Giant Intramural Left Ventricular Rhabdomyoma in a Newborn

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A 1-day-old newborn was referred with prenatal echocardiographic diagnosis of cardiac mass. Born by caesarean section at 38 + 3 weeks of pregnancy, he presented with cyanosis and bradycardia at birth, requiring immediate intubation and mechanical ventilation. Chest x-ray demonstrated cardiomegaly with a cardiothoracic index of 0.7 (Figure 1). Echocardiographic examination (Figure 2 and Movie I in the online-only Data Supplement) confirmed the diagnosis of a large cardiac mass located at the posterior wall and at the apex of the left ventricle (LV).

The child presented with several episodes of paroxysmal supraventricular tachycardia, which were treated with DC shock and various pharmacological agents, such as amiodarone, adenosine, flecainide, digoxin, and metoprolol, and 1 episode of ventricular fibrillation with cardiac arrest requiring resuscitation. MRI showed a single large mass (5.5×5 cm) arising from the left atroioventricular groove and extending to the LV lateral wall and apex, up to the interventricular septum, with no LV inflow or outflow obstruction, and was suggestive for cardiac fibroma (Figure 3A through 3C and Movie II in the online-only Data Supplement). The left bronchus was partially obstructed by external compression from the mass. However, further MRI tissue characterization was compatible with a LV rhabdomyoma, because the gradient echo sequences showed signal intensity of the mass similar to that of the myocardium. In addition, turbo-spin echo sequences with fat saturation ruled out the presence of fatty tissue. Finally, the images obtained after gadolinium chelate injection (Figure 3D) did not show any enhancement of the tumor, and this finding was significant enough to rule out a fibroma.

Figure 1. Postnatal chest x-ray. The cardiac silhouette is dilated (cardiothoracic index, 0.7). Note the endotracheal tube and umbilical line.

Figure 2. Postnatal echocardiographic examination, showing a large cardiac mass, located at the posterior wall and apex of the left ventricle, and involving the interventricular septum. LV indicates left ventricle; RV, right ventricle.

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Further preoperative evaluation of the coronary arteries was not performed. Finally, neurological investigation excluded the association with tuberous sclerosis.

At 14 days of life, the patient was scheduled for surgical resection of the cardiac mass. After median sternotomy, on cardiopulmonary bypass, the large solid mass (Figure 4A) was partially resected (Figure 4B) to preserve the coronary branches that were running close to it, as evidenced intraoperatively. The postoperative course in the intensive care unit was unremarkable, on low-dose inotropes, without supraventricular tachycardia recurrence. The patient required mechanical ventilation for 4 days, and was discharged from the intensive care unit on the 8th postoperative day and eventually discharged from the hospital on the 17th postoperative day. Histopathologic examination revealed the presence of enlarged, vacuolated cells with sparse cytoplasm and so-called spider cells, ie, cells characterized by a typical centrally located nucleus with radial extensions to the cell periphery. Immunohistochemical studies showed features of striated muscle cells (myoglobin positive) confirming the diagnosis of cardiac rhabdomyoma (Figure 5). Ten months after the operation, the child is alive and in good clinical condition, with no oral medications. Follow-up ECG Holter monitoring documents sinus rhythm, and echocardiographic assessment demonstrates a good LV function (shortening fraction 37%) and only a small residual mass in the apical portion of the interventricular septum, not hemodynamically significant.

Primary cardiac tumors are rare entities, occurring with a lifetime incidence of 0.0017% to 0.02%.1 In the pediatric population, cardiac tumors have been identified in 0.02% to 0.04%,2,3 and the majority of them are benign rhabdomyomas. Early identification with fetal echocardiography, associated with postnatal MRI tissue characterization, enables prompt postnatal treatment with lifesaving surgical resection of the mass when significant hemodynamic compromise or life-threatening arrhythmias occur.4

Disclosures

None.

References


Figure 4. Intraoperative view. A, The large solid mass is exposed after median sternotomy. B, Operative view of the tumor during resection. LV indicates left ventricle; RV, right ventricle.

Figure 5. Histopathologic study. A, Enlarged swollen myocytes with clear cytoplasm and centrally located nucleus, connected with the periphery of the cell by strands of cytoplasm (spider cells) (hematoxylin-eosin stain, ×80). B, the enlarged cells are vacuolated because of the abundant glycogen deposits in the cytoplasm (Alcian PAS stain ×160); C) immunohistochemistry reveals striated muscular cells in keeping with cardiac rhabdomyoma (myosin ×160).
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