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ABSTRACT

Primary cardiac tumors are usually rare, within them the atrial myxoma is the most common. The presentation of a clinical case is made where the debut of presentation of atrial myxoma was with intermediate-risk pulmonary thromboembolism. The location site was at the level of the left atrium, with an integral interatrial septum, so paradoxical embolism was not the reason for the pulmonary embolic event. The presentation by primary thrombotic event by myxoma is unusual. A brief review of the literature on atrial myxoma is performed, with few cases where the reason for pulmonary thromboembolism is presented. Atrial myxoma resection was performed, with adequate clinical evolution and without complications during the perioperative period, with discharge due to patient improve- ment. The importance of the review of this clinical case plus the review of literature is key to suspect unusual sites of thrombotic events triggered by atrial myxoma.

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Pulmonary Thromboembolism as the First Manifestation of Left Atrial Myxoma. Case Report and Literature Review

Vergara Orduña Fernando Iván[α], Solis Mendoza Karla Ivonne[σ], Rivera Hermosillo Julio César[ρ], Vela Huerta Agustín [Ѡ], Peréz Moreno Israel David [Ѡ], Narváez Oriani Carlos Alfredo[Ѡ], González Coronado Vidal José[Ѡ]& Rivera Capello Juan Miguel [Ѡ]

RESUMEN

Tromboembolismo pulmonar como primera manifestación de mixoma auricular. Reporte de caso y revisión de literatura.

Los tumores cardíacos primarios son usualmente raros, dentro de ellos el mixoma auricular es el más común. Se realiza la presentación de un caso clínico donde el debut de presentación de mixoma auricular fue con tromboembolismo pulmonar de riesgo intermedio. El sitio de localización era a nivel de aurícula izquierda, con septum interauricular íntegro, por lo que el embolismo paradójico no fue el motivo del evento embólico pulmonar. La presentación por evento trombóticos primarios por el mixoma son poco usuales. Se realiza una breve revisión de literatura sobre el mixoma auricular, encontrándose escasos casos donde el motivo de presentación por tromboembolia pulmonar. Se realizó resección del mixoma auricular, con adecuada evolución clínica y sin complicaciones durante el perioperatorio, con egreso por mejoría de la paciente. La importancia de la revisión de este caso clínico más la revisión de literatura es clave para sospechar sobre sitios inusuales de eventos trombóticos desencadenados por mixoma auricular.

Palabras Clave: mixoma, auricular, tromboembolismo.

ABSTRACT

Primary cardiac tumors are usually rare, within them the atrial myxoma is the most common. The presentation of a clinical case is made where the debut of presentation of atrial myxoma was with intermediate-risk pulmonary thromboembolism. The location site was at the level of the left atrium, with an integral interatrial septum, so paradoxical embolism was not the reason for the pulmonary embolic event. The presentation by primary thrombotic event by myxoma is unusual. A brief review of the literature on atrial myxoma is performed, with few cases where the reason for pulmonary thromboembolism is presented. Atrial myxoma resection was performed, with adequate clinical evolution and without complications during the perioperative period, with discharge due to patient improvement. The importance of the review of this clinical case plus the review of literature is key to suspect unusual sites of thrombotic events triggered by atrial myxoma.

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I. INTRODUCTION

The presence of primary cardiac tumors is a rare nosological entity, the presence of cases tends to be dramatically where systemic embolism or sudden death is usually the first manifestation until the asymptomatic course of said entity where the diagnosis is made in a incidental. The debut

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with the presence of embolism is less common, the presentation of the clinical case is made where the initial manifestation was pulmonary thromboembolism as a thrombotic manifestation, with a subsequent bibliographic review of said pathology.

II. CLINICAL CASE

Female of 68 years of age, originary and resident of Mexico City, with no significant familiarly antecedents, only pathological personal history performing bilateral tubary ligation 30 years ago as a family planning method. With cardiovascular history unknown until current condition.

Began with sintomatology a previous month with the presence of volume increase at the level of the left pelvic limb as well as pain of intensity 3/10 on the VAS scale without attenuators or exacerbations, so it goes to an emergency department assessment of the second level Hospital , where deep vein thrombosis is documented, the diagnostic protocol is performed, a simple chest angioTAC is performed, where the presence of segmental pulmonary thromboembolism of the right pulmonary artery is documented without evidence in this hospitalization of dyspnea or desaturation. The image is evaluated where there is evidence of incidental findings in the left atrium, sending a corresponding third level unit for evaluation.

The physical examination of apparent age similar to the chronological, cooperative and oriented in its three spheres, with adequate coloration of teguments, without characteristic fascies, scleras with adequate coloration, hydrated oral mucosa, neck without yugular regurgitation data, normolyne thorax, movements of thorax conserved, respiratory noises with disseminated vesicular murmur, adequate voice transmission. Rhythmic precordium of good tone and intensity, S1 present, clean systole, S2 with physiological split, clean diastole, with no evidence of murmur or added noises. Globose abdomen at the expense of adipose panicle with normal active peristalsis, without visceromegaly, extremities with symmetrical, homocrotic and synchronous pulses with the carotid. Slight edema in the left + pelvic limb, without pain on palpation, with the presence of red venous.

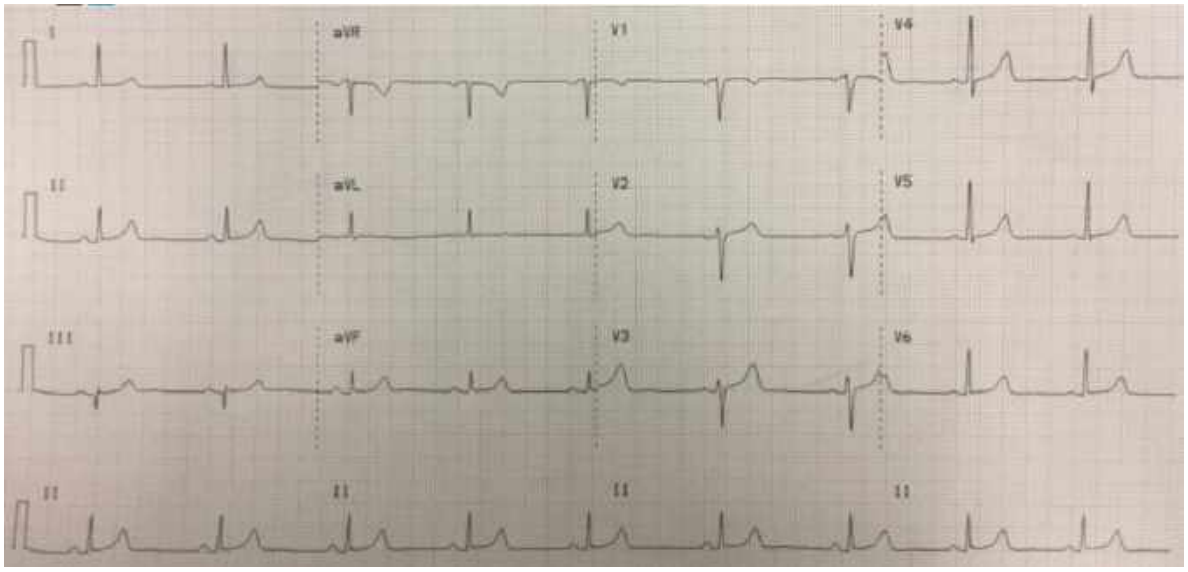

Fig 1: (12 lead electrocardiogram, sinus rhythm)

Transesophageal echocardiography is performed, showing cardiac tumor, probable left atrial myxoma, not pediculated and attached to the interatrial septum, with dimensions of 14 by 17 mm, which does not condition dynamic obstruction. Left ventricle of normal size with

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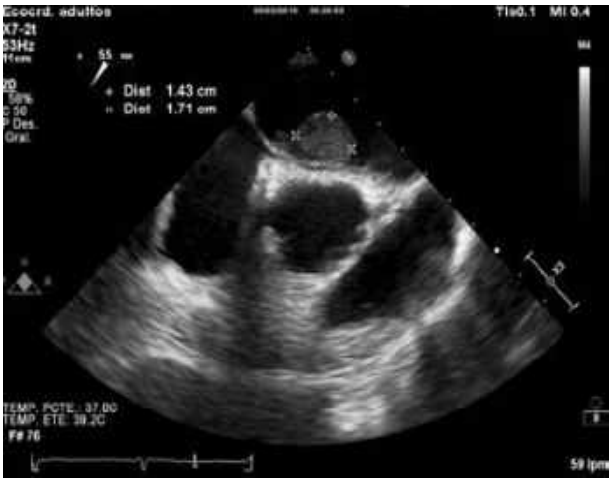
LVEF 67% Simpson Biplane. Diastolic function preserved. Non-dilated atria without thrombi. Right dilated ventricle with preserved systolic function. Mitral, pulmonary, aortic and tricuspid insufficiency all light grade. Low probability of pulmonary hypertension, PSAP 11 mmHg. Normal pericardium. Without thrombi or short intracardiac circuits.



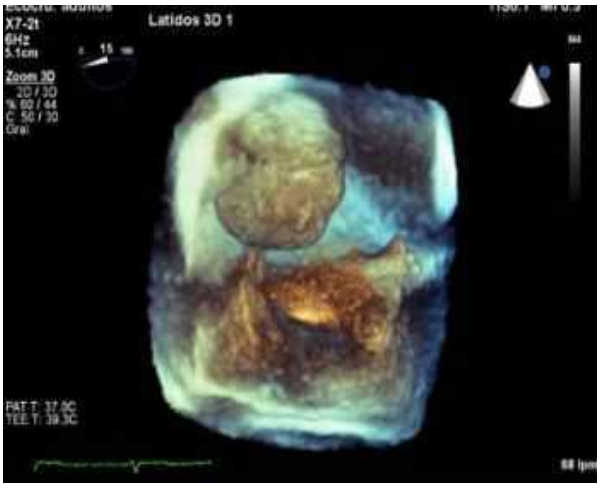
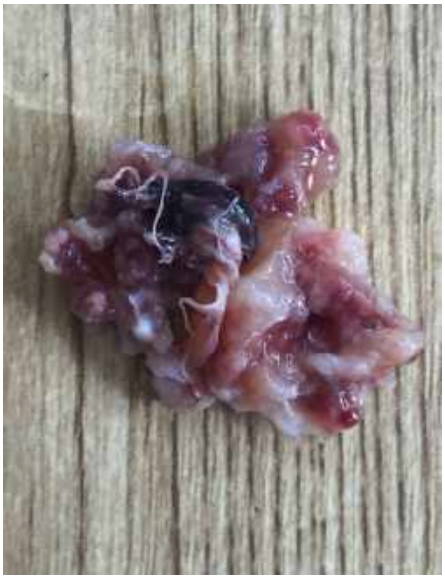


Fig. 2 and 3: Transesophageal echocardiogram in 2D and 3D mode respectively, showing a well-defined left atrial tumor of 14 by 17 mm.

Based on these findings, the presentation of the case to the surgical committee of our unit is decided, being accepted for tumor resection with a high suspicion of atrial myxoma diagnosis. This procedure is carried out without any complications, and a surgical piece is sent to the pathology department where histopathological diagnosis is confirmed for Atrial Myxoma.



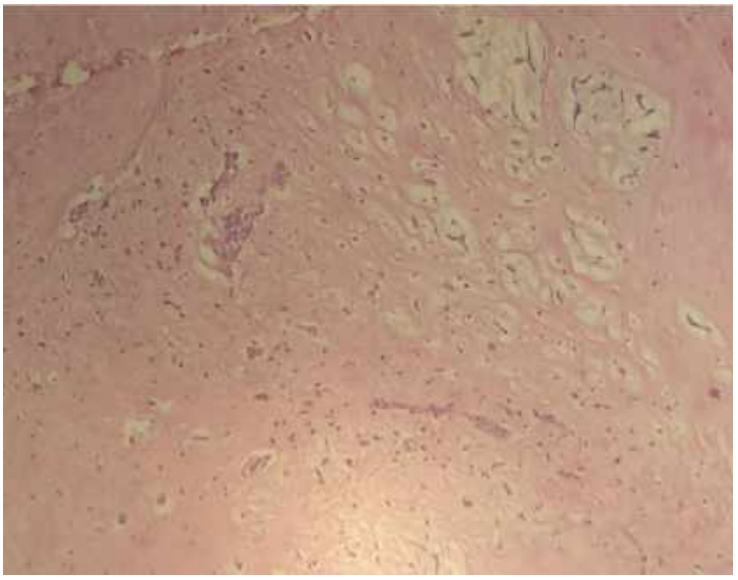


Fig. 4 and 5: Macroscopic piece, where evidence of a pink-brown sessile surgical piece is evident. The surgical piece was sent to pathology where polygonal cells are evidenced under staining.

2.1 Revision

The first description of cardiac myxoma was made in 1845 by King.[1] The presence of primary cardiac tumors is a rare entity in international reports, with an incidence between 0.0017 to 0.19% in autopsy studies in unselected patients. More than

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60% of the tumors are benign, of these, more than half are comprised of myxomas, the rest are comprised of rhabdomyomas, lipomas and fibroelastomas[10], have a clear predominance of presentation in adult patients, with a peak presentation between 20 to 50 years of age. With a distribution of incidence at a 1: 1 ratio between men and women.[2] In some cases they are associated with complex family genetic syndrome where tumor formation is the rule, the Carney complex is the largest syndrome identified. Multiple oncogenes have been identified that are related to the formation of benign cardiac tumors, mainly myxoma; the presence of c-MYC (oncogen), vimentin (mesenchymal-epithelial transition protein), p53 and HIF-1α (metabolic transcription factors associated with neoplasia); they have been related to tumor activity, development and even their recurrence, however it has not been demonstrated at the moment in large series of cases.[9]

2.2 Location

The ideal location site is at the endocardial level, where the left atrium is the main affected with 75% of the cases, being the *fossa ovalis* where they are located more frequently; In a smaller case, the right atrium affecting 15 to 20% of the cases (patient condition reported in the clinical case), the rest of the cases are distributed in ventricular location with less than 5%. [3]

2.3 Histological and macroscopic findings

The mytopathological study of myxoma shows that it is a tumor of endocardial and subendocardial origin, composed of multi-potential cells of mesenchymal line from which they can be distinguished: fibrocytes, myocytes or endothelium (histology). They consist of myxoid matrix composed of stromal-rich mucopoly-saccharides, with polygonal cells. As a characteristic finding of myxoma with eosino-philic cytoplasm: multinucleated cells can be found without mitotic activity. The macroscopic characteristics are generally polypoid, with an oval and pedunculated form; The mobility of the tumor within the cavity is dependent on the sessile appendix adhered to the endocardic wall and the amount of collagen that composes it, this characteristic also correlates with the consistency and emboligenic capacity of the tumor. [4]

III. CLINICAL MANIFESTATION

The clinical manifestations are varied, are dependent on the location as well as the friability of the tumor, causing; as previously mentioned emboligenic manifestations. The classic triad described in the reports are emboligenic pheno-mena, constitutional symptoms and cardiac obstruction, although in many cases patients are asymptomatic. Constitutional symptoms are secondary to the production of interleukin -6 for the same tumor, creating symptoms such as fever, malaise, arthralgia, rash and unintended weight loss. The same production of this cytokine[5] ,tends to cause anemia with normocytic normochromic pattern, in minor cases there may be thrombocytosis or leukocytosis; In rare cases there is Raynaud's phenomenon and digital hypocratism.

Embolism events occur in 30% of patients, the location of the tumor in the left atrium will have embolic events at the brain level, where obstruction by microemboli at the level of the retinal atherias can cause loss of transient or permanent vision. Embolism at the peripheral level can affect splanchnic circulation, abdominal aorta and renal arteries, with the manifestations of ischemic events in these organs and systems. The presence of right atrial myxoma can cause pulmonary embolism. [7]

Intracardiac obstruction is dependent on the size, friability and mobility of the tumor, an obstruction at the level of the tricuspid and mitral valve will lead to a syncope event, if the obstruction is not resolved, sudden death can develop. In case of permeable oval foramen or some other interatrial septum defect, it can cause paradoxical embolism.

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3.1 Physical exploration

The physical examination directed mainly to cardiac auscultation, must be performed dynamically so that the auscultatory findings of the tumor, appearance and disappearance with position changes is highly suggestive of atrial myxoma where the “tumor stroke” is the classic finding in myxoma. In case of Location in the left atrium can be confused with mitral stenosis, where the "tumor stroke" can be confused with an opening click and a presystolic rumble. On the other hand, the location in the right atrium can be confused with tricuspid valvulopathy. The findings can be searched intentionally for peripheral systemic and embolic manifestations. [12]

3.2 Diagnostic assistants

The use of electrocardiogram and chest radiography have been relegated to the most modern imaging methods, this is because the findings are nonspecific. Electrocardiographic abnormalities are few unless the tumor is altering the hemodynamics of the patient, while the chest x-ray may show atrial growth or data of venocapillary congestion. [10]

The echocardiogram has become the diagnostic assistant of choice in these patients, since its sensitivity and specified reach almost 100% in some case series. The transesophageal approach is the one of choice to be able to delimit the structures and characteristics of the tumor well, previously the 2D mode was the only one used, however the realization of 3D echocardiography entails a better clinical characterization of the tumor. [13]

Other diagnostic methods used are chest tomography and magnetic resonance imaging, which will be requested in case the echocardiographic findings are inconclusive and there is a high suspicion of atrial myxoma diagnosis. [11, 14]

3.3 Differential diagnosis

In the presence of an intracardiac tumor, a differential diagnosis must be made with primary or metastatic tumors, neoplasms with cardiac metastases are 20 times more common than primary cardiac tumors. The presence of intracavitary thrombus is another differential diagnosis that must be taken into account, however, this is done in the presence of alterations of cardiac mobility. Finally the cardiac vegetation as manifestation of infective endocarditis is another differential diagnosis that must be taken into account.

3.4 Treatment

The treatment of choice is surgical resection, which; Because it is pedunculated it is easy to remove. Carried out by sternotomy with subsequent cardioplegia so that the surgical bed is adequate. When performing the resection, a Dacron or bovine graft patch can be placed where the extraction was performed. One of the care that should be taken during the intervention is the prevention of tumor fragmentation with subsequent embolization. The prognosis is good, with a perioperative complication rate of less than 5% and a 3% mortality rate. In case of recurrence despite surgical treatment, as it could occur in polypoid or myxomatous syndrome with genetic alteration, surgery can be reconsidered, however there are reports of cases where recurrence can occur in more than three occasions cardiac transplantation. [8]

IV. CONCLUSION

Atrial myxoma is the most common primary cardiac tumor, particularly in this clinical case, in which the initial manifestation was the pulmonary embolic event; and in which the presence of permeable oval foramen was ruled out because the interatrial septum was integrated, this in view of the probability of a supposed trigger for a paradoxical embolism, this being ruled out. Therefore, the thrombotic origin could be a

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manifestation of the constitutional findings of the previously described tumor. The surgical procedure was performed with this patient successfully. The approach protocol is usually simple, however, there should be high suspicion to carry out an adequate diagnostic follow-up.

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