

Perioperative Management of Critical Right Ventricular Inflow Obstruction from Right Atrial Rhabdomyoma

Abstract

Rhabdomyoma is the most common cardiac tumor in infancy and commonly located in the ventricles causing outflow obstruction or arrhythmias. We report a rare pediatric (7 month old) case of a right atrial rhabdomyoma presenting with severe cyanosis and low cardiac output from significant tricuspid inflow obstruction with right to left shunt across a stretched patent foramen ovale. We present an emergency cardiac surgery for right atrial tumor resection, and the management of separating the patient with failing right ventricle from cardiopulmonary bypass using a Glenn shunt, since extracorporeal membrane oxygenation (ECMO) or nitric oxide was not available.

Keywords: Cardiothoracic surgery, extracorporeal membrane oxygenation, pediatric oncology, right atrium tumor

Introduction

Rhabdomyoma is considered the most common cardiac tumor in childhood.^[1-8] In general, rhabdomyomas are multiple, well circumscribed, noncapsulated nodules, found most commonly in the ventricles. They may cause obstruction of blood flow and or arrhythmias.^[1,3,5-7] Outflow tract obstruction is the most common complication; inflow obstruction has rarely been reported.^[3]

Case Report

A 7-month-old female infant was referred due to severe cyanosis. She had been normal at birth but developed feeding difficulties 6 weeks before presentation and cyanosis 2 weeks later. At the time of admission, she weighed 6.1 kg, was critically ill with low cardiac output (CO), weak peripheral pulses (heart rate 144 bpm), blood pressure (82/40 mmHg), and central cyanosis (oxygen saturation 63%).

Echocardiography revealed a massive RA tumor measuring 33 cm × 31 mm causing almost complete inflow obstruction of the tricuspid valve. The right ventricle (RV) appeared underfilled. A stretched patent foramen ovale (PFO) was allowing a right to left shunt across, resulting in venous blood entering the systemic circulation with consecutive drop in oxygen saturation. No

other tumors were seen in any of the other cardiac chambers [Figures 1 and 2].

While the chest radiograph appeared normal, the electrocardiogram showed RA enlargement. During emergency cardiac surgery, a solitary tumor was removed from the RA. It was found to be encapsulated, measured 30 mm × 30 mm, had a broad-based stalk and arose from the RA appendage. The RA was markedly dilated, with the tumor occupying its entire cavity, while the RV was much smaller. The tumor was completely excised along with part of the appendage and the PFO closed. An initial attempt to separate from cardiopulmonary bypass (CPB) failed despite high inotropic support with dobutamine (10–15 mcg/kg/min) and epinephrine (0.1 mcg/kg/min). The small RV in combination with moderate tricuspid regurgitation led to a low CO state. Subsequently, a bidirectional Glenn shunt operation was performed, and the child weaned off from CPB with dobutamine 5 mcg/kg/min and epinephrine 0.05 mcg/kg/min after a total CPB time of 66 min. The immediate postoperative echocardiogram showed a large RA and small RV with moderate tricuspid incompetence and systolic dysfunction. Histologic examination of the excised tumor revealed a rhabdomyoma. The child remained hemodynamically unstable for 3 postoperative days requiring high doses of

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dobutamine (up to 10 mcg/kg/min). She gradually improved and was successfully extubated on the 5th postoperative day. RV function was normal by the 6th day [Figure 3]. Inotropes were tapered off, and the patient was fit for discharge by the 8th postoperative day with normal central venous pressure and an oxygen saturation of 94%. She was subsequently discharged home on aspirin.

Discussion

Cardiac tumors in children are rare (0.03%–0.32% of patients) and present early in life.^[7] A multi-institutional database of children undergoing cardiac surgery for tumors showed 42 out of 120 children had rhabdomyoma,^[9] most often noted in ventricles. In a study by Nield *et al.*,^[11] 28% of the patients had tumors in the right ventricular outflow tract ($n = 13$) or left ventricular outflow tract ($n = 23$). In a single-center study of 6 patients, the tumors mostly originated from the left ventricle.^[2] A more detailed analysis by Beroukhim *et al.*^[4] described 78 patients with 85 tumors of which only 10 were located in the RA (11%).

The most common indication for surgery has been flow obstruction,^[3,6] of which outflow obstruction is significantly more common than inflow obstruction. Surgery has been the definitive treatment, occasionally on an urgent basis. Pucci *et al.* involved 24 postnatal cases, of which 7 patients underwent surgery with 3 mortalities.^[5]

Patients usually present with arrhythmia.^[7] Tumor obstruction is rarely the cause for emergent surgery. One of the largest series of cardiac tumors presented with arrhythmia including ventricular tachycardia requiring surgical management; others have reported combined surgery and radiofrequency ablation.^[8]

Cavopulmonary connection has been previously performed in a case with rhabdomyoma, but without details described.^[7] In our case, the inflow obstruction seen in preoperative echocardiography had not allowed the RV to facilitate adequate CO.

In our case, the tumor was completely excised along with part of the RA appendage and the PFO closed. After tumor excision, an initial attempt to separate from CPB failed since the small RV in combination with moderate tricuspid regurgitation led to a low CO state. Subsequently, a bidirectional Glenn shunt operation was performed and the child was weaned off from CPB. At the time when the decision for the Glenn shunt was made, other options such as the use of venoarterial (VA) extracorporeal membrane oxygenation (ECMO) for circulatory support or nitric oxide ventilation for decrease of pulmonary pressure to help offloading the RV were not available and the Glenn shunt was considered the best available option. In recent years, the use of ECMO as one form of extracorporeal life support has been increased throughout the world and presently may represent the state of the art. In this context, Maybauer *et al.*^[10] reported cases of patients with Ebstein's anomaly

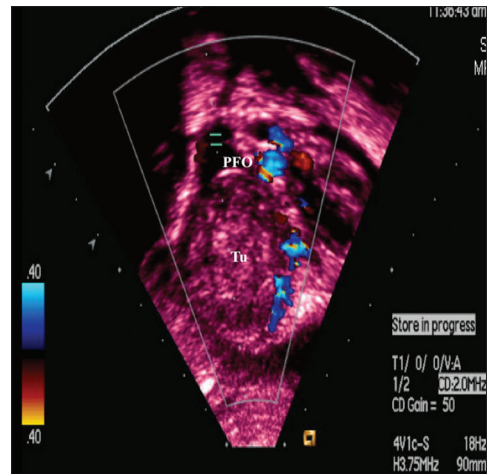


Figure 1: Subcostal view showing the huge right atrial mass (Tu), small patent foramen ovale, and very little blood flow across the tricuspid valve

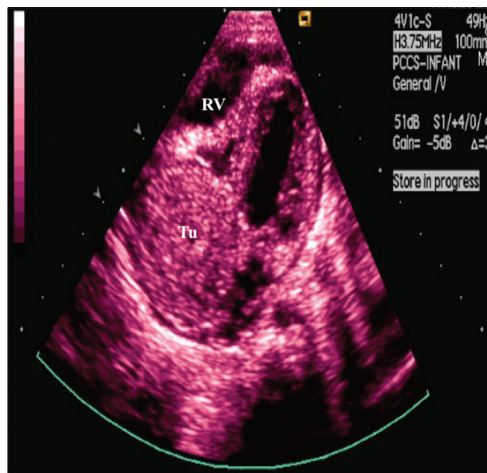


Figure 2: Apical 4-chamber view showing entire right atrium filled with a tumor (Tu) and a small right ventricle with a tumor pushing interatrial septum to the left

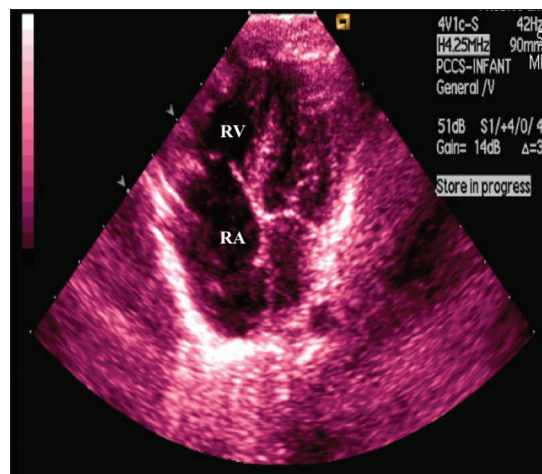


Figure 3: Apical 4-chamber view 6 days after surgery shows right atrium and good size right ventricle

who presented in cardiogenic shock, one postcardiotomy and one with severe biventricular failure caused by an acute

thrombotic event with anatomical narrowing and inflow obstruction similar to the present case. VA-ECMO was initiated and was considered as bridge to either recovery, or ventricular assist device, or to the heart transplantation. However, the patients were stabilized and recovered on VA-ECMO within days because time was crucial to adapt to the increased RV blood volume. A similar approach could have been possible in our patient if ECMO was available at the time and may have been more beneficial than having a Glenn shunt with its own long-term issues.

Conclusion

Right ventricular inflow obstruction is rare presentation of rhabdomyomas. In cases of RV failure, nitric oxide ventilation reduces RV afterload and is helpful to improve RV performance. In severe cases, a cavopulmonary shunt (Glenn shunt) as surgical treatment and ECMO for circulatory support are very useful tools to separate from CPB.

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Conflicts of interest

There are no conflicts of interest.

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