Casos clinicos

Right atrium myxoma in chilhood

Mixoma auricular derecho en la infancia

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Abstract: Primary cardiac tumours are rare and account for only 5% of all cardiac tumours. Seventy-five percent of these tumours are benign, and myxoma is the most common benign tumour in adults (50%). Most cardiac myxomas are located in the left atrium, only 18-20% are located in the right atrium, and the occurrence of a bilobed tumour is even rarer, the signs and symptoms with which they present are non-specific and vary according to their behaviour. We present a case with these characteristics in a 9-year-old school girl who was admitted to the Hospital del Niño Manuel Ascencio Villarroel.

Keywords: myxoma, cardiac tumor, right atrium.

Resumen: Los tumores cardíacos primarios son poco frecuentes y representan solo el 5% de todos los tumores cardíacos. El 75% de estos tumores son benignos, y el mixoma es el tumor benigno más común en el adulto (50%). La mayoría de los mixomas cardíacos se localizan en la aurícula izquierda, solo el 18-20% se localizan en la aurícula derecha, y es aún más infrecuente la aparición de un tumor bilobulado, los signos y síntomas con los que se presentan son inespecíficos y varían de acuerdo a su comportamiento. Presentamos un caso con estas características en una paciente escolar de 9 años de edad que ingresó al Hospital del Niño Manuel Ascencio Villarroel.

Palabras clave: mixoma, tumor cardiaco, aurícula derecha.

Primary cardiac tumors are rare, most are benign, myxomas are the most common cardiac tumors2 and account for 50% of these[1,2].

Myxomas can remain asymptomatic for a long time, but once diagnosed they must be removed[2] .

We present the case of a patient who presented a lobulated right atrial myxoma, with implantation in the right atrium that intermittently prolapses into the right ventricular cavity and who underwent surgery. The purpose of this study is to analyze the clinical presentation, the anatomopathological characteristics, and the postoperative results after surgical resection of right atrial myxoma.

Case presentation

A 9 year old school girl with no personal pathological history, who was asymptomatic until approximately one day prior to her admission, decided to go to the doctor for medical consultation when she began to experience abdominal pain starting in the epigastric region and radiating to the lumbar region of moderate to great intensity, continuous pungent type, accompanied by quantified thermal rises of 38°C and a persistent clinical picture and increased pain in the right flank region, anorexia, non-productive, non-cyanotizing and non-emetizing cough.. He presented pathological findings on physical examination, in relation to the respiratory aspect there was evidence of slight suprasternal retraction, pulmonary fields were auscultated with isolated cramp in the right subscapular region. In relation to the cardiovascular aspect; tachycardic with a heart rate of 128 bpm, blood pressure 100/70 mmHg, on auscultation with audible systolic murmur in the 4 auscultation foci with intensity II/VI. Complementary examinations with haemogram reporting leucocytes of 11,900 mm3 at the expense of segmented 84%, haemoglobin of 15 g/dl and haematocrit 45.3%; pCr of 61mg/ dL, arterial blood gasometry with compensated metabolic acidosis pH: 7.49; pCO2: 20.1; pO2: 89.4; HCO3 13; chest X-ray showing right basal alveolar infiltrates and cysuritis, suggestive of right basal pneumonic process. An abdominal ultrasound was performed, reporting findings compatible with bacterial endocarditis without ruling out atrial tumour (myxoma), liver with discrete periportal reinforcement, gall bladder with presence of biliary microlithiasis, basal pneumonia of the right lower lobe. Antibiotic coverage was indicated, and due to suspicion of pulmonary thromboembolism, enoxaparin was started and a D-dimer was requested, which reported 4763. An echocardiogram was requested (Figure 1) which reported an enlargement of the right cavity, a mobile heterogeneous mass measuring 8.5 x 6 cm at the level of the right atrium which prolapses intermittently towards the ventricular cavity compatible with myxoma, a cardiothoracic index of 0.6 mm and mild to moderate pulmonary hypertension.

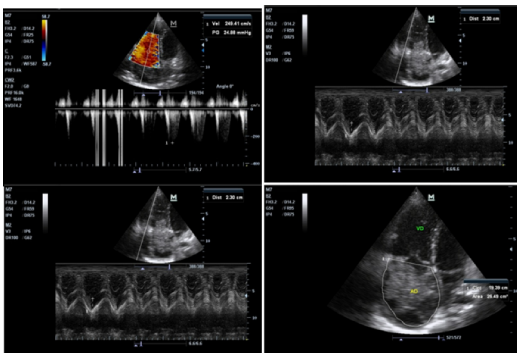

FIGURE 1.
Echocardiogram showing an 8.5 x 6 cm heterogeneous mobile mass in the right atrium.

She was assessed by a cardiovascular surgeon who carried out the diagnostic approach and requested a general CT scan to rule out possible dissemination. Based on the data obtained, it was decided to treat the basal pneumonia in the first instance, and when she was discharged by the pulmonology department, surgery was scheduled under extracorporeal circulation. A right atriotomy was performed and a large reddish-brown mass was observed occupying a large part of the right atrium, bilobed, with a yellowish second lobe, mobile, which penetrated the right ventricle through the tricuspid valve (Figure 2), with implantation in the interatrial septum.

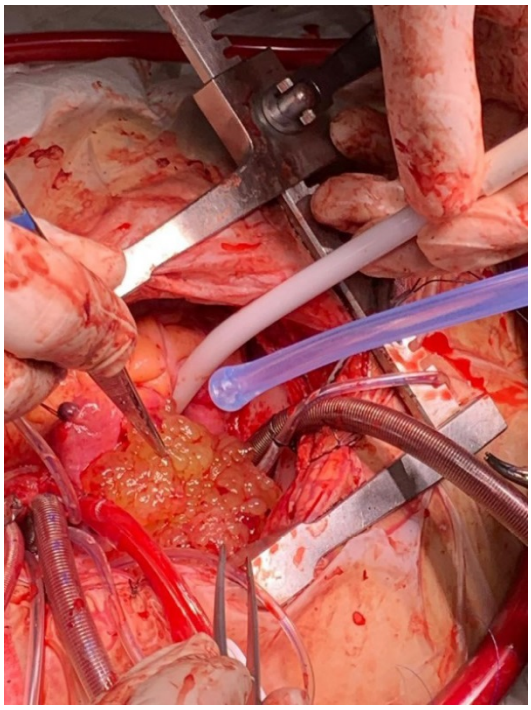

FIGURE 2.
The image shows the myxoma in the right atrial cavity during the trans-surgical period.

The mass was resected, including the segment of the septum where it was implanted and part of the posterior wall of the left atrium where the tumour infiltrated, the remaining defect in the posterior wall of the left atrium was repaired with direct suture and the interatrial septum was reconstructed with a pericardial patch (Figure 3). The patient was transferred to the intensive care unit with favourable postoperative recovery and was discharged in good condition.

FIGURE 3.
The image shows the surgical specimen, the size and weight of the myxoma, which has a length of 13 centimeters and a weight of 0.08 gram

Discussion

Primary cardiac tumours are rare and account for only 5% of all cardiac tumours; the remaining 95% are secondary or metastatic tumours. Of primary cardiac tumours, 75% are benign, and of these, 50% are myxomas, making myxoma the most common tumour in adults (and the third most common in paediatric patients, with 10% incidence); the other half corresponds to lipomas, rhabdomyomas, fibroelastomas and other rarer varieties[1,2,3].

Macroscopically, these tumours are usually polypoid, pedunculated, rarely sessile, with a soft, gelatinous consistency, of variable size, usually between 5 and 6 cm. Histologically they are characterised by a myxoid matrix, with typical polygonal cells with eosinophilic cytoplasm. Foci of calcification are seen in only 10% of cases[4].

Although they have been reported in neonates, and age ranges up to 95 years, they are most frequent between 30 and 70 years of age. There appears to be a small female predominance[5,6].

Myxomas may remain asymptomatic for a long time, but once diagnosed they must be removed and in some situations, pulmonary or systemic embolism, syncopal episodes or echocardiographic evidence of multilobular masses, surgery must be performed promptly.

Most patients present with at least one feature of the classic triad of cardiac obstruction, constitutional symptoms and embolic events. The size, postural changes and location of myxomas determine the clinical manifestations[9,11].

There is usually a single tumour (although multiple myxomas affecting more than one cardiac chamber have been described), and they are frequently pedunculated. They may be sporadic (>90%), recurrent or occur in association with other clinical manifestations, as in Carney syndrome. In 75-86% of cases they are located in the left atrium [6,7,11].

In our patient, the myxoma was located in the interatrial septum in the right atrium, infiltrating it and prolapsing into the right ventricle. Right atrial myxomas, in particular, can obstruct the tricuspid valve, causing symptoms of right heart failure, peripheral oedema, hepatic congestion or sudden death, which has been reported to be more common among patients with right atrial myxomas that protrude through the tricuspid valve, as is the case in our patient[10,13].

Systemic embolism may occur in 25-50% of patients with left-sided myxomas and more than half of the emboli are directed to the central nervous system, including the retina. In 30% of these patients, the neurological manifestations appear before the rest of the symptoms[7] .

The first M-mode echocardiographic diagnosis was made in 1959. Echocardiography is the diagnostic tool of choice, but it is impossible to define with certainty the type of tumour and whether it is a tumour or a thrombus. However, the size, fixation, mobility and prolapse of the mass across the heart valves can be defined. The sensitivity for diagnosing myxoma on transthoracic ultrasound is about 95%, and can reach close to 100% with transesophageal ultrasound[7,11].

The treatment of choice for cardiac tumours, especially myxomas, is surgical. The first surgical resection of a left atrial myxoma was described by Clarence Crafford in 1954, and since then the mandatory surgical nature of this disease has been established[6,8]. Although surgery is generally curative, recurrences have been reported up to 25 years later, with recurrences reported in 3-5% of cases of sporadic aetiology and 22% in cases associated with Carney syndrome [11,12].

Postoperative mortality worldwide is around 2.2% approximately[13] .

As it is an unusual presentation and its clinical features may go unnoticed or, as in the present case, an incidental finding in the presence of signs that may be mistaken for acute abdominal or lower respiratory tract symptoms, a comprehensive and rational approach is important in order to reach an accurate diagnosis.

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