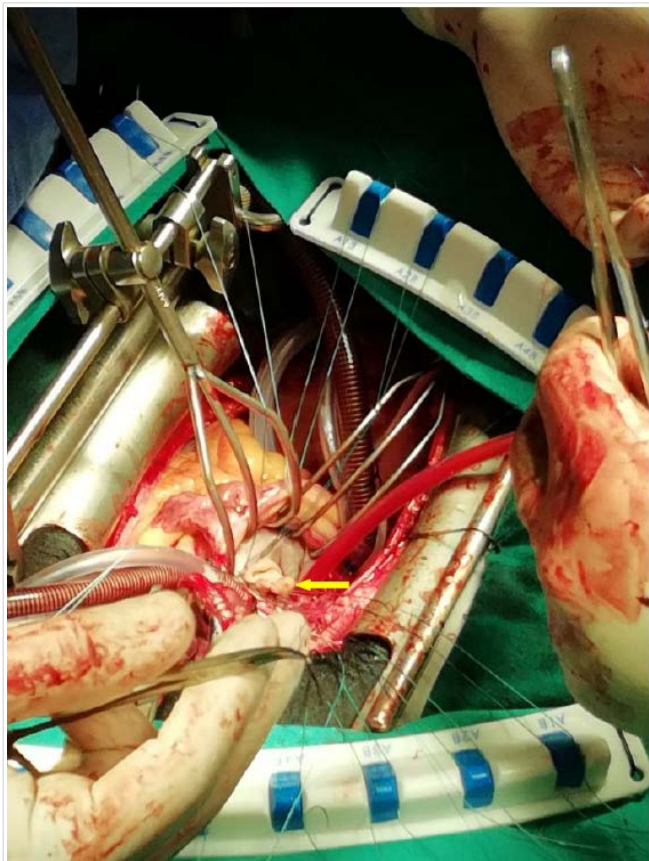


Figure 2. Surgical intervention: the well-demarcated rounded mass (arrow) is visualized on the atrial side of the anterior mitral leaflet (scallop A1).

and mitral valve repair.

Scheduled cardiac catheterization was performed as part of the pre-surgical study, which showed epicardial coronary arteries without angiographic lesions. The patient underwent surgery and a nearly 1 cm rounded tumor in segment A1 was found (**Figure 2**). Triangular resection of a portion of A1, including the tumor, closure of the surgically excised segment with 5-0 monofilament, and n° 34 ring annuloplasty were carried out. Intraoperative TEE control showed minimal mitral regurgitation. Postoperative recovery at the intensive care unit and later on the hospital ward was uneventful.

An echocardiogram was performed before hospital discharge which showed good outcome of mitral valve annuloplasty, with no significant diastolic gradient (mean 4 mmHg), at a heart rate of 65 beats per minute, and minimal central regurgitation. Pathological study of the mitral mass (**Figure 3**) reported: whitish, pedunculated, and fluffy lesion. Total inclusion in a paraffin wax block after hemisection. Final

diagnosis: papillary fibroelastoma (elastic hamartoma).

Outpatient follow-up has shown a favorable outcome. The patient has remained asymptomatic from the cardiovascular point of view without recurrence of cardiocerebrovascular events.

COMMENT

Papillary fibroelastomas are small benign cardiac tumors, typically smaller than 1 cm. In adults, papillary fibroelastomas are usually found on the aortic valve, and occasionally on the mitral valve, although they can be detected in any cardiac region. Valvular papillary fibroelastomas may involve any area of the valve leaflets; tending to recede from the cusps of the aortic valve and its corresponding edges, and from the mid-portion of the atrial side of the atrioventricular valves. Papillary fibroelastomas generally appear as solitary tumors, although occasional cases of multiple fibroelastomas have been reported³.

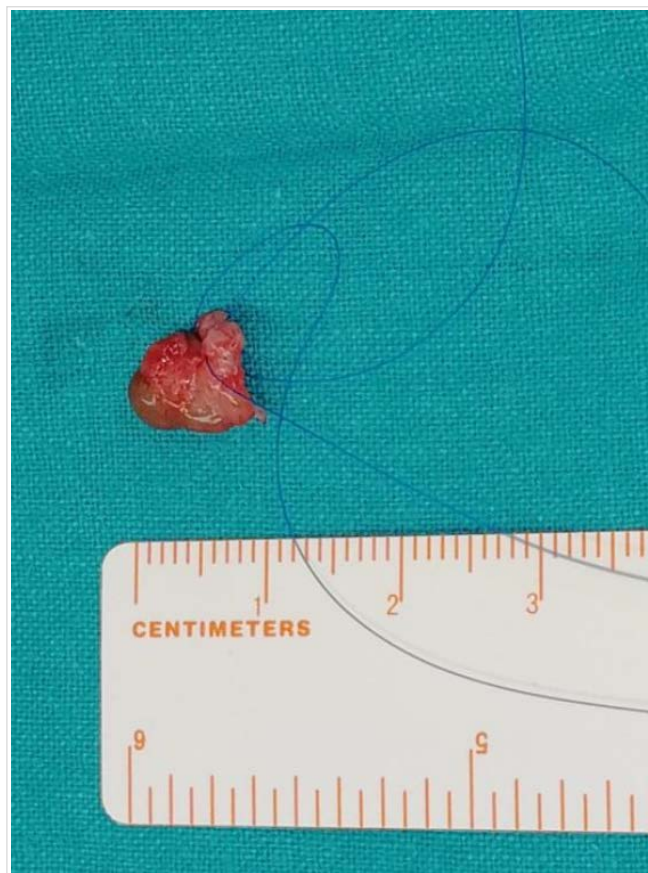


Figure 3. Surgical specimen: excised tumor mass.

Echocardiography, especially TEE, is considered the diagnostic method of choice given its greater ability to detect these tumors and rule out other possible embolic sources. Transesophageal echocardiography will show fibroelastomas as homogeneous, rounded, encapsulated, prolapsing masses attached to the valves⁴. Their variable size and mobility is considered an independent predictor associated with the occurrence of embolism and mortality⁵. The advantages of TEE versus cardiac magnetic resonance for assessment of possible cardiac masses on the mitral valve should be emphasized as these are highly mobile structures and therefore, echocardiography –with greater temporal resolution– will allow for better imaging.

Valvular vegetations (with lower echodensity and more heterogeneous appearance) and myxomas are important for the differential diagnosis, mainly due to their location; since myxomas are more likely to arise from the limbus of the fossa ovalis of the left atrium, while papillary fibroelastomas have a tendency to be located on the atrial side of the atrioventricular valves or the ventricular side of the semilunar valves.

Complete surgical resection is recommended due to the risk of embolic events (stroke, peripheral embolisms, myocardial infarction and even sudden cardiac death, as these tumors may cause blockage of coronary ostial openings). A higher frequency of embolic stroke in cases located on the mitral valve has been reported⁶. In our case we opted for surgical removal due to the risk of embolism associated with tumorization. Recurrence is unlikely if the tumor has been removed. No report has demonstrated the

benefits of anticoagulation in these patients, if there is no other indication for it⁷.

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