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Intracardiac tumor causing left-ventricular outflow-tract obstruction in a newborn



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The following report describes the case of newborn girl with an asymptomatic systolic murmur, which on imaging revealed a nearly obstructive mass in the left-ventricular outflow tract. The mass was resected and found to be consistent with a rhabdomyoma. Here, we describe the pathologic and clinical characteristics of this tumor.

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Keywords: Intracardiac tumor, Rhabdomyoma, Congenital heart disease

Case report

A newborn girl was evaluated in the first few hours of life for an asymptomatic systolic murmur. Three-dimensional transthoracic echocardiography revealed a nearly obstructive 9 mm solitary left-ventricular outflow-tract mass arising from the interventricular septum, which moved against the aortic valve during systole (Fig. 1). Doppler echocardiography demonstrated a small patent ductus arteriosus with left to right flow and no evidence of retrograde diastolic flow in the transverse aortic arch. The infant remained stable on room air with no evidence of metabolic

acidosis or arrhythmia. On Day 3 of life, resection of the intracardiac mass was performed via aortotomy. A white, fibrous, rather friable mass was identified abutting the right coronary cusp. The mass was resected in its entirety with needle-point electrocautery. Postoperative echocardiography demonstrated no residual lesion without evidence of aortic stenosis or insufficiency (Fig. 2). Pathology was consistent with rhabdomyoma (Fig. 3). The infant was extubated within 24 hours of the procedure. A genetic evaluation, including brain magnetic resonance imaging and renal ultrasound was completed to rule out tuberous sclerosis, and a serum tuberous sclerosis panel was negative. The patient was discharged

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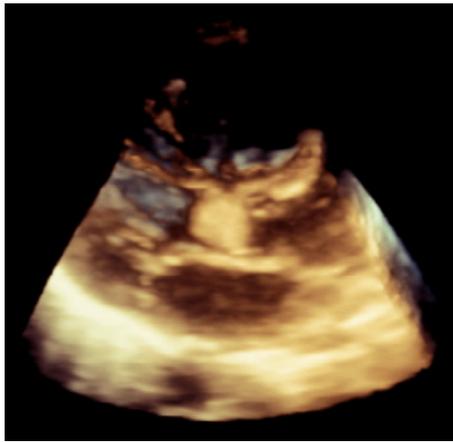


Figure 1. Echocardiogram. Three-dimensional transthoracic echocardiography (parasternal long axis) demonstrates a large mass extending from the interventricular septum into the subaortic region of the left-ventricular outflow tract.

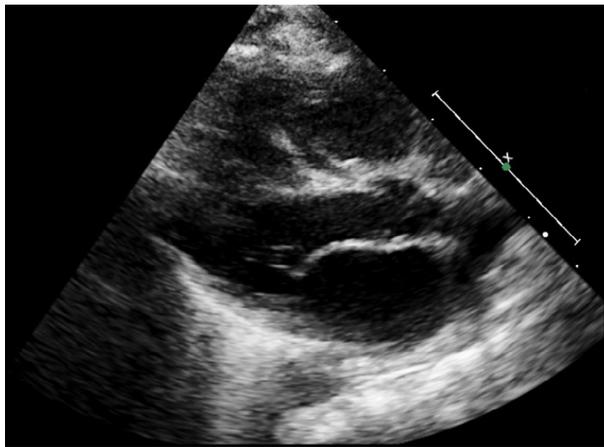


Figure 2. Echocardiogram. Transthoracic echocardiography reveals postoperative resolution of the outflow-tract obstruction.

to home on postoperative Day 10 in sinus rhythm. At 12 months following surgical resection, the child is doing well with no evidence of tumor recurrence.

Discussion

Rhabdomyomas comprise the majority of intracardiac tumors in the newborn, with more than half associated with tuberous sclerosis [1]. Atrial myxomas, fibromas, and pericardial teratomas comprise the remainder of tumors occurring with extreme rarity [2,3]. Although rhabdomyomas may be bilateral and the majority are multiple, a meaningful proportion of these masses are solitary. These tumors tend to be well-demarcated and histologically are composed of enlarged cells,

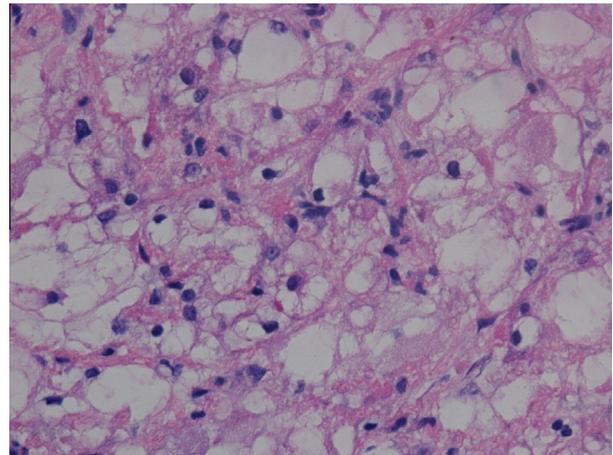


Figure 3. Rhabdomyoma. High-power view (40 \times) of large, vacuolated cells and a "spider cell" (cell with a central nucleus from which radial wisps of cytoplasm extend to the cell membrane). Special stains are most often not necessary due to the striking features of the neoplasm.

mostly with clear or vacuolated cytoplasm, or pale pink cytoplasmic granular material. "Spider cells" (Fig. 3) are the classic feature, although they may not always be appreciated. Mitoses or atypical nuclear features are not present.

Clinical presentation can vary from an incidental finding to symptoms of congestive heart failure or life-threatening arrhythmias. Cases presenting with complete cardiovascular collapse and sudden death have also been reported [4]. The natural history of cardiac rhabdomyomas is spontaneous regression, and as such, most incidentally found tumors can undergo surveillance with echocardiography. Indications for resection include outflow tract or cavitory obstruction and intractable arrhythmia [2,3].

The patient discussed presented with a tumor causing near complete obstruction of her left-ventricular outflow tract during systole. As such, it was felt that this mass had high potential to cause hemodynamically significant obstruction at any point. Given this risk, surgical resection was undertaken to prevent the potential of catastrophic cardiovascular collapse. Although successful conservative management of smaller, less problematically placed tumors is well described [3], we present this case to highlight the importance of aggressive surgical management in patients with echocardiographic findings to suggest the potential of hemodynamic compromise.

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