

# An Unusual Association of Aortic Valve Fibroelastoma and Mitral Valve Stenosis

*Una asociación inusual de válvula aórtica Fibroelastoma y estenosis de la válvula mitral*

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## Abstract

The etiology of mitral valve stenosis is due to degenerative calcific disease. We report a case of a woman with an aortic supravulvar mass associated to a mitral valve stenosis with previous history of ischemic stroke.

**Keywords:** Mitral valve stenosis, Aortic valve, Echocardiography, Papillary fibroelastoma.

## Resumen

La etiología de la estenosis de la válvula mitral se debe a una enfermedad calcificada degenerativa. Presentamos un caso de una mujer con una masa supravulvar aórtica asociada a una estenosis de la válvula mitral con antecedentes de accidente cerebrovascular isquémico.

**Palabras clave:** Estenosis de la válvula mitral, válvula aórtica, ecocardiografía, fibroelastoma papilar.

## Introducción

Cardiac papillary fibroelastoma is a benign papilloma of the endocardial tissue. Surgical resection is recommended in symptomatic patients and when these tumors are found incidentally during preoperative evaluation.<sup>1</sup> With improved echocardiographic resolution, due to higher frequency transducers and new imaging modalities, small and ill-defined valvular lesions are increasingly recognized.<sup>2</sup> Papillary fi

## Case presentation

A 61-year-old female presents to the emergency department with symptoms of blurred vision, confusion and light-headedness. Upon admission to the hospital, she had also been experiencing chest pain, fatigue and progressive dyspnea to moderate efforts. She has history of an ischemic stroke two years before. The physical examination showed blood pressure of 94/60 mmHg, heart rate of 40-45/minute, respiratory rate of 20/minute and temperature of 36.5°C. On auscultation a bradyarrhythmic heart sounds with heart rate of 40-45 beats/min and diastolic murmur with opening click in mitral focus of grade II/IV intensity were detected. A right hemiparesis was found. The electrocardiogram shows atrial fibrillation with an average heart rate of 45 bpm and electric axis at +70°.

Transthoracic and transesophageal echocardiography showed severe mitral valve stenosis, mitral area of 0.92 cm<sup>2</sup> with the combination of two methods (pressure half-time and proximal isovelocity surface area), mild to moderate tricuspid regurgitation and normal pulmonary artery sys-

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tolic pressure (36 mmHg). Also a pedunculated and mobile aortic supra-valvular mass of 30 mm x 14 mm, which seemed to be adhered to the non-coronary cusp.

Computed tomography angiography corroborated the presence of a mass on the non-coronary cusp of the aortic valve, suggestive of papillary fibroelastoma of the aortic valve.

#### Discussion

Primary tumors of the heart are extremely rare. Papillary fibroelastoma is the most common benign tumor of heart valves and the third most common benign tumor of the heart, with an incidence of 0.33 % in autopsy series.<sup>3</sup>

It occurs in patients older than 50 years, although it can be found at any age.<sup>4</sup> Clinical manifestations include insidious, non-specific cardiac symptoms such as arrhythmias, precordial pain, dyspnea, syncope, pericardial effusion, amaurosis and cerebral vascular event, which occurs in 53 % of cases, being in many the only symptom.<sup>3,5,6</sup> Angina, myocardial infarction or sudden death may also occur, due to embolization. The probability of papillary fibroelastoma should be considered in patients with cerebral and cardiac embolic events without evident etiology.<sup>7</sup>

Four days later the patient underwent surgery for resection of the aortic valve mass and for mechanical mitral valve replacement. A large mass with speckled appearance and multiple papillary fronds attached to the non-coronary cusp of the aortic valve was found. A histopathological examination of the tumor revealed a benign cardiac papillary fibroelastoma. The patient's postoperative course was complicated with 3 episodes of bleeding and 3 days monitoring in the intensive care unit (ICU). A follow-up transthoracic echocardiogram at 5th day did not demonstrate aortic lesion and the prosthetic mitral valve was normal, also the pulmonary artery pressure. After 13 days the patient was discharged in optimal clinical conditions.

In the present case, a large mass was incidentally detected by echocardiogram. Echocardiography easily detects the pa-

pi- lary fibroelastoma, usually appearing as a mobile and pedunculated valvular mass; the superior temporal resolution of transesophageal echocardiography makes this technique, the definitive imaging modality for the diagnosis of this kind of tumors of the heart.<sup>8</sup> Preoperative transesophageal echocardiography is an important diagnostic tool that allows to define the location of the tumor and to assess the valvular abnormalities.<sup>9</sup> A papillary fibroelastoma should be evaluated by a noninvasive computed angiotomography, which permits also the characterization of the coronary anatomy and avoid its high cardiac embolic risk.<sup>10</sup> However, during the evaluation of this case an extremely large mass was detected, papillary fibroelastoma are typically small tumors of about 9 to 12 mm in diameter.<sup>11</sup> In addition to unusual features of the tumor in our patient, a combined pathology of mitral stenosis and papillary fibroelastoma was detected, which is extremely rare; only a few cases have been reported in the literature.<sup>12</sup>

Tumors of the left heart can cause ischemic stroke, which is the most common presentation of asymptomatic heart tumors.<sup>13</sup> Our patient had a medical history of ischemic stroke and her symptoms included pre-syncope, dyspnea and chest pain. As occurred in this patient, surgical resection is the treatment of choice for primary cardiac tumor in symptomatic patients.<sup>14</sup>

#### Conclusions

The association of aortic valve fibroelastoma with mitral stenosis is rare. The echocardiogram especially the transesophageal technique is the method of choice in the diagnosis of fibroelastoma. Surgical resection of the fibroelastoma is the treatment of choice when it is highly mobile, even in asymptomatic patients, because of its embolic risk.

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