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CASE REPORT

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Huge right ventricular myxoma in a 15-year-old female patient: a case report and literature review

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Abstract

Background Myxomas represent about 5–10% of primary cardiac tumors in children, and only about 2.5–4% of cardiac myxomas originate from ventricular chambers. Symptoms and signs of right ventricular tumors depend mainly on the size and the site of the tumor and its effect on the inflow and outflow of the blood.

Case presentation A 15-year-old female patient presented with syncope on exertion and symptoms and signs of congestive heart failure. She had a history of dyspnea and palpitation on exertion for 2 years before admission. Echocardiography and cardiac magnetic resonance imaging (MRI) diagnose a huge right ventricular mobile mass (9 × 4.6 × 3.7 cm) prolapsing during systole into the main pulmonary artery and severe tricuspid regurgitation. The mass showed no contrast enhancement in early dynamic and delayed gadolinium images, suggestive of a thrombus. The mass was safely excised and tricuspid annuloplasty repair was done using right atriotomy and vertical right ventriculotomy approaches. The patient was discharged in a well condition on postoperative day 6. Pathologically, the mass was confirmed as cardiac myxoma with old extensive hemorrhage inside.

Conclusions Early echocardiography is essential for patients complaining of unexplained cardiac symptoms for early diagnosis and management of rare cases. Multimodality imaging is needed for the diagnosis and planning of the surgical procedure of right ventricular masses. Hemorrhage inside myxoma may lead to no contrast enhancement of the tumor.

Keywords Right ventricular tumors, Right ventricular mass, Right ventricular myxoma, Cardiac tumors in children, Intraventricular mass

Background

Primary cardiac tumors are rare in children with an incidence of 0.0017 to 0.28% in autopsy series and about 90% of them are benign [1]. The most common primary cardiac tumors in children are rhabdomyomas (42–75%) followed by fibromas (6–25%). Myxomas represent only 5 to 10% of primary cardiac tumors in children, while more than 50% of primary cardiac tumors in adults are myxomas [2, 3]. Most of the cardiac myxomas are located in the left atrium (75–85%) followed by the right atrium (15–20%). Only about 2.5–4% of cardiac myxomas originate from ventricular chambers [4–7].

We present a case of a 15-year-old female with a huge right ventricular myxoma protruding into the pulmonary artery during systole, as myxoma is rare in this location and this size. Also, cardiac myxoma is rare in this age group.

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Case presentation

Fifteen-year-old Egyptian female student admitted to the hospital because of syncope on exertion. She had a history of dyspnea and palpitation on exertion for 2 years before admission. Dyspnea exaggerated 2 weeks before admission with the appearance of bilateral lower limb edema. The patient had no personal or familial history of cardiac diseases, and she had no history of a previous admission to a hospital or chronic diseases.

On examination, her blood pressure was normal (110/70 mmHg) with a regular heart rate (98/min). Neck veins were congested with bilateral mild lower limb edema. A systolic murmur on the lower left border of the sternum was heard. Complete blood count, renal function tests, serum alanine aminotransferase (ALT), serum aspartate aminotransferase (AST), and serum alkaline phosphatase were within normal limits. There were slight increases in serum total bilirubin (1.93 mg/dl), serum direct bilirubin (1.50 mg/dl), prothrombin time (16.1 s), and international normalized ratio (INR) (1.41). Electrocardiogram revealed a normal sinus rhythm with a right bundle branch block. A plain chest X-ray showed mild right pleural effusion.

An echocardiography was done for the patient and revealed the presence of a big mass in the right ventricle (RV) and its outflow tract (54 mm × 43mm) (Fig. 1A), movable and protruded into the pulmonary artery; dilated right ventricle (right ventricular dimension = 48 mm); dilated right atrium (right atrial dimension = 45 mm); severe tricuspid regurgitation (Fig. 1B); increased right ventricular systolic pressure (RVSP = 75 mmHg); and dilated inferior vena cava. Differential diagnosis of right ventricular mass was considered on echocardiography and the patient was subjected to cardiac magnetic resonance imaging (MRI).

Cardiac MRI revealed a large right ventricular outflow tract (RVOT) soft tissue mass, about 9 × 4.6 × 3.7 cm, mobile, prolapsing during systole into the main pulmonary artery (Fig. 2A, B), benign looking, attached to the interventricular septum (IVS), and exhibited an isotense to high signal in T2 W1. The mass showed no contrast enhancement in early dynamic and delayed gadolinium images, suggestive of thrombus formation (Fig. 2C, D). The RV was dilated with increased indexed volumes and impaired RV systolic function (EF ± 29%) with global hypokinesia of RV walls. The left ventricular (LV) function was mildly impaired (EF ± 40%) with a D-shaped interventricular septum and paradoxical motion denoting increased RV pressure. Also, MRI revealed no definite abnormal macroscopic mural enhancement in RV and LV walls, dilated right atrium (about 30 cm^2) with a normal size of the left atrium, severe tricuspid regurgitation, dilated inferior vena cava, dilated hepatic veins, hepatomegaly, minimal pericardial effusion, and minimal pleural effusion that it was more on the right side.

According to the absence of contrast enhancement in the mass, in early dynamic and delayed gadolinium images, the possibility of thrombus formation was raised, but because of the presence of manifestations of congestive heart failure and the presence of severe tricuspid regurgitation that need tricuspid repair and to eliminate the risks of distal embolization, we decided surgical intervention to excise the mass and repair the tricuspid valve.

The patient was prepared for complete resection of the right ventricular mass and tricuspid valve (TV) repair under cardiopulmonary bypass (CPB). Standard

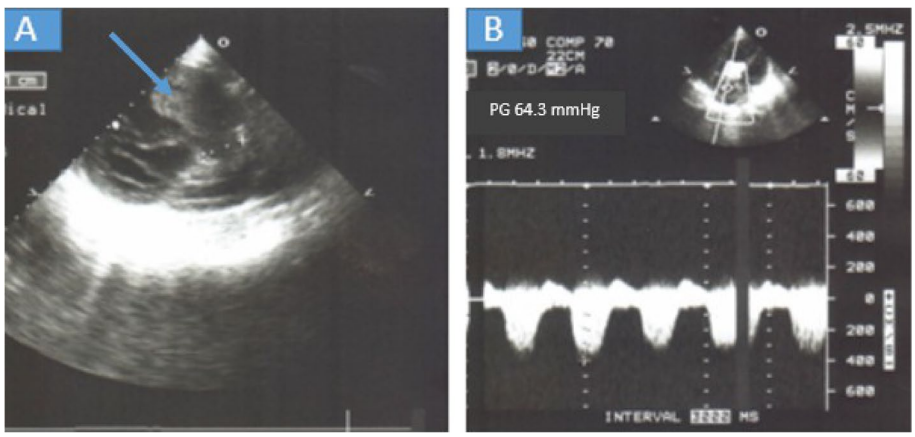

Fig. 1 **A** Two-dimensional transthoracic echocardiogram (TTE), parasternal long-axis view showing a large mass in the right ventricle. **B** Two-dimensional TTE, apical view, with continuous wave Doppler showing severe tricuspid regurgitation with a pressure gradient of 64.3 mmHg across the tricuspid valve

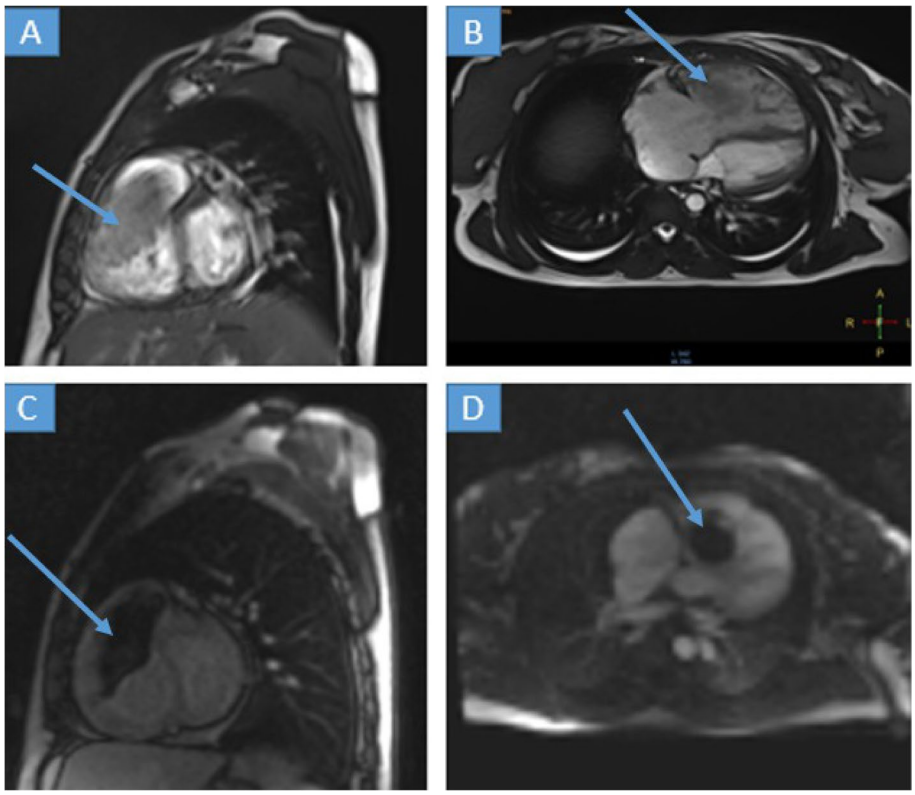

Fig. 2 Cardiac magnetic resonance images: **A** sagittal view, showing a large mass in the right ventricle and the right ventricular outflow tract; **B** cross-sectional view, showing a right ventricular mass; **C**, **D** delayed post-contrast-enhanced imaging showing no contrast enhancement of the mass

median sternotomy and CPB with aortic and bicaval cannulation and warm blood cardioplegia were used. Through a longitudinal right atriotomy, the right atrium was explored. The mass was seen protruded from the tricuspid valve. The tricuspid annulus was dilated with normal tricuspid leaflets. The large size of the ventricular mass did not allow us to safely excise it through the right atriotomy approach. A vertical incision of the RVOT was performed and the upper part of the mass was delivered through the incision (Fig. 3A). Then, the mass was completely excised from its attachment to the septum and it was delivered outside. Saline washing of the RV cavity and exploring the integrity of the TV apparatus, septum, and ventricular wall were done followed by the closure of the ventricular incision by two layers with proline sutures, a continuous suturing layer and a pledgeted transverse mattress layer (Fig. 3B). Tricuspid annuloplasty repair was done with a pericardial strip before the closure of the right atriotomy. The patient was weaned from cardiopulmonary bypass (CPB) easily and transferred to our intensive care unit. The mass had a gelatinous consistency with hemorrhagic areas on gross examination (Fig. 4A).

The postoperative course was uneventful, and the patient was discharged on postoperative day 6 in a well condition to be followed in our outpatient clinic.

Microscopic examination of the mass revealed neoplastic growth displayed as proliferating polygonal/stellate myxoma (lepidic) cells embedded upon loose myxoid matrix with wide old hemorrhage, indicating cardiac myxoma with extensive old hemorrhages (Fig. 4B–D).

The presence of extensive old hemorrhages in the myxoma mass may be the cause of the absence of contrast enhancement in early dynamic and delayed gadolinium images in cardiac MRI.

A follow-up echocardiography was done 2 months after the operation and revealed successful resection of the RV mass without residual, and the TV displays normal thickness and morphology with mild functional central regurgitation.

Discussion

Cardiac tumors may be asymptomatic and found incidentally. Symptoms are usually related to the cardiac location and the tumor size. Cardiac tumors may be presented with constitutional symptoms as fever, weight loss, and

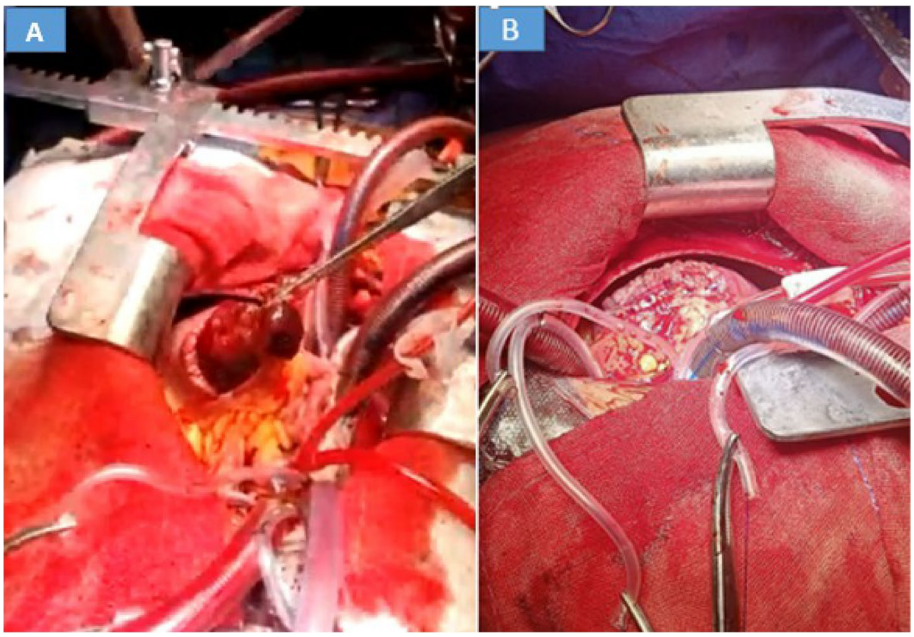

Fig. 3 **A** The image showing the upper part of the mass delivered through the right ventriculotomy. **B** The image showing the right ventriculotomy closed with 2 layers of proline sutures, a continuous suturing layer and a pledgeted transverse mattress layer

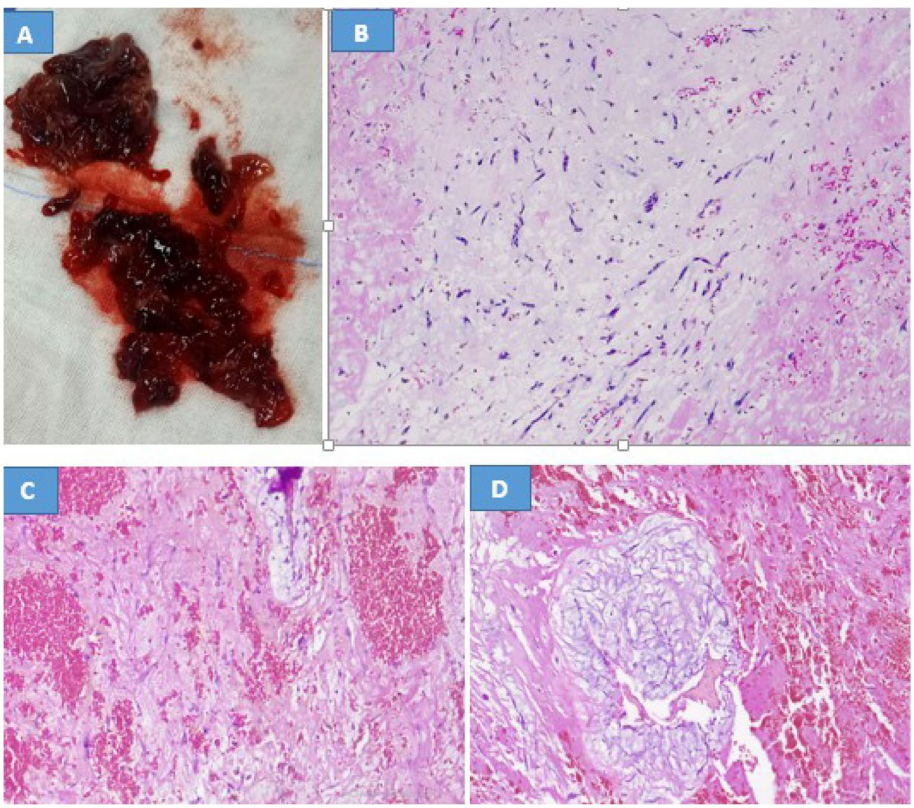

Fig. 4 **A** The image showing the excised tumor. The tumor had a gelatinous consistency with hemorrhagic areas. The hematoxylin and eosin histology sections (**B**–**D**) showing polygonal/stellate myxoma cells embedded upon loose myxoid matrix with wide hemorrhage

fatigue or mass effect that interfere with blood flow, heart valves, and myocardial function resulting in arrhythmias, dyspnea, chest discomfort, or syncope. Also, it may be presented by embolic manifestations. Usually, intracavitary tumors are more symptomatic than intramural tumors [1, 3, 6, 8].

Factors that are helpful in establishing a diagnosis of intracardiac mass are the location of the mass, age at presentation, and imaging characteristics [8].

RVOT obstruction produced by right ventricular tumors will lead to elevated right-sided pressure and low cardiac output, and patients will present with dyspnea, peripheral edema, hepatomegaly, and ascites. Sometimes, they present with symptoms and signs of pulmonary embolism as the first presentation [3, 9–13].

Our patient admitted to the hospital because of syncope, but she had a history of dyspnea and palpitation on exertion for 2 years before admission. So, early echocardiography is essential for patients with unexplained cardiac symptoms for early diagnosis and management. The existence of a large tumor in the RV cavity and significant elevation of right ventricular pressure due to partial obstruction of the RVOT induces dilatation of the cavity and subsequently dilatation of the tricuspid annulus that leads to tricuspid regurgitation [11], and we think that this was the cause of severe tricuspid regurgitation in our patient.

Echocardiography is the first noninvasive step for the evaluation of cardiac masses. If echocardiography cannot clearly distinguish the mass, computed tomography (CT) or cardiac magnetic resonance imaging (MRI) can be used [10]. Cardiac MRI is superior to CT in detecting anatomic details and avoids radiation [14, 15]. Cardiac MRI with gadolinium enhancement has been found to be integral in the accurate diagnosis of cardiac masses, with an accuracy of more than 80% [16].

Echocardiography for our patient gave us a valuable information regarding the mass and its effect on the chambers and valves of the heart but we cannot differentiate whether is it a tumor or a thrombus. So, a cardiac MRI was requested.

Contrast enhancement in early dynamic and delayed gadolinium images in cardiac MRI is very useful in differentiating myxomas from a non-enhancing thrombus [15, 16]. In our patient, the mass showed no contrast enhancement suggesting thrombus formation. But, considering the huge size of the mass ($9 \times 4.6 \times 3.7$ cm) with the absence of symptoms and signs of pulmonary embolism, the age of the patient (15 years), and the absence of diseases that may cause a hypercoagulability state, the thrombus was less likely and the mass was likely a primary cardiac tumor. Also, the presence of manifestations of congestive heart failure and severe tricuspid regurgitation that need tricuspid repair encouraged us to decide on surgical intervention to excise the mass and repair the tricuspid valve.

On gross examination, the excised mass had a gelatinous consistency with hemorrhagic areas. Pathological examination revealed that the excised mass was a cardiac myxoma with extensive old hemorrhage inside it. We believe that the presence of extensive old hemorrhage in the myxoma was the cause of the absence of contrast enhancement in the MRI images. Katiyar et al. [17] reported a case of myxoma in RVOT with no enhancement on post-contrast scan subjected to contrast-enhanced computed tomography (CT) and on delayed post-gadolinium sequences in cardiac MRI, and enhancement occurred only at the site of attachment and at the thin rim of the peripheral fibrous cap. They attributed this to the central thrombotic component of the myxoma.

The approaches for resection of right ventricular masses are right atriotomy, right ventriculoctomy, and pulmonary arteriotomy [9, 12, 18]. As there was severe tricuspid regurgitation, right atriotomy was mandatory for tricuspid repair, and as we found that excision and extraction of the huge mass through the tricuspid valve could be hazardous, right ventriculotomy was done and the mass was excised and extracted through it without any complications.

Conclusions

The location of cardiac myxomas in the right ventricle in children is rare. The tumor mostly presented with symptoms and signs of congestive heart failure mainly due to obstruction of RVOT. Early echocardiography is essential for patients with unexplained cardiac symptoms for early diagnosis and management of rare cases. Multimodality imaging is needed for the diagnosis and planning of the surgical procedure. Hemorrhage inside myxoma may lead to no contrast enhancement of the tumor.

Abbreviations

MRI	Magnetic resonance imaging
ALT	Serum alanine aminotransferase
AST	Aspartate aminotransferase
INR	International normalized ratio
RV	Right ventricle
RVSP	Right ventricular systolic pressure
RVOT	Right ventricular outflow tract
IVS	Interventricular septum
EF	Ejection fraction
LV	Left ventricle
CPB	Cardiopulmonary bypass
TV	Tricuspid valve
CT	Computed tomography
TTE	Transthoracic echocardiogram

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Authors' contributions
AD decided and did the surgical intervention; designed the case report, analysis, and interpretation of the data; and drafted the manuscript. HE was involved in the analysis of data and in drafting the manuscript. AB shared in the decision and in doing the surgical intervention and was involved in the analysis of data and in drafting the manuscript. The authors read and approved the final manuscript.

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