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Case Report

Right Ventricular Obstructive Myxoma

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Abstract

The cardiac tumors are infrequent in pediatric patients. Intracavitary tumors generally cause more symptoms and different complications are described. I present a case of prolapsing tumor into the pulmonary trunk causing severe dynamic obstruction.

Keywords: Myxoma; Right out flow; Obstruction

Case Report

Female 17-years- old, without previous hospitalizations. 6 months ago symptoms of dyspnea of medium effort, chest pain with exercise. She was referred to the Cardiology Department for cardiac murmur work up. The patient was in good general condition. On physical examination she had normal heart rhythm, normal first and second heart sound, normodynamic precordial activity, sistolic murmur in pulmonary focus with vertical irradiation. Electrocardiogram: in normal sinus rhythm, QRS right axis deviation +120°, Late R wave in aVR, abnormalities in QRS and abnormal repolarization in V1 (Figure 1). Chest X-ray was normal. Transthoracic echocardiogram showed: cardiac tumor with irregular borders adhered to right ventricular outflow tract on infundibular septum, pedunculated and mobile. In systole the tumor prolapse into the pulmonary trunk causing severe dynamic obstruction with maximum gradient of 71 mmhg (Figures 2 & 3). The study was completed with transesophageal echocardiography, this one not found secondary tumors (Figure 4). She was referred to urgent surgery. The size tumor was 4.5 x 6 cm. Surgical resection without consequences and the histopathology confirmed myxoma.

Discussion

Primary cardiac tumors are rare heart disease, constituting between 0.001% and 0.028% of autopsy studies [1]. Most are of mesenchymal origin, with a broad spectrum of tissue differentiation. 75% are benign and 25% malignant. The benign tumors in adults in order of frequency are: myxomas 50%, followed by lipomas, papillary fibroelastomas and rhabdomyomas. They have a 0.17% frequency in pediatric patients [2] and in order of frequency they are: rhabdomyomas 45-75 %, fibromas 6-25 %, and myxomas 5-10 % [3,4]. Symptoms depend on their location (pericardium, intramural or endocavitary), the chamber involved, their size and their infiltrative nature. Patients may be asymptomatic or debut with sudden death, pulmonary or systemic thromboembolic events, direct invasion of the myocardium causing altered contractility, arrhythmias, heart block and pericardial effusion, ischemia secondary to compression of the coronary arteries, heart failure, valve obstruction in the ventricular inflow or outflow tract, and respiratory symptoms due to compression of adjacent structures; rarely, they present with weight loss and fever [5]. Intracavitary tumors generally cause more symptoms than intramural ones. Up to 47% of cases have electrocardiographic abnormalities [6]. Chest X-ray is usually normal or show cardiomegaly. Diagnosis by transthoracic or transesophageal echocardiogram is sufficient, with a 93% and 97% sensibility, respectively [7,8]. A complete echocardiographic study should report:

1. Morphology: border characteristics irregular, smooth, rounded, presence of foci of calcification, bleeding.

2. Location: relationship to important structures, valves, coronary arteries, pulmonary veins.

3. Insertion site: septum, free wall, valves, atrium.

4. Mobility: pedunculated, with or without independent movement with adjacent myocardial contraction, prolapse into cavities or large vessels.

5. Hemodynamic impact: valve, ventricular inflow and outflow tracts, large vessel and pulmonary vein entry doppler studies, and presence of pericardial effusion.

Magnetic nuclear resonance gives a precise evaluation of the location and functional impact of cardiac masses in any image plane. Compared with echocardiography, it is more precise in pericardial tumors with or without extension to adjacent structures [9].

Myxoma is the most frequent primary tumor in adults, and is usually diagnosed between 30 to 60 years old. It may appear at any age including newborns, and it may even be diagnosed prenatally [10]. It is predominantly seen in females (70%) [11-13]. There are three clinical presentations: sporadic, complex and familial. The sporadic form is the most common (90%), being a single tumor with a less than 1% recurrence [14]. The complex form is characterized by multiple growths, associated cutaneous lesions such as lentiginosis or pigmented nevus, breast myxoid fibroadenomas, pituitary adenomas, Cushing´s syndrome, and rarely testicular tumors. It has a greater

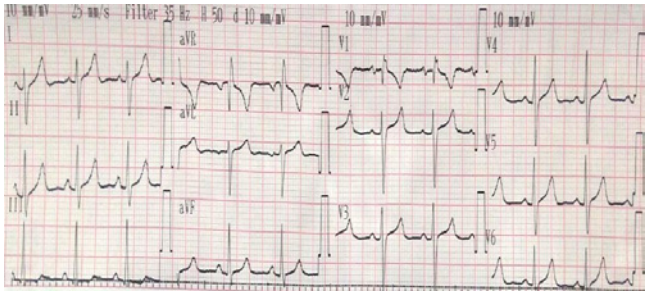

Figure 1: Electrocardiogram showed right axis deviation +120°, negative T wave in V1 and late R wave in aVR.

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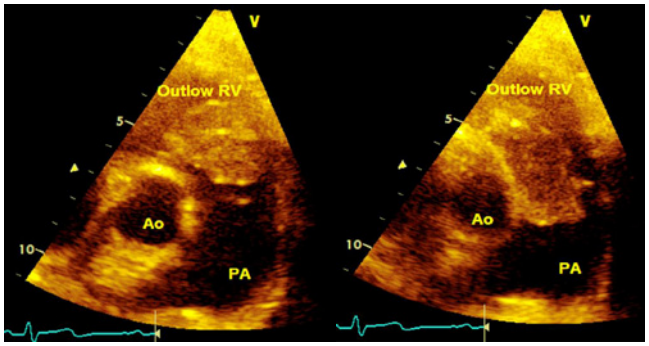

Figure 2: Short axis view. A) Diastole, tumor adhered to right ventricular outflow tract on infundibular septum. Ao: Aorta; PA: Pulmonary artery; *: Closed pulmonary valve. The Same View. B) Sistole, tumor prolapse into the pulmonary trunk.

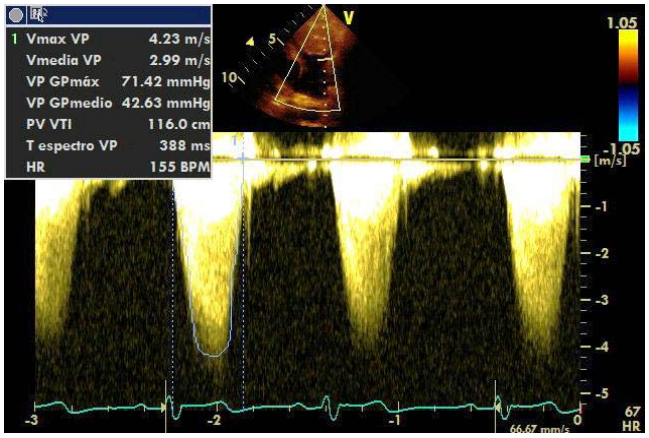

Figure 3: Transthoracic continuous doopler echocardiogram showed severe dynamic obstruction with maximum gradient of 71mmhg.

recurrence at 20%. The familial form7% of cases is autosomal dominant, affecting young people. It has a recurrence of 10%. It is part of the Carney complex: acromegaly, gigantism, pituitary adenomas and hyper pigmentation of the face, trunk and lips. Other familial forms are the NAME (Nevus, Atrial myxoma, Myxoid neurofibroma, Ephelides) syndrome, and the LAMB (Lentiginosis, Atrial Myxoma, Blue nevus) syndrome [15]. They are found in order of frequency in the left atrium (75-86 %), right atrium (20%), right ventricle (5%), left ventricle (3%), and less frequently in the aortic valve and inferior vena cava [11-13]. They are rapidly growing tumors (1 to2 grams/month), with a usual size of 4-8 cm, although they have been reported at up to 15 cm. They are masses with a grayish, soft, gelatinous, mucoid appearance, frequently having areas of bleeding or thrombosis. They are mobile or fixed pedunculated tumors. This case is of the sporadic variety: the tumor was single and large, arising from a stalk or pedicle on the infundibular septum, which gave it great mobility and made it prolapse during systole into the pulmonary trunk, causing severe dynamic stenosis which increased with exercise. It manifested as dyspnea on exertion, and the chest pain could have been secondary to left coronary compression due to the proximity of the tumor. There was a high risk of embolism and sudden death due to its size, which could occlude the whole pulmonary trunk, thus requiring emergency surgery. There was no history of arrhythmias or cardiac

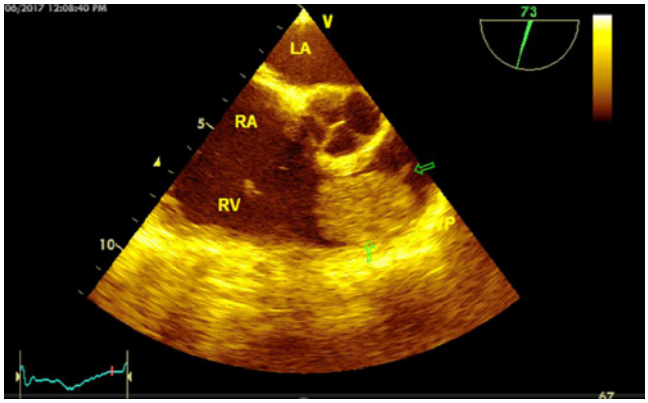

Figure 4: Transesophageal echocardiography study: Right ventricle and outflow views, this one not found secondary tumors. LA: Left atrium; RA: Right atrium; RV: Right ventricle; Green arrows show tumor.

emboli. Surgical resection was easy. The tumor was found to also be slightly attached to the ventricular surface of the pulmonary valve and it was freed without consequences. As reported in the literature, these tumors are unlike other benign growths such as rhabdomyomas, which are more frequent in the pediatric population, are usually multiple, may spontaneously regress through apoptosis, and rarely have complications. Myxomas have more complications, do not have spontaneous regression and require surgical resection, as in this case. The sporadic variety has little recurrence and a good long-term prognosis. Paladino et al. reported 89 cases of tumors in patients under the age of 18, achieving complete resection in 62 cases (69.7%), incomplete resection in 21 (23.6%), and four cases (4.5%) requiring heart transplantation [16].

Conclusion

Myxomas are uncommon in pediatric patients. These complicate more easily. In the presence of cardiac murmur it required to perform an echocardiographic study, which has sensitivity in more than 90% of the cases for diagnosis. Serious complications such as obstruction of ventricular outflow or inflow tract are a surgical urgency because of the high risk of sudden death or embolic events.

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