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When a Cerebrovascular Accident Reveals a Cardiac Myxoma: A Clinical Case Report.

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ABSTRACT:

Primary cardiac tumors in adults are rare, but benign in 80% of cases. The myxoma is the most common benign primary tumor, typically located in the left atrium and, rarely, at the level of the heart valves. The main risk is associated with potential complications, particularly embolic events, including cerebrovascular accidents (CVA), which may reveal the tumor. Diagnosis is based on echocardiography and often cardiac MRI, with surgical excision being the sole therapeutic approach. We report the case of an 84-year-old female patient, admitted for management of a CVA, whose etiological workup revealed a myxoma located at the small mitral valve. The patient underwent surgical excision of the myxoma with uncomplicated postoperative recovery, and the myxomatous origin of the tumor was confirmed. We present this case with a literature review to detail the diagnosis and treatment of myxoma in the context of cardioembolic etiologies of CVA.

KEYWORDS: CVA, myxoma, cardiac tumor, mitral valve.

INTRODUCTION:

Although myxomas are benign tumors, their severity lies in the complications they cause, which can affect both the vital and functional prognosis. Neurological complications may include embolic infarctions, cerebral aneurysms, or meningeal or intracerebral hemorrhages [1]. The vascular embolus can originate from the tumor itself or from a thrombus on the tumor's surface. The discovery of a myxoma may, therefore, be secondary to the occurrence of neurological signs of cerebral ischemia in nearly 50% of cases [2]. The characteristics of the tumor on echocardiography and cardiac MRI may suggest a myxomatous origin, but only histopathology can confirm it following surgical excision.

Consent: The authors confirm that written consent for the submission and publication of this case, including the images, was obtained from the patient, in accordance with the guidelines of the Committee on Publication Ethics (COPE).

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LIST OF FIGURES:

Figure 1: Transthoracic echocardiographic views in the 5-chamber apical position showing a mobile element on the small mitral valve.

Figure 2: Transesophageal echocardiography images showing the mitral valve at different angles and cuts, highlighting the myxoma on the posterior mitral valve.

CASE REPPORT:

B.A., an 84-year-old female patient with cardiovascular risk factors including age and menopause, with no significant medical history, presented following the sudden onset of left-sided hemiparesis. Upon admission, the patient was conscious, hemodynamically and respiratory stable. The cardiovascular examination revealed a sinus rhythm at 76 bpm and a blood pressure of 94/72 mmHg, with a palpable point of maximum impulse, clear heart sounds without murmurs or added sounds, and no signs of right heart failure. Examination of the major vascular axes showed hyperpulsatility in

both carotid arteries without murmurs along their course. The neurological examination revealed left-sided hemiparesis, with muscle weakness in the left upper limb (3/5) and left lower limb (2/5), without sensory disturbances or dysarthria.

The patient was hospitalized and underwent a brain CT scan, which was consistent with an ischemic stroke in the territory of the anterior cerebral artery. As part of the etiological workup, the patient had an ECG and a 24-hour Holter ECG to assess for arrhythmia, which showed a regular sinus rhythm without any rhythm or conduction abnormalities. Regarding the Doppler ultrasound of the supra-aortic trunks, it revealed well-modulated flows and normal arterial velocities, without plaques or significant stenosis.

As part of the cardiac evaluation, the patient underwent a transthoracic echocardiography, which revealed a mobile element on the atrial side of the posterior mitral valve. Transesophageal echocardiography confirmed the presence of a multilobular mass attached to the atrial side of the small mitral valve, originating at its proximal portion, measuring 12mm x 5mm, suggesting a myxoma. The mitral valve showed mild regurgitation, and the interatrial septum appeared normal. The patient was then transferred to the cardiovascular surgery department and was admitted to the operating room.

After a median sternotomy and pericardotomy, cardiopulmonary bypass (CPB) was established with an aortic cannula and two venous cannulas, with the left heart vent placed through the right superior pulmonary vein. After initiating CPB and clamping the aorta, a left atriotomy was performed anterior to the pulmonary veins, which allowed visualization of a thickened mitral valve with a myxomatous-looking mass.

It was decided to preserve the native valve and perform a resection with biopsy of four fragments. The atriotomy was closed with two half-sutures of 4-0 prolene, followed by a gradual weaning from CPB. The CPB was successfully discontinued, and the postoperative course was uncomplicated, with a follow-up echocardiogram showing normal cardiac contractility and no mitral valve regurgitation or stenosis. Histopathological examination of the resected specimen confirmed the myxomatous nature of the mass.

Figure 1: Transthoracic echocardiographic views in the 5-chamber apical position showing a mobile element on the small mitral valve.



Figure 2: Transesophageal echocardiography images showing the mitral valve at different angles and cuts, highlighting the myxoma on the posterior mitral valve.



DISCUSSION:

Myxoma is the most common type of primary cardiac tumor [6, 8]. Its incidence in women is 2 to 3 times higher than in men, with an average age of onset around 50 years [7]. Myxomas primarily develop within the left atrium, often attached to the interatrial septum, and in only 20% of cases, they are located in the right atrium, and very rarely at the level of the mitral valve or in contact with it [1,6,10]. The majority of myxomas are solitary, but multiple myxomas have been described in familial forms (Carney complex), representing less than 10% of cases [5,11].

The histological origin of this tumor has not been easily determined, and several hypotheses have been suggested, including the development from preexisting thrombi [12]. The most widely accepted explanation is that the tumor develops from embryonic subendocardial pluripotent vestigial remnants that remain sequestered in the fossa ovalis of the interatrial septum [13, 14].

This tumor leads to a polymorphic clinical presentation, which may sometimes explain diagnostic delays. The most common mode of presentation is an embolic event or a valvular obstruction syndrome [4], or signs of heart failure [15,16]; their association in the form of a triad is also possible [5]. Embolic events occur in 30% of cases [15] and can be cerebral (in 50% of cases) or pulmonary for right-sided lesions. Other embolic locations, such as hepatic, coronary, and ocular, have also been reported [18,19]. Myxomas can cause arrhythmias and may be asymptomatic in 10% of cases [15].

Most myxomas are pedunculated and can obstruct diastolic ventricular filling when they prolapse through the mitral valve. The rest of the tumors are broad-based and sessile. They may be smooth, firm, lobulated, or friable and irregular, which significantly increases the risk of systemic embolism [5].

Echocardiography is the diagnostic modality of choice, with a sensitivity of approximately 93.3% and a specificity of 96.8% for transthoracic imaging [4]. Due to its superior temporal resolution, it not only allows visualization of mobile cardiac tumors but also enables the detailed description of their characteristics [5]. This exam can assess the mobility, base of implantation, size, and satellite lesions of the myxoma [1, 20]. It also helps predict the embolic risk of the myxoma by evaluating its morphology. Thus, villous and irregular polypoid tumors are more fragile and may carry a higher risk of embolization compared to smooth, regular tumors [21, 22]. The heterogeneous appearance is often associated with calcified or hemorrhagic areas [1]. Tumor size can range from a few millimeters to several centimeters, without a direct correlation to the embolic risk.

Transesophageal echocardiography plays an important role in characterizing the implantation site of the myxoma and provides a better assessment of atrial tumors [4,5], with a sensitivity of 97% [23].

Other imaging techniques can be used [24, 25], particularly cardiac MRI. This technique allows for good characterization of the tumor, which appears iso-intense on T1-weighted images, hyper-intense on T2-weighted images, and demonstrates late enhancement after gadolinium injection [1]. The heterogeneous nature of the late enhancement may suggest areas of necrosis, calcifications, hemorrhage, or small thrombi on the tumor surface that do not take up the contrast agent.

Cardiac CT can detect calcifications with a posterior shadow cone and help determine the anatomical relationship with adjacent structures [5].

The main differential diagnoses for myxomas include intracavitary thrombus, valvular vegetations, and other benign and malignant cardiac tumors, highlighting the importance of imaging and histopathological examination [1, 4].

The definitive treatment is surgical excision, which should be performed as soon as possible to avoid complications [2]. It must be done in a single block to prevent embolism due to fragmentation of the tumor mass [26]. The resection must be complete to prevent recurrence [4]. The valvular tissue is often preserved unless the tumor has a large base of implantation, in which case excision of part or the entire valve, followed by valvuloplasty or valve replacement, may be necessary. It has been shown that electrocoagulation of the base of the myxoma with resection of part of the healthy endocardium prevents recurrence [27].

Histopathological analysis of the resected mass confirms the diagnosis when the tumor appears gelatinous and friable, with lipid cells within a myxoid stroma rich in glycosaminoglycans [28].

According to several series, the surgical outcome is excellent, with an operative mortality rate of 1 to 2% [1, 29]. Postoperative morbidity is primarily related to the risk of tumor recurrence or the development of another myxoma, and even the occurrence of distant metastases. In fact, the frequency of tumor recurrence is around 2% [17, 30]. The recurrence risk is higher in familial forms, reaching 10%. The incidence of postoperative complications, such as arrhythmias or conduction disturbances, is observed in bi-atrial approaches and large excisions [31]. A resection via mini-thoracotomy with the use of video-endoscopy has recently been reported to reduce this risk [32].

CONCLUSION:

Myxoma is one of the main benign cardiac tumors. It is a rare pathology with a highly variable mode of presentation depending on its location. This tumor is often located in the left atrium, attaching to the interatrial septum; mitral valve involvement is very rare, which is characteristic of our patient. Embolic events, particularly cerebral, are one of the main complications that reveal the tumor and worsen its prognosis. The diagnosis is made through cardiac imaging, starting with transthoracic and transesophageal echocardiography, sometimes with the addition of cardiac MRI. The only definitive treatment is surgical, with or without preservation of the mitral valve when the tumor is attached to it. Excision is urgent and should be as complete as possible to reduce the risk of recurrence, which remains low at less than 2%, and to prevent serious complications

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