Cardiology in the Young

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Right ventricular myxoma in a child causing right ventricular outflow tract obstruction

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Brief Report

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Abstract

Primary cardiac tumours are uncommon in the paediatric population, accounting for fewer than 0.5% of paediatric cases of cardiac disease. Right ventricular tumours, including myxomas, are particularly rare and may be asymptomatic or demonstrate varying degrees of cardiac dysfunction based on the location and size of the tumour, inducing conduction abnormalities, syncope, embolism, and potentially, sudden death. We report a rare case of right ventricular myxoma causing severe right ventricular outflow tract obstruction and surgical intervention in a paediatric patient.

Case report

A previously healthy 11-year-old girl experienced nausea, lightheadedness, and loss of consciousness while playing outdoors with her sister. She was taken to the local emergency room where she also described a history of headaches, occasional palpitations on exertion, and episodes of dizziness and pre-syncope over the previous 2 weeks. An electrocardiogram demonstrated a prolonged QTc interval of 519 msec, and an echocardiogram revealed a 4 × 4 cm obstructive lesion in the right ventricle with a peak pressure gradient of 78 mmHg across the right ventricular outflow tract. She was then transferred to a tertiary health centre for specialist evaluation.

Physical examination revealed a III/VI harsh, holosystolic murmur loudest at the left sternal border. Chest radiograph showed a normal cardiac silhouette. Because of the history of syncope and severe obstruction in the right ventricular outflow tract, surgery was recommended. Using cardiopulmonary bypass and cardioplegic arrest, the main pulmonary artery was opened, and the mass was identified just below the pulmonary valve with a visible attachment point along the anterior free wall of the right ventricle (Fig 1). Due to the large size of the myxoma and apparent extension towards the apex of the right ventricle, additional exposure was required through the right atrium. A short pedunculated attachment was localised with gentle traction on the tumour, and the mass and a small amount of adjoining endocardium were resected via the right atrium, as the mass could not be extracted through the pulmonary valve annulus due to its size (Fig 2). The inferior portion of the mass was somewhat adhered to the right ventricular trabeculations, giving the appearance of multiple attachment points. Several small areas of muscle were resected with the tumour along with tissue near the apex that appeared to be organised thrombus which was adherent to the tumour. At completion of the resection, no additional tumour or thrombus was visible within the ventricle. The incisions were closed, the heart reperfused, and the patient was weaned from cardiopulmonary bypass without difficulty. Intra-operative transesophageal echocardiogram demonstrated relief of the right ventricular outflow tract obstruction with low normal right-sided systolic function and no residual mass. The patient was extubated in the operating room and taken to the paediatric cardiac ICU in stable condition.

Because of the appearance of multiple attachment points and the rarity of myxomas in the right ventricular outflow tract, a frozen section was obtained during the procedure. This demonstrated a multilobulated, low-grade myxoid tumour without evidence of myocardial infiltration. The pathologic evaluation of permanent sections confirmed the diagnosis of benign myxoid neoplasm consistent with myxoma.

After an uncomplicated postoperative course, the patient was discharged on postoperative day 6 on furosemide, as well as prophylactic clopidogrel because of the complexity of the tumour resection within the trabeculated right ventricle and the suspicion for thrombus around the tumour site. She underwent additional evaluation to rule out Carney complex, which is associated with paediatric cardiac myxomas, but no other findings were identified. She was referred for additional genetic work-up. Follow-up echocardiogram evaluation at 22 months post-resection demonstrated no visible tumour recurrence or right ventricular outflow obstruction and normal right ventricular function. The patient remains asymptomatic and in normal sinus rhythm.

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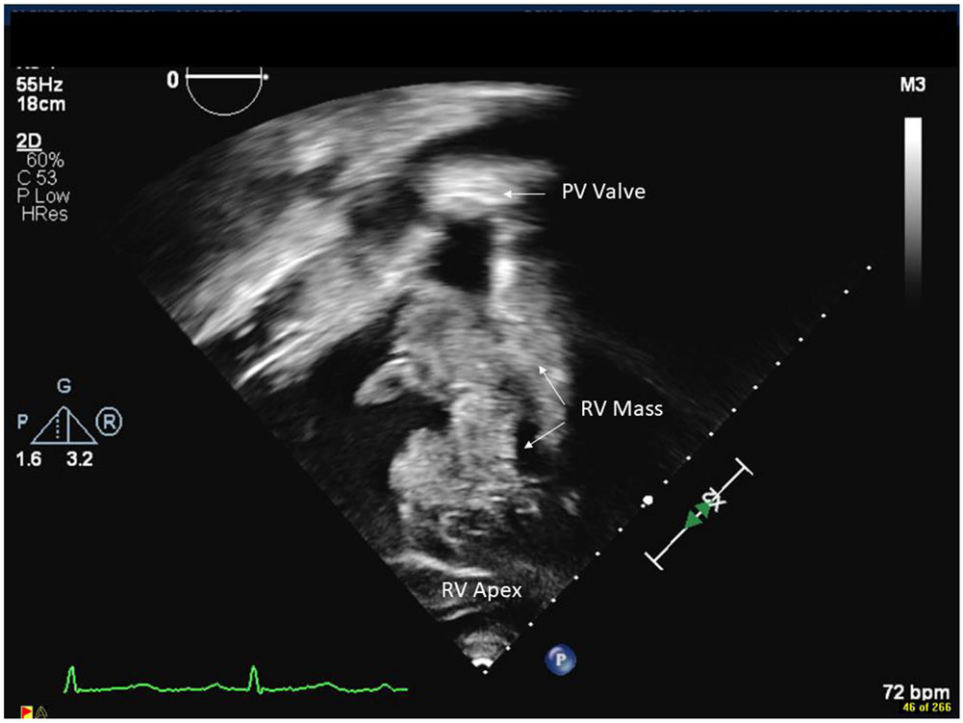


Figure 1. Right ventricular mass.

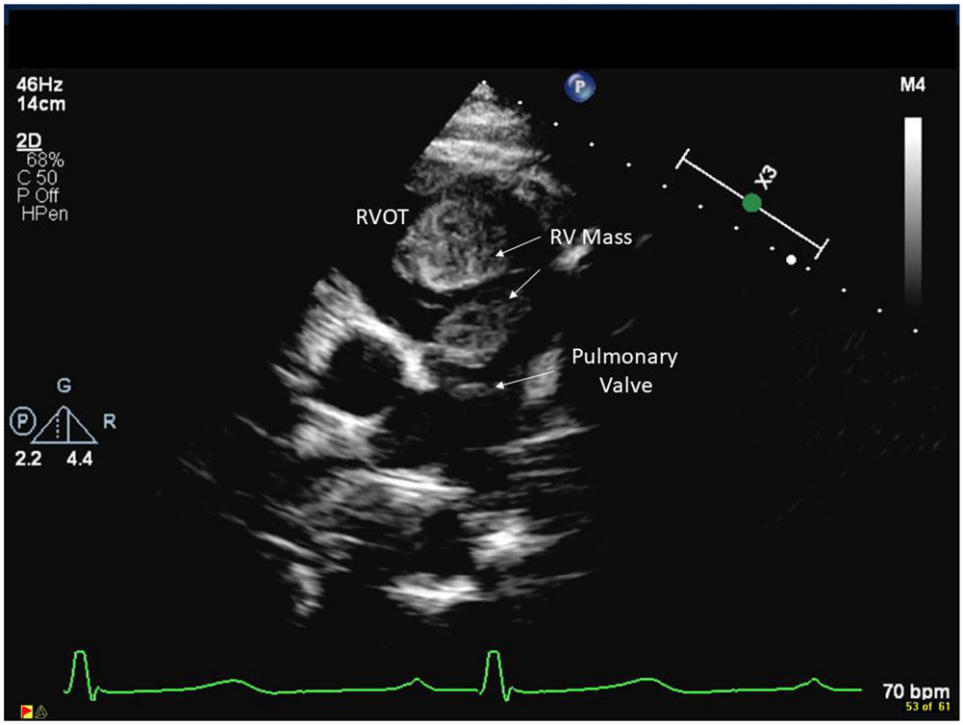


Figure 2. Right ventricular mass.

Discussion

An estimated 15% of paediatric cardiac tumours are myxomas, primarily occurring in the left atrium.[1] Sporadic right ventricular myxomas are extremely rare with only isolated case reports describing their incidence and clinical outcomes.[2] Myxomas are the commonest intracardiac tumours in adults, yet they are one of the rarest cardiac tumours found in children, occurring in 0.17% of total cardiac lesions.[3,4] The histology, prevalence, and

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location of paediatric cardiac tumours differ considerably from their counterparts in adults. Rhabdomyomas are the commonest paediatric cardiac tumours, followed by teratomas, fibromas, myxomas, and rhabdomyosarcomas.[5–7] Except for the latter, these tumours are histologically benign, causing symptoms through either obstruction or dysrhythmias depending on tumour size and location. As with left-sided masses, two-dimensional echocardiography can be utilised to evaluate the tumour size and the presence of inflow or outflow obstruction. Severity of right ventricular outflow tract obstruction can help predict future risk of syncope, pulmonary embolism, and sudden death and help dictate urgency of removal.[8]

Isolated right ventricular myxomas in childhood are extraordinarily rare, though their incidence seems to be increasing due to improved detection.[3] Clinical presentation and insights into surgical prognosis are based predominantly on few case reports rather than cohort studies. Most documented cases of right ventricular myxomas in children have been linked to familial myxoma syndrome, or Carney complex, a rare autosomal dominant collection of mucocutaneous, visceral, and endocrine disorders and tumours.[8] Other than the myxoma location, no other signs of Carney complex were identified in this patient, and her family history was unremarkable for similar findings consistent with an underlying genetic condition. For asymptomatic paediatric cardiac tumours without significant obstruction to flow, surgical removal is not recommended as first-line treatment. Rather, careful follow-up to observe tumour size and patient symptoms is recommended, as many tumours are well-documented to regress spontaneously.[7] If significant clinical manifestations are present, complete surgical excision of the myxoma is recommended to prevent further cardiac deterioration or complications since surgical outcomes are favourable. [3,7–9] Close post-resection follow-up imagining is indicated to monitor for tumour recurrence.

Paediatric cardiac myxomas are uncommon, and they are particularly rare in the right ventricle. Their size and location-dependent symptoms may pose diagnostic and surgical challenges. Despite their rarity, the diagnosis of a cardiac tumour should remain within the differential for evaluation of syncope and dysrhythmia in a child. Surgical removal in the symptomatic patient is indicated to prevent complications of obstruction of flow, dysrhythmia, embolisation, syncope, and sudden death.

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Conflicts of interest. None.

Ethical standard. Not applicable.

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