CASE REPORT

Left ventricular myxoma in pediatric population: a rare presentation. A case report

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Cardiac neoplasms are rare, with myxoma being the most common benign tumor. Its most frequent location is in the left atrium. Typical clinical presentation is between the third and sixth decades of life, being more frequent in females. We report here the case of a 17-year-old patient diagnosed with thrombophilia secondary to protein S deficiency and acute lymphoblastic leukemia in remission. The patient developed data of acute heart failure, with a diagnosis suggestive by imaging of an intracardiac tumor in the left ventricle, causing left ventricular outflow tract obstruction. The tumor was successfully removed surgically.

Key words: Cardiac tumor; Myxoma; Left ventricle; Pediatrics.

Las neoplasias cardíacas son raras, siendo el mixoma, el tumor benigno más común. Su localización más frecuente es en la aurícula izquierda. Su presentación clínica predominante es entre la tercera y sexta décadas de la vida, siendo más frecuente en el sexo femenino. Reportamos aquí el caso de una paciente de 17 años de edad con diagnósticos de trombofilia secundaria a deficiencia de proteína S y leucemia linfoblástica aguda en remisión. La paciente presentó datos de falla cardiaca aguda, con diagnóstico sugestivo por imagenología de tumoración intracardiaca en ventrículo izquierdo, produciendo obstrucción al tracto de salida del ventrículo izquierdo. La tumoración fue removida quirúrgicamente con éxito.

***Palabras clave**: Tumor cardiaco; Mixoma; Ventrículo izquierdo; Pediatría.*

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Cardiac Myxoma are the most common primary tumor in the adult population. Presentation encountered in infants and children is rare. Though it is a benign tumor, behavior within the heart cavities can potentially make it clinically quite aggressive so that it would require immediate surgical treatment [1]. In general, cardiac myxoma is presented in 75-80% of cases in the left atrium, 20% in the right atrium, 3% in the left ventricle, and 3% in the right ventricle. It is more commonly presented in women. Overall presentation is typically seen between the third and sixth decade of life, although it can be present at any time of life [2]. We present herein a 17-year-old female patient with diagnosis of thrombophilia and history of cerebrovascular event in the past, who presented to the emergency room with heart failure due to a left ventricle myxoma. It is relevant to point out that this patient presented a stroke event in the past as well as the clinical history of the coagulation disorder as the cause. However, intracavitary heart tumors, specifically myxomas which are a slow-growing benign tumors, should always be ruled out as it an important cause of emboli. In this case, heart failure data and rapidly clinical deterioration was an important factor to search for cardiac related causes. In addition, imaging studies showed a tumor in the left ventricle which is a rare location to be presented in pediatric population.

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CLINICAL CASE

This is a case of a 17-year-old male with a diagnosis of thrombophilia secondary to protein S deficiency and acute lymphoblastic leukemia in remission, a history of cerebrovascular event with sequelae of hemiplegia and epilepsy. Patient developed acute heart failure including edema of the lower limbs, dyspnea on exertions. She was admitted emergency room. A transthoracic echocardiogram demonstrated a heterogeneous, lobulated, mobile, and pedunculated mass with irregular edges in the left ventricle of 12.8 cm by 5.3 cm by 2.33 cm in the anterosuperior wall that generates a dynamic obstruction to the left ventricular outflow tract. Pulsed Doppler shows flow acceleration of up to 40 mmHg, without evidence of pericardial effusion, suggestive of obstructive intracardiac tumor.

Diagnosis of intracardiac tumor with left ventricular outflow tract obstruction was confirmed with imaging studies, so emergency surgery was scheduled for cardiac exploration and

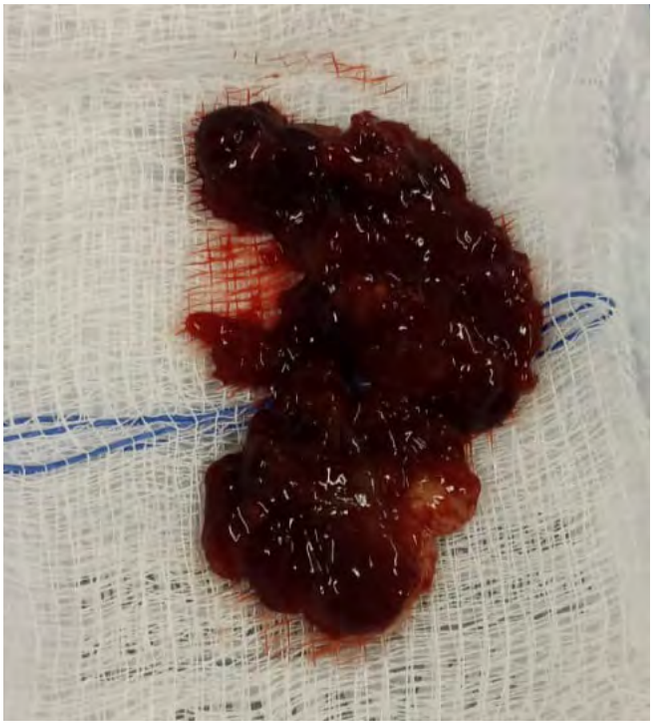

Figure 1. The image shows the resected intracardiac tumor, which has pedunculate and friable characteristics and jelly-like consistency.

resection. A surgical approach was performed through median sternotomy and cardiac exploration without evidence of extracardiac tumors. Extracorporeal circulation was established. An aortotomy was performed, and a tumor with myxoid characteristics of approximately 9 cm by 5 cm was identified that depended on the free wall of the left ventricle and protruded towards the aortic valve and partial occlusion of the mitral valve. A resection of the base was performed (**Fig. 1**). The rest of the procedure was uneventful and successful.

The histopathology report was consistent to cardiac myxoma. The patient presented an adequate surgical recovery and was discharged to continue ambulatory care.

COMMENT

In patients with benign myxoma-type tumors, the signs and symptoms are varied and non-specific depending on their behavior, as they may be asymptomatic or produce systemic or local effects and embolic phenomena [3]; cardiac symptoms (obstruction of the atrioventricular area) [4], general symptoms (fatigue, fever, myalgia, weight loss, Raynaud's phenomenon, skin lesions), it should be noted that the presentation of this patient is not found in any of the groups with higher incidence, by age or by location of the tumor, it is worth maintaining a broad criteria when faced with symptoms of probable cardiac origin to rule out a tumor. 90% of tumors in the pediatric population are benign in the sense that they are not invasive, although they can cause serious hemodynamic alterations due to their location [5]. Within the differential diagnoses in this age group are found in order of frequency: rhabdomyomas, fibromas, myxomas, teratomas, and sarcomas (the latter has as a characteristic that it is the most common malignant tumor, and the most frequently associated type is angiosarcoma) [6]. Within myxomas specifically, these tend to increase in frequency with age.

Clinical manifestations derived from the tumor is that when they occur they tend to affect the cardiac structures due to the location of the mass, to extrinsic compression and less frequent due to infiltration into the tissues. The most frequent clinical data are murmurs present during auscultation, as well as right ventricular dysfunction due to obstruction in the right ventricular outflow tract; pulmonary edema, low output and syncope in the case of obstruction of the left ventricular outflow tract, at earlier ages we can find data of cardiac tamponade due to complete extrinsic compression that could generate sudden death. All of them may suggest intrinsic or extrinsic compression. Further hemodynamic repercussions may appear as a result. High level of clinical suspicion and prompt therapeutic response are mandatory [6].

The electrocardiogram can provide with sensitive and relevant information on the location of the lesion, such as the deviations of the axis of the QRS complex. Chest X-ray can be useful when showing an increase in the size of the cardiac silhouette or an extrinsic growth at the expense of extracardiac structures. However, all of them are non-specific findings.

The transthoracic echocardiogram is considered the fundamental key for the diagnosis of this condition, since the cause of the obstruction can be visualized under direct vision, in case of heart failure, although its definitive treatment will be surgical excision and the definitive diagnosis will be due to histopathological results.

The recurrence rate after resection is 4-7%. Recurrences are due to inadequate resection, recurrence in multiple areas such as in familial cases, or associated with endocrine abnormalities. This these cases, the mortality rate is greater than 50% [6]. Controls are recommended at least every six months, which should be more frequent in cases of tumors with hemodynamic significance or mild arrhythmias that are not amenable to surgical treatment at that time.

Due to the fact that these tumors are not so frequently found in these patients, high level clinical suspicion should be taken into consideration. In this case, due to the clinical characteristics of this patient, the probability of having an intracavitary thrombus should be considered. Also, the same is applicable because of the history of hypercoagulable condition for thrombophilia background. The diagnostic suspicion was oriented towards an intracavitary mass or thrombus, given this background and the high suspicion of a probable thrombus, the diagnosis of the intracardiac tumor was facilitated, for which it is suggested to take these differentials into account. Although the age group is not so specific, it is recommended to perform imaging and paraclinical studies in case of clinical data suggestive of low output or incipient acute heart failure.

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