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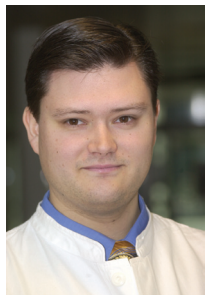
# Rhabdomyoma Operation—Intraoperative Video of a Very Rare Tumor Entity



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Primary cardiac tumors are a rare entity, with an incidence ranging from 0.001% to 0.03%. Among these tumors, congenital rhabdomyomas are the most common benign tumors of the fetal heart. Because of their tendency for regression, surgical removal is rarely indicated [Bosi 1996; Verhaaren 2003; Günther 2008]. In the present case, a third child was born at full term to healthy parents. Body measurements were normal (weight, 3120 g; length, 53 cm). A routine examination revealed a systolic murmur. An echocardiography evaluation showed situs solitus and atrioventricular concordance; however, several tumors were detected in the left and right ventricular cavum. Within the left ventricle, one of the tumors (approximately 1.3 × 0.8 cm) was located within the outflow tract and originated from the lateral wall; another tumor (approximately 0.9 × 1.2 cm) was lateral to the mitral annulus (Video I online). A third tumor (approximately 0.4 × 0.8 cm) was located in the right ventricle and originated from the interventricular septum, and a fourth was located within the outflow tract (approximately 1.0 × 1.1 cm). During the hospital stay, the patient's oxygen saturation dropped on various occasions. The maximum Doppler gradient within the right ventricular outflow tract (RVOT) was 40 mm Hg. Because of the continuous aggravation of the respiratory situation, urgent relief of the high degree RVOT obstruction was considered the only therapy of choice. The operation was performed on day 20 after delivery. Cardiopulmonary bypass was instituted, the aorta was clamped, and cold crystalloid cardioplegia was administered in an antegrade fashion. The cross-clamp time was 14 minutes. Access was achieved through the tricuspid valve. We found an almost round and even-surfaced tumor, approximately 1.0 × 0.5 cm in size, that expanded from the infundibulum to the pulmonary valve (Video II online).

After careful resection of the broad origin, the tumor was removed in toto. Intraoperative transesophageal echocardiography control showed no residual tumor within the RVOT

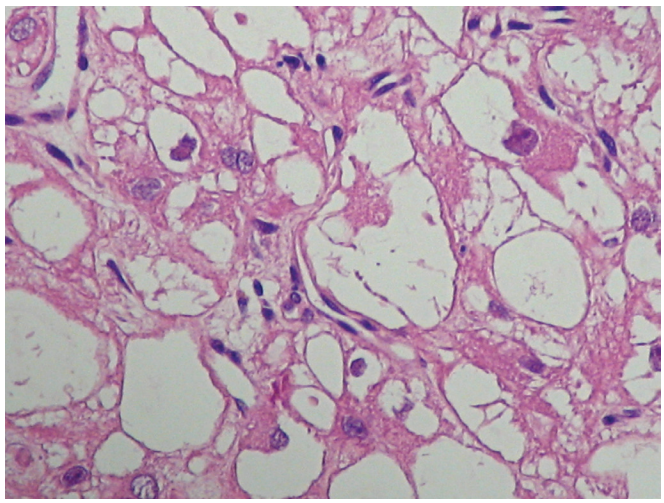

Figure 1. Hematoxylin and eosin staining shows large polygonal cells with ample eosinophilic and sometimes vacuolated cytoplasm and small round nuclei. Necrosis or mitoses are absent.

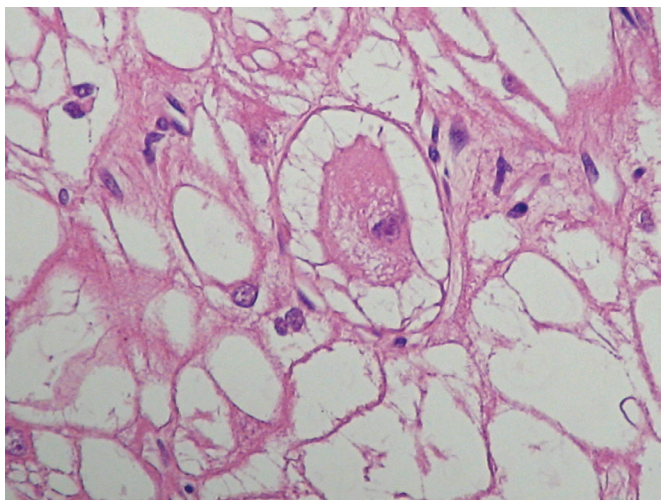

Figure 2. Cardiac rhabdomyoma cell with vacuolated cytoplasm, with strands of cytoplasm extending to the cell periphery ("spider cell").

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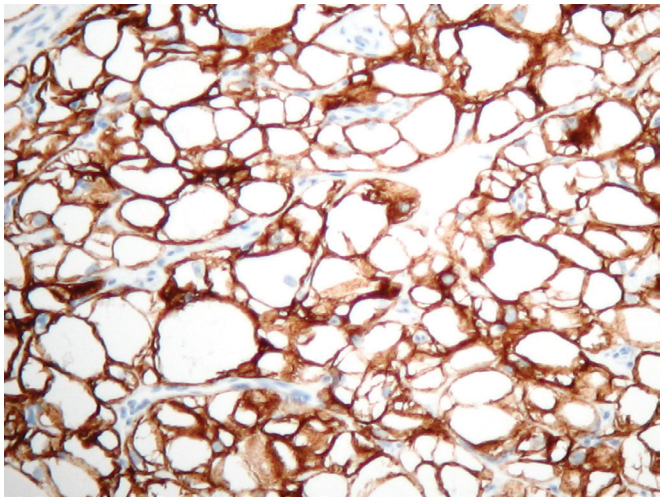

Figure 3. Desmin immunohistochemical stain demonstrating intense positivity in lesional cells.

and no Doppler gradient. The patient's postoperative course was uneventful. Recovery of the neonate was normal. A pathologic examination confirmed the diagnosis of a rhabdomyoma (Figures 1-3). An echocardiography evaluation documented no residual obstruction at the level of the RVOT and showed normal valvar function.

Although spontaneous tumor regression in rhabdomyoma is common and therefore the necessity for surgical procedures is extremely rare [Stiller 2001; Schreiber 2006; Günther 2008], some life-threatening hemodynamic situations may require instantaneous surgical intervention as rescue therapy.

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