Myofibroblastic tumor of the heart: A rare intracardiac tumor

Kimberly L. Gandy, MD, PhD,^a Matthew A. Burtelow, MD, PhD,^b V. Mohan Reddy, MD,^a and Norman H. Silverman, MD,^c Stanford, Calif

Cardiac tumors in infancy are rare, with an incidence of less than 1 in 10,000. The types of cardiac tumors found in adults differ from those found in children, with myxomas being the most common tumors found in adults and rhabdomyomas being the most common tumors found in children. We report a case of a rare intracardiac tumor, an inflammatory myofibroblastic tumor, in a 2-month-old child, this being the eighth reported case in the literature. We also review the differential diagnosis and treatment of cardiac tumors in children, with specific reference to the role of echocardiography.

Clinical Summary

A 2-month-old, asymptomatic 5.6-kg girl had a heart murmur. Preoperative echocardiography documented a solitary 2×2.5 cm mass attached to the right atrial free wall (Figure 1, A and C).

From the Division of Pediatric Cardiac Surgery,^a the Department of Pathology,^b and the Division of Pediatric Cardiology,^c Stanford University School of Medicine, Stanford, Calif.

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Address for reprints: Kimberly Lynn Gandy, MD, PhD, 300 Pasteur Dr, Falk Cardiovascular Research Institute, Stanford University Medical Center, Palo Alto, CA 94305 (E-mail: kimberly@stanford.edu).

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There was no associated superior or inferior vena caval obstruction, although the tumor appeared to obstruct the tricuspid orifice. No other intracardiac, visceral, or extremity soft tissue masses were appreciated. Our initial diagnosis was that of an atrial myxoma.

Dual venous cannulation was used for cardiopulmonary bypass. On opening the right atrium, we noted that the tumor was attached to the atrial appendage (Figure 1, B). The mass was resected en bloc (Figure 2, A) with the attached pectinate muscle and a small piece of right atrial free wall. A small patent foramen ovale was closed. Recovery was unremarkable. The patient is well and free of recurrence, as determined by echocardiography, at 1 year.

Pathologic examination of the mass revealed a grossly smooth and homogeneous mass without evidence of necrosis or hemorrhage. The tumor was composed of a proliferation of cytologically bland fibroblastic cells in random arrays without a discrete fascicular pattern within a myxoid background (Figure 2, B). Immunohistochemical staining confirmed the diagnosis of an inflammatory myofibroblastic tumor (Figure 2, C).

Discussion

Cardiac tumors are rare in children, making the analysis of incidence, clinical history, and treatment problematic. There are 2 large series of intracardiac masses reported to date, both recently reviewed by Becker.¹ Burke² reported a series from the Armed Forces Institute of Pathology and reported 386 tumors, 55 of which arose in infants and children. Becker reported a series from the Academic Medical Center in Amsterdam and reported 113 tumors, 21 of which originated in children. Recent case series report that

Figure 1. Echocardiographic analysis and appearance in situ. **A**, Four-chamber apical view showing a large homogenous tumor occupying the right atrium prolapsing through the tricuspid orifice in diastole. **B**, Intraoperative view at the time of surgical excision showing the tumor still attached to its stalk from the crista terminalis. **C**, Intraoperative transesophageal image in the long-axis view of the right atrium showing the mass with a peduncular attachment to the right atrial wall in the region of the crista terminalis. **D**, Image of the right atrium as for panel B after separation from cardiopulmonary bypass. LA, Left atrium; T, tumor; RV, right ventricle; LV, left ventricle; SVC, superior vena cava; RA, right atrium.

Figure 2. Pathologic analysis of the tumor. A, The tumor is smooth and homogeneous in gross appearance. B, Hematoxylin and eosin staining demonstrates the cellular configuration of the tumor with the myxoid background. C, Special stains of the myofibroblasts reveal evidence of smooth muscle actin (shown) and no evidence of desmin, S100, and cytokeratin Ael/CAM5.2 (not shown).

rhabdomyomas are the most common tumors in infancy, with fibromas and teratomas being the second and third most common, respectively. The remaining tumors are rare and include heman- giomas, myxomas, lipomas, and Purkinje cell tumors. Primary heart tumors are rare, and metastatic lesions are 10 to 20 times more common than primary malignant tumors.

With the advances in infant echocardiography and the advent of fetal ultrasonography, an increase in primary cardiac tumors has been reported.³ Most cardiac masses in children are now diagnosed before the second year of life, with most of the rhabdomyomas and fibromas being found in the first year of life in the aforementioned series.¹ Previous reviews reported that most rhabdomyomas were

associated with the tuberous sclerosis complex and that most rhabdomyomas found in children less than 4 years of age regressed in size over time. Most reviews therefore recommend nonoperative management of asymptomatic cardiac masses in children, with the expectation that most tumors are rhabdomyomas and will regress with time.¹ Resection is recommended for cases associated with hemodynamic or respiratory compromise, severe arrhythmia, or embolic potential.^{1,4} When resection is recommended, most suggest removal of as much tumor as possible while preserving cardiac function, even if some tumor is necessarily left behind.⁵

The tumor described in this case report is an inflammatory myofibroblastic tumor, which is synonymous with an inflammatory pseudotumor (IPT), a rare entity with only 7 previously reported cases, as reviewed by Li and associates.⁶ IPTs show a predilection for the atria, and approximately 25% recur locally,⁶ most of which appear to behave in a benign fashion. These tumors never metastasize, but a small percentage show the potential for malignant transformation over time. At present, there are no reliable clinical or pathologic predictors for the behavior of IPTs. Tumors, however, that show evidence of cytologic atypia or increased mitotic activity should be treated with greater suspicion.

We believe that the treatment algorithm previously described should be modified to further reflect clinical presentation and imaging characteristics suggesting that the tumor might not be a rhabdomyoma or fibroma. When the tumor is not associated with the tuberous sclerosis complex, is solitary, involves the atrial free wall, and does not decrease in size within 2 months after diagnosis, consideration should be given to early operative excision.

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