Calcified inferior vena cava and right atrial myxoma in an 18-month-old male

A case report

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Abstract

Introduction: Cardiac myxomas are the most frequent primary cardiac tumor in adults. The incidence in pediatric patients is extremely low. Heavy calcification of an atrial myxoma is uncommon in children.

Case presentation: An 18-month-old boy was admitted for a significant precordial systolic murmur. Transthoracic echocardiography revealed a cardiac mass extending from the inferior vena cava across the right atrium and tricuspid valve into the right ventricle with severe tricuspid regurgitation. According to the echocardiography result, the patient was diagnosed with an inferior vena cava and right atrial tumor with tricuspid regurgitation. After the diagnosis, the patient underwent removal of the tumor via median sternotomy. The mass was removed under cardiopulmonary bypass with deep hypothermia circulatory arrest. The tricuspid valve was repaired by valvuloplasty and annuloplasty. The postoperative recovery was unremarkable. Follow-up echocardiogram at 1 month revealed moderate tricuspid regurgitation without myxoma recurrence.

Conclusion: Heavy calcification of an atrial myxoma is uncommon especially in children. Definitive therapy for myxomas requires prompt surgical excision and long-term follow-up is recommended in children although recurrence after excision is rare.

Abbreviations: IVC = inferior vena cava, RA = right atrium, TTE = transthoracic echocardiography.

Keywords: calcification, inferior vena cava, myxoma, pediatric, right atrial

1. Introduction

Cardiac myxomas are the most common primary neoplasm of the heart. About 80% of myxomas are within the left atrium.[1] Calcification appears to be more frequent in the right atrium rather than in the left atrium and is present in 10% to 20% of myxomas.[2] Heavy calcification of an atrial myxoma is uncommon in children. We recently cared for a previously healthy 18-month-old male who was found with a significant systolic heart murmur without associated symptoms.

The patient provided informed consent for the publication of his clinical and imaging data. This case report was approved by Medical Ethical Committee of Hunan Children’s Hospital.

An 18-month-old boy who was found with a significant systolic heart murmur was admitted to our hospital. He had no symptoms and there was no family history of cardiac disease. Transthoracic echocardiography (TTE) revealed a cardiac mass extending from the inferior vena cava (IVC) across the right atrium (RA) and tricuspid valve into the right ventricle. The sessile mass sized 41 × 10 mm and was extensively calcified (Figs. 1 and 2). The tricuspid was severely regurgitated. According to the echocardiography result he was diagnosed with an IVC and RA tumor with tricuspid regurgitation. Physical findings showed a regular pulse of 122 beats/min and his blood pressure was 86/52 mm Hg at admission. There was a significant systolic murmur over the right sternal border at the third intercostal space. Laboratory evaluation showed no abnormalities. Chest x-ray revealed cardiomegaly with an oval radiopaque mass within the cardiac silhouette. Electrocardiogram revealed a normal sinus rhythm. Computed tomography showed an extensively calcified RA mass of 4 cm in diameter which was located from the IVC across the RA and partly into the right ventricle (Figs. 3 and 4).

He subsequently underwent removal of the tumor via median sternotomy. Cardiopulmonary bypass was achieved via aorta and right atrium cannulation. The aorta was cross-clamped with cardioplegic arrest. While the core temperature decreased to 18°C circulatory arrest was initiated. The right atrium was incised longitudinally toward the IVC. This revealed a whitish-yellow, stone-hard calcified mass that attached to anterior wall of the IVC and atrial septum through a wide base. The tumor extended within the tricuspid valve annulus with direct attachment to the...
anterior and posterior leaflets (Fig. 5). The tumor was excised en bloc together with some firmly attached endocardium. It weighed 28g and measured about 41 × 21 × 10mm. The tricuspid valve was repaired using valvuloplasty and De Vega’s annuloplasty. The patient was weaned from cardiopulmonary bypass straightforward and the postoperative recovery was unremarkable. Pathological evaluation of the mass showed extensive calcification, with only a few typical myxoma spindle cells observed (Fig. 6). The final diagnosis was atrial myxoma with extensive calcification and tricuspid regurgitation. Follow-up TTE at 1 month revealed moderate tricuspid regurgitation without myxoma recurrence.

3. Discussion

Although cardiac myxomas are the most frequent primary cardiac tumor in adults, the incidence in pediatric patients is extremely low.[3] Such pediatric patients with cardiac myxomas are usually less symptomatic and discovered on TTE or computed tomography.[4]

In this patient, a precordial systolic murmur was found upon a routine medical examination before immunization and led him to diagnosis and treatment. This case of myxoma was characterized by severe calcification, which is more common in the right atrium rather than in the left atrium. whatsoever, heavy calcification of an atrial myxoma is uncommon especially in children.[5]

Definitive therapy for myxomas requires prompt surgical excision and long-term follow-up is recommended in children although recurrence after excision is rare.[3]

Unlike other pediatric intracardiac tumors, such as rhabdomyomas, definitive therapy for myxomas requires prompt surgical excision. A calcified right atrial myxoma is usually associated with disruption to the tricuspid valve, and after the mass has been removed, a repair attempt of tricuspid valve should always be made.
In conclusion, we report a rare case of a heavy calcification of right atrial myxoma in children, long-term follow-up is necessary for recurrence.

**Author contributions**

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