Multiple fibroelastomas of the aortic valve with mitral caseous calcification: Case report and review of the literature

Fibroelastomas múltiples de la válvula aórtica con calcificación caseosa mitral: reporte de caso y revisión de la literatura

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Eighty-six-year-old woman with a history of hypertension, mixed dyslipidemia, overweight, hyperuricemia, permanent atrial fibrillation, cardioembolic stroke in 2012, chronic renal failure, and ischemic heart disease with lower myocardial infarction, with no reperfusion, without the date being specified.

In June 2015, she experienced an episode of general malaise, asthenia, adynamia, nausea, vomiting, dizziness, and oppressive chest pain, which drove her to attend our institution. The electrocardiogram showed blocked atrial fibrillation that required temporary and later permanent pacemaker placement. Two-dimensional and three-dimensional transesophageal echocardiogram showed multiple fibroelastomas in the aortic valve, caseous calcification of the mitral valve (CCMV) with moderate mitral insufficiency and left atrial dilation without evidence of thrombi (Fig. 1).

During her evolution, the patient experienced Stokes-Adams episodes due to pacemaker malfunction and hypovolemic shock that required volume resuscitation and vasopressors. Three hours later, she developed abdominal pain, which was diffuse on palpation.

Abdominal ultrasound and transthoracic echocardiogram showed pericardial effusion that required pericardiocentesis. During the procedure, she developed cardiac arrest, and thus she was taken to an urgent pericardial window, and due to the persistence of bleeding, she underwent a chest examination for sternotomy. Two lesions were found on the right ventricular free wall that was repaired, unfortunately, the patient died.

Primary cardiac tumors are very rare. Fibroelastoma is a benign and avascular neoplasm, and it is the second most common cardiac tumor in adults, which affects the heart valves (aortic and mitral) in up to 80% of cases and is localized in the ventricular surfaces of semilunar valves and on the atrial surfaces of atrioventricular valves¹. These tumors have an appearance similar to sea anemones with multiple papillary frond-like projections attached to the endocardium by a short stem. They can occur with cerebral and heart embolism, acute valvular dysfunction, and sudden death²⁻⁴.

Our patient was in the ninth decade of life, and searching in the literature, we found two cases similar to ours regarding the age at diagnosis and localization

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Figure 1. Two-dimensional (2D) and three-dimensional (3D) transesophageal echocardiogram. **A:** 135° image showing multiple fibroelastomas in the aortic valve **B:** 3D image of the aortic valve in diastole with multiple fibroelastomas (red arrow). **C:** 3D image of the aortic valve in systole with fibroelastomas in the leaflets. **D:** 2D images and with color Doppler showing caseous calcification of the mitral annulus and moderate mitral regurgitation (red arrow). **E:** 3D atrial view of the mitral valve with caseous calcification. **F:** ventricular view of the mitral valve showing important calcification.

of the fibroelastomas: one, published in 2004, of a 77-year-old patient with two fibroelastomas in the aortic valve, who was brought to surgical resection, with good evolution both in the post-operative and in the follow-up period⁴; and another, published in 2007, of a 73-year-old woman whose pre-operative transesophageal echocardiogram showed an image consistent with an aortic valve fibroelastoma. The patient underwent surgical resection of the mass, but with a conservative approach to the valve and myocardial revascularization with the placement of a venous and an arterial graft⁵.

In our patient, transesophageal echocardiogram showed multiple images suggestive of fibroelastomas in the aortic valve, and CCMV. Retrospectively, it can be observed that the patient had multiple risk factors (female gender, old age, hypertension, and dyslipidemia) for having a stroke.

The risk of atrial fibrillation in patients with CCMV has been shown to be higher in comparison with those in sinus rhythm⁶. Two studies were analyzed; the first one, a cohort study, with a hazard ratio (HR) = 1.6 (95% confidence interval [CI]: 1.1-2.2), which was corrected with regard to atrial size, with a HR = 1.4 being obtained (95% CI: 0.9-2.0)⁷. In the second, the Multi-Ethnic Study of Atherosclerosis, a HR = 1.9 (95% CI: 1.5-2.5), was found,

which was consistent in all subgroups of age, gender, ethnicity/origin (Caucasians vs. non-Caucasians), hypertension, diabetes, and left atrial enlargement⁸.

The relationship between CCMV and stroke was described by Kizer et al.⁹, who found that the presence of CCMV is a strong risk factor for stroke.

The coexistence of fibroelastomas with stroke has been described by Anastacio et al.¹⁰, in their analysis of 23 patients operated for fibroelastomas over a period of 15 years, where most patients were symptomatic, with a 30% prevalence of embolic events. The association of aortic fibroelastomas with caseous calcifications is uncommon and increases the risk of stroke, atrial fibrillation, and sudden death.

Conflicts of interest

The authors declare that they have no conflicts of interest.

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Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors have obtained written informed consent of the patients and/or subjects mentioned in the article. The corresponding author is in possession of this document.

Right to privacy and informed consent. The authors declare that no patient data appear in this article.

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