Journal of african clinical cases and reviews / Journal africain des cas cliniques et revues

www.jaccrafrica.com ISSN 1859-5138 Open access

Original article

Large myxoma of the right cavities in children: a clinical case report withliteraturereview

Large myxome des cavités droite chez l'enfant : cas clinique avec revue de littérature

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Abstract

Myxomas are the most common primary cardiactumors, mostfrequentlyfound in the left atrium. We report the case of a large myxoma of the right atrium in a 6-year-old child, extending to the right ventricle and to the trunk of the pulmonaryartery and complicated by bilateralpulmonaryembolism. The right localization is rare and uncommon in children.

Cardiac scanner and echography are the key examinations for the diagnosis as well as the anatomopathological examination.

Keywords: Atrial Right Atrial myxoma, right ventricular myxoma, heart failer, pulmonary embolism.

Résumé

Les myxomes sont les tumeurs cardiaques primitives les plus courantes, le plus souvent trouvées dans l'oreillette gauche. Nous rapportons le cas d'un volumineux myxome de l'oreillette droite chez un enfant de 6 ans, s'étendant au ventricule droit et au tronc de l'artère pulmonaire et compliqué d'une embolie pulmonaire bilatérale. La bonne localisation est rare et peu commune chez les enfants.

Le scanner cardiaque et l'échographie sont les examens clés pour le diagnostic ainsi que l'examen anatomopathologique.

Mots-clés : Myxome auriculaire droit, myxome ventriculaire droit, insuffisance cardiaque, embolie pulmonaire.

Introduction

Right atrial myxomaremains a rare pathology whose clinicalpresentationis not veryspecific, most often revealed by signs of heartfailure.

The prognosisremainsdominated by the occurrence of complications, the most frequent of which are pulmonaryembolism, or obstruction of the tricuspid valve.

Myxomasrarely affect children.We report the case of a large myxoma of the right atrium in a 6-year-old child, extending to the right ventricle and the trunk of the pulmonary artery and complicated by bilateral Pulmonaryembolism.

Clinical case

This is a 6-year-old patient with no history who underwent surgery following the fortuitous discovery of a mass in the right atrium extending to the right ventricle and to the trunk of the pulmonary artery on echocardiography prompted by a dyspnea of recent installation and progressive aggravation.

The clinical examination shows diastolic rolling at the tricuspid focus, with no signs of heart failure.

The chest x-ray: shows cardiomegaly with supra-diaphragmatic tip (ICT at 0.58), the ECG registers a sinus tachycardia with CVM 127.

Biological examinations did not find any inflammatory syndrome (C-reactive protein and normal BNP).

Trans thoracic echocardiography shows the presence in the right atrium of a homogeneous polylobed tumor mass, with a wide implantation base measuring 10 /4cm, wedged into the tricuspid orifice and protruding into the pulmonary infundibulum. With Grade I Tricuspid Insufficiency and normal lung pressures.

A left ventricle of normal size and systolic function (LVDTD at 31mm/LVDSD at 20mm, ejection fraction: 62%)

Non-dilated right ventricle, of good systolic function.

Chest CT showed a right heart mass extending to the trunk of the pulmonary artery with bilateral pulmonary embolism.

The patient was operated under extracorporeal circulation established between the aorta and the two-vena cava. The surgery was performed via the right auricular approach. Intraoperative exploration found a double myxoma of the right atrium and the left atrium with interatrial septum (AIS) and interventricular septum (IVS) intact and tricuspid leak on the water test.

The gesture consists of a resection of the myxoma of the right cavities with suture. Duration of extracorporeal circulation: 127 min, aortic clamping: 87 min and circulatory support: 36 min.

The immediate postoperative follow-up was marked by a complete AV block requiring definitive equipment.

Stay in intensive care unit: 48 hours. Duration of intubation: 12 hours. Duration of postoperative stay: 7 days. The patient is discharged on the 8th day.

The anatomopathological study came back in favor of a myxoma of the right cavities, with absence of histological signs of malignancy.

Control echocardiography performed on the 1st, 3rd, 6th and 9th months did not show any cardiac recurrences.

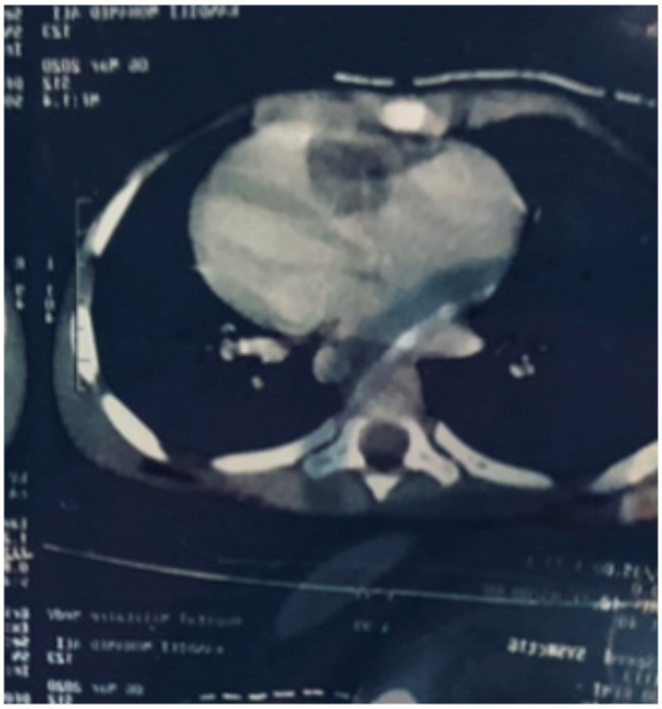

Figure 1: CT appearance of myxoma of the right cavities.

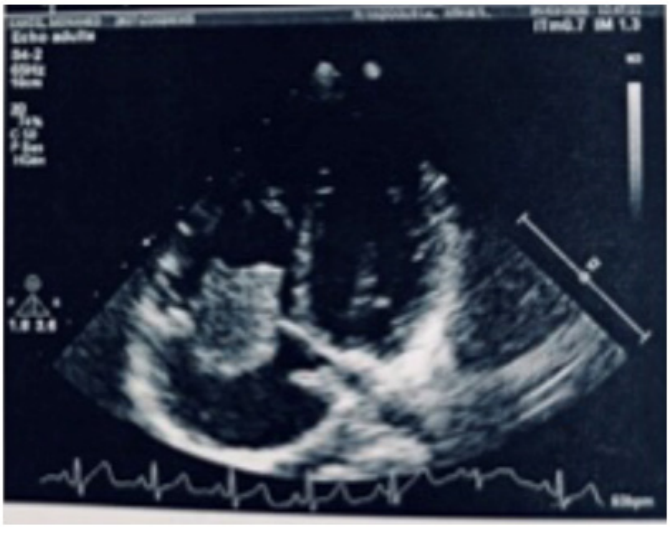

Figure 2: Sonographic appearance of large right chamber myxoma.

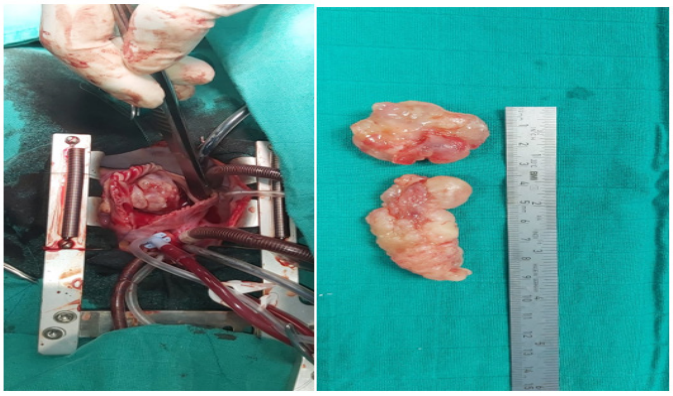

Figure 3: Operative view of the mass of the right cavities. Macroscopic appearance of the large myxoma.

Discussion

Atrial myxoma represents more than half of the Primary intracardiac tumors in adults [1]. The localisation in the right atrium is rare, found in 20% of cases.

The average age of revelation is 50 years old [2] with a female predominance.

Most often single localized, multiple localizations are more frequently found in familial forms which constitute less than 10% of myxomas [3]. Myxoma in children is rare, often associated with endocrine syndromes such as the Carney complex.

The usual mode of clinical revelation [4–8] is the appearance of signs of heart failure in 70% of cases. Right atrial myxoma is most frequently revealed by pulmonary embolism [4]. There is also a risk of enclaving the tumor in the tricuspid valve which may be responsible for syncope.

Cases of tamponade have also been described [9].

Embolic events occur in 30% of cases [4] (cerebrovascular accident, pulmonary embolism for right locations), other embolic locations (liver, eye, coronary arteries) have also been reported [10].

Transthoracic echocardiography remains the gold standard for diagnosis.

The myxoma presents as a mobile mass, rounded, polylobed or smooth, pedunculated or with a broad base of implantation, hanging most often at the level of the inter auricular septum, of heterogeneous appearance due to calcified and hemorrhagic zones.

Cardiac CT plays a vital role in studying the relationship between tumors and adjacent structures, in particular the tricuspid valve and the AIS, as well as the extension of the tumor into the pulmonary tract.

Cardiac MRI shows a heterogeneous appearance isointense in T1 mode, hyperintense in T2 mode, with Heterogeneous contrast uptake during gadolinium injection.

Histological examination confirms the diagnosis, showing on inspection a gelatinous and crumbly appearance, in addition to calcified and hemorrhagic areas.

The diagnosis is based on the observation of lipid cells within a myxoid stroma rich in glycosaminoglycans [6].

The main differential diagnosis of myxoma is thrombus. In 1 to 5% of cases, a recurrence or the appearance of a second myxoma has been reported after resection of an initial myxoma [11].

Recently, a mini thoracotomy approach with the use video endoscopy has been reported [12].

Conclusion

Right atrial myxoma remains a rare pathology, the mode of revelation of which is dominated by heart failure and pulmonary embolism. The attack of the child is most often part of the Carney syndrome with multiple localizations.

Transthoracic echocardiography remains the examination of choice for making the diagnosis, with histological confirmation.

Treatment is based on urgent and complete surgical excision to avoid complications and recurrences.

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Available online : May 31, 2022

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Conflict of interest : None

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To cite this article :

S Bellouize, N Loudiyi, A Meskine, A Benyass. Large myxoma of the right cavities in children: a clinical case report withliteraturereview. Jaccr Africa 2022; 6(2): 327-330