



Surgical Resection of Cardiac Rhabdomyoma in a Neonate

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Abstract

Rhabdomyomas are the most common cardiac tumors in infancy and childhood. Surgery is reserved for those patients with hemodynamic compromise. We present a case of surgical resection of a large mass obstructing the left ventricular outflow tract in a neonate.

Keywords: Rhabdomyoma; Cardiac tumor; Left ventricular outflow tract; Neonate

Case Presentation

A 38-weeks gestational age newborn girl was derived to our institution with the diagnosis of a cardiac tumor. On her first day of life, a cardiac murmur and signs of low cardiac output were detected. The echocardiogram performed after birth revealed the presence of a large mass in the left ventricular outflow tract. Clinical presentation resembled that of critical aortic stenosis. Therefore, prostaglandin infusion was started to restore adequate systemic blood flow and perfusion of vital organs. The patient was derived to our institution and arrived in good condition. The diagnosis was confirmed. The large tumor (19 x 17 mm) infiltrated the ventricular septum and protruded into the left ventricular outflow tract, occluding 90% of its diameter (Figure 1). No other cardiac masses were found.

All other organs were scanned for associated lesions using ultrasound and MRI. No other tumor was found.

After diagnostic confirmation, the patient was taken to the operating room. Median sternotomy was performed. Cardiopulmonary bypass was started using aortic and single venous cannulation. The aorta was crossclamped. The ascending aorta was transected some millimeters above the sinotubular junction. Through the aortic valve, the left ventricular outflow tract was explored. A large homogeneous white mass was found just below the leaflets of the aortic valve. It completely occluded the subaortic region, making visualization of the ventricular cavity impossible (Figure 2).

After identifying its free margins, the cardiac mass was detached from the ventricular septum using an ophthalmic micro scalpel. The protruding portion was completely resected (Figure 3), but part of the tumor remained infiltrating the ventricular septum. Extended resection in that area was considered impossible without causing severe damage to vital anatomic structures. The aorta was then sutured. Air was removed and the aortic cross clamp was released. The heart showed adequate contractility with sinus rhythm. Cardiopulmonary bypass was ended and the chest was closed.

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Figure 1: Long paraesternal view of the left ventricular outflow tract. The tumor infiltrates the ventricular septum and protrudes into the subaortic region, leaving only a narrow pathway. The red line delimitates the margins of the cardiac mass (Ao: Aorta; LV: Left Ventricle; T: Tumor).

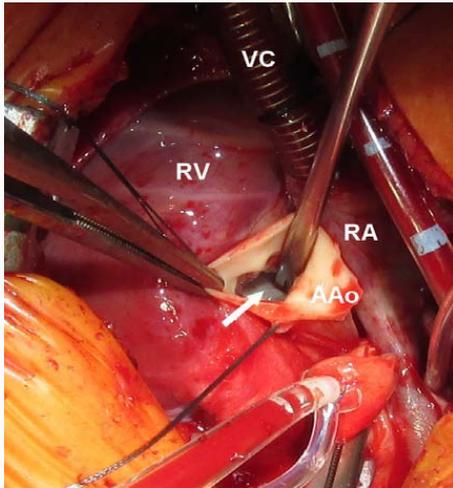


Figure 2: The aorta has been transected above the sinotubular junction. Through the aortic valve, a white mass can be identified occupying the left ventricular outflow tract in almost all its diameter (AAo: Ascending Aorta; RA: Right Atrium; RV: Right Ventricle; VC: Venous Cannula; Arrow; Tumor).

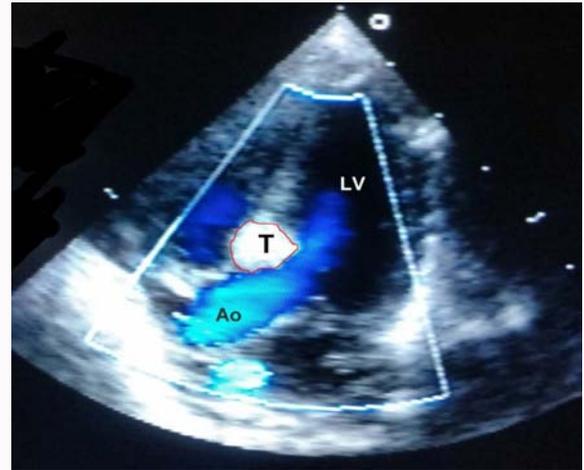


Figure 4: Five chamber apical view of the subaortic area showing unobstructed pathway from the left ventricle to the ascending aorta. The size of the tumor has been considerably reduced (Ao: Aorta; LV: Left Ventricle; T: Tumor).

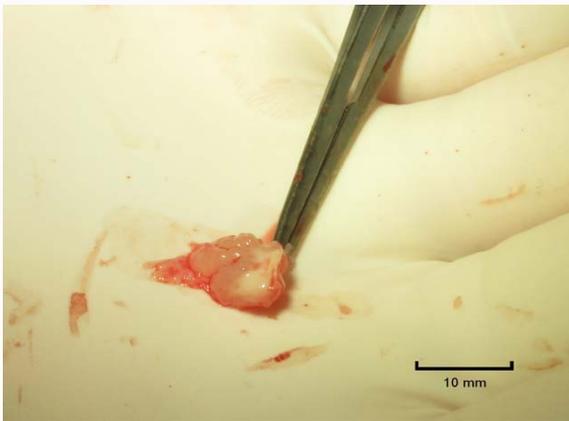


Figure 3: The tumor has been partially excised from the ventricular septum.

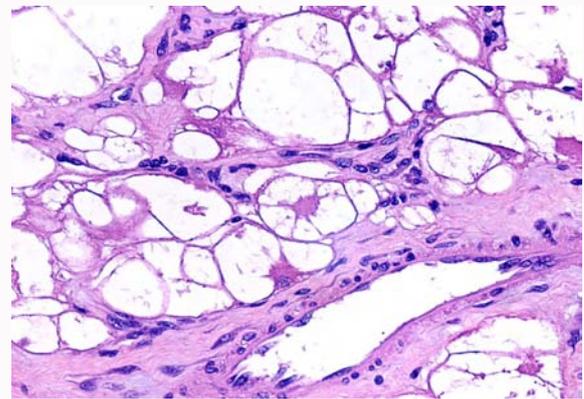


Figure 5: Haematoxylin and eosin staining of the cardiac rhabdomyoma.

The patient was extubated on the following day, and discharged 5 days later in an excellent condition. Postoperative echocardiogram showed unobstructed left ventricular outflow tract without aortic regurgitation or stenosis (Figure 4).

Anatomic pathology confirmed the diagnosis of rhabdomyoma (Figure 5). Although there were no obvious signs of tuberous sclerosis, the patient was derived to the geneticist after hospital discharge.

Discussion

Rhabdomyoma is the most prevalent of all cardiac tumors in infancy and childhood [1]. It accounts for 35% of the cases. More than 70% of the patients present with symptoms. It is a benign tumor with a clear tendency to spontaneous regression and unreported malignant degeneration. However, it can be life-threatening in those cases in which it causes severe mechanical obstruction within the heart. Left ventricular outflow tract obstruction is the most frequent cause of death, and it is usually associated to a single tumor located in the septum [2].

Surgical resection is reserved for these situations or to prevent potentially lethal acute complications such as embolization or ventricular arrhythmias [3]. Reported mortality rate is less than 5%.

Previous studies have demonstrated no difference between total and partial resection of cardiac rhabdomyomas in terms of postoperative complications and mortality [4]. Besides, regrowth of the tumor after partial excision is not common. Therefore, resecting only those parts that produce hemodynamic alterations may be the safest strategy [3]. Masses with subaortic location are especially challenging, because surgical approach must be done through the aortic valve, which has a small diameter in the neonatal period [5].

This tumor is strongly related to tuberous sclerosis. Depending on the series, 30% to 90% of patients with a cardiac rhabdomyoma suffer from this disease. This is a genetic disorder with variable compromise caused by benign tumors in many vital organs such as the heart, central nervous system, eyes, kidneys, lungs and skin. Prognosis is extremely variable and depends on the severity of the manifestations [6].

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