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Primary Cardiac Fibroma: A Rising Giant in a Small Cavity—Size Does Matter

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ABSTRACT

We report the case of a 5-month-old girl who presented with cardiac failure due to a cardiac fibroma originating from the interventricular septum. We also include some remarks about the etiology and the most favorable therapeutic strategy for the disease.

INTRODUCTION

Primary cardiac tumors in infants and children are extremely rare and reflect a benign process in more than 90% of the cases. Because of their rare prenatal diagnosis, rapid growth, and crucial localization, large cardiac fibromas often present with cardiac arrhythmias and heart failure, and they can lead to sudden death. We report the case of a 5-month-old girl who presented with cardiac failure due to a cardiac fibroma originating from the interventricular septum.

CASE REPORT

A 5-month-old girl was referred to our hospital with a diagnosis of an intracardiac mass originating from the interventricular septum and causing biventricular obliteration. The girl weighed 5.2 kg and was a firstborn of healthy parents without any familial history of cardiac disease or genetic anomalies. Both the pregnancy and childbirth had been unremarkable, and prenatal ultrasounds were normal. A suspicion arose when failure to thrive became undeniable at the age of 18 weeks and was accompanied by cyanotic spells during feeding. On admission, the patient was dystrophic, tachycardiac, and tachypneic and showed signs of chronic cardiac decompensation.

Chest radiography revealed impressive cardiomegaly (computed tomography index of 0.8). Echocardiography revealed a septal mass of 52 × 38 mm and signs of chronic decompensation (Figure 1A). Magnetic resonance imaging (MRI) confirmed the presence of a large tumor arising from the inferoseptal myocardium and inferior wall. The tumor was isointense in T1-weighted MR images and slightly hyperintense in T2-weighted MR images (Figure 1B).

Heart transplantation was considered, but because of rapid deterioration in the patient's general condition, we performed urgent debulking through a left ventriculotomy (Figures 2 and 3). Examination of a frozen section confirmed the tentative diagnosis of myofibroma obtained in the radiology evaluation.

The postoperative course was complicated by arrhythmias, impaired left ventricular function, and catheter sepsis that required extracorporeal life support (ECMO) and the administration of broad-spectrum antibiotics. On the sixth postoperative day, we performed a second intervention with the resection of another 4 g of the tumor. At the same time, we weaned the patient from ECMO; however, she developed life-threatening arrhythmias 1 day later that required another ECMO run for 4 days. We extubated the patient 2 days later and discharged her from the intensive care unit 1 week later. The patient's subsequent recovery was uneventful, and she was discharged 6 weeks after the primary hospital admission and was receiving beta-blockers, an angiotensin-converting enzyme inhibitor, and a diuretic. One year later, the patient maintains a stable cardiac condition with normal neuromotor development, but she manifests a psychogenic aversion toward oral feedings.

Histologic and immunohistochemical studies of the 44-g homogeneous white mass showed a proliferation of spindled cells surrounded by scarce elastic fibers, which indicated a recent onset for the growth of the cardiac fibroma. There was focal expression of α smooth muscle actin, desmin, and vimentin (Figure 4).

COMMENT

Cardiac fibromas are rare benign tumors, accounting for approximately 3.2% of all tumors of the heart and pericardium [Furguson 1996]. In the infant and child, cardiac fibroma is

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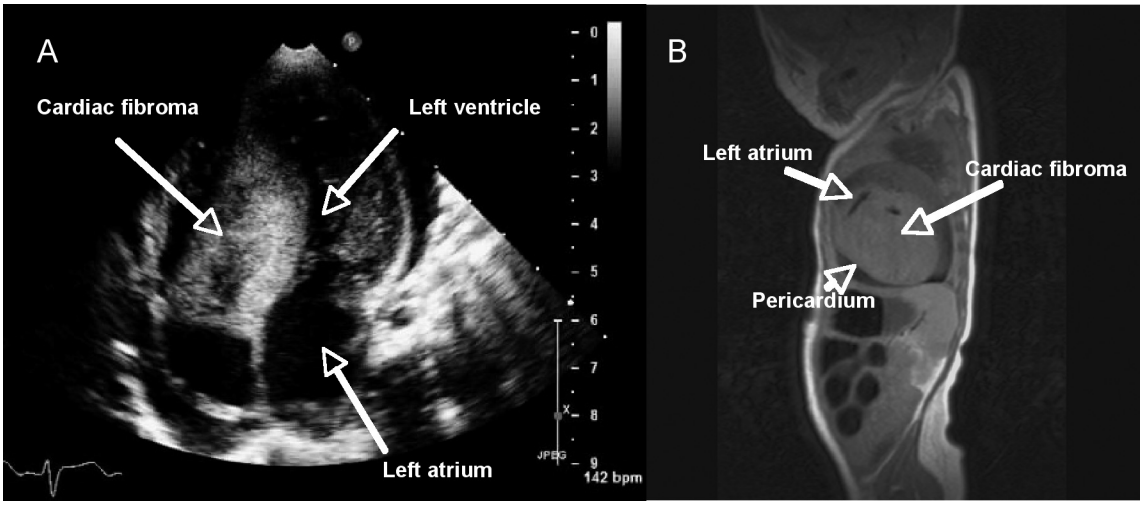

Figure 1. Cardiac fibroma in a 4-chamber echocardiography evaluation (A) and slightly hyperintense in T2-weighted magnetic resonance images (B).

the second most common benign cardiac tumor, following rhabdomyoma [Furguson 1996]. There is no sex predominance, and the frequency of such tumors tends to decline with age. These tumors tend to grow rapidly [Padallino 2005]. Clinical findings depend on the size and location of the tumor and the impact on blood flow and myocardial function [Isaacs 2004]. Detection is rarely incidental. Patients mostly present with congestive heart failure, respiratory distress, cyanosis, cardiac arrhythmias, or sudden death. The evolution and refinement of noninvasive imaging techniques, such as ultrasonography, echocardiography, MRI, and multidimensional echocardiography, have allowed optimal visualization and decision making at earlier stages of tumor development.

Although some cardiac tumors are detectable as early as 20 weeks of gestation, most tumors belong to the group of late-onset congenital heart diseases and first become apparent during the third trimester [Geipel 2001].

Unlike rhabdomyomas and teratomas, prenatal diagnosis of cardiac fibromas has been rarely reported.

Pathologically, cardiac fibromas are solitary, solid, unencapsulated tumors in the ventricular septum or in the free wall of the ventricle [Isaacs 2004]. Microscopically, they represent a benign proliferation of connective tissue arising

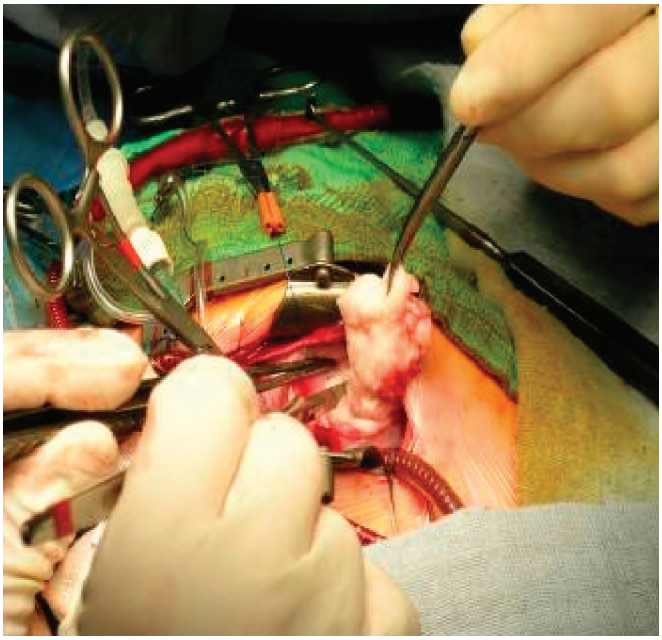

Figure 2. Intraoperative view.

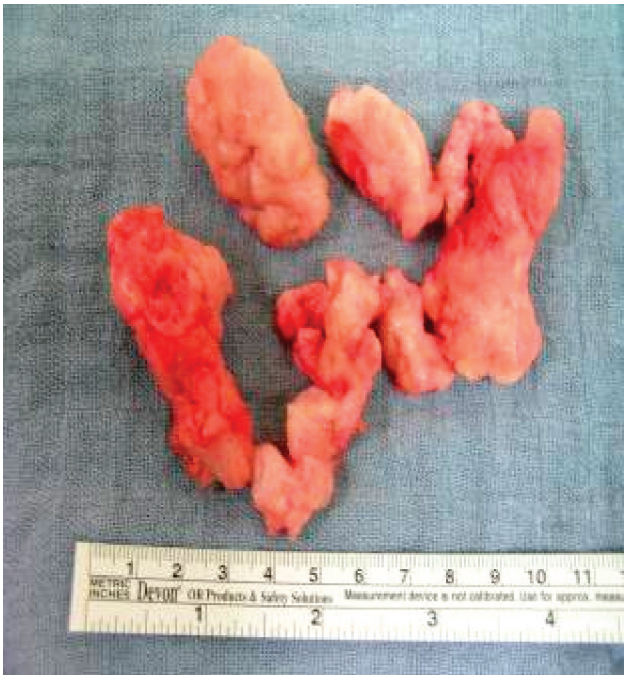

Figure 3. Gross appearance of the left ventriculotomy.

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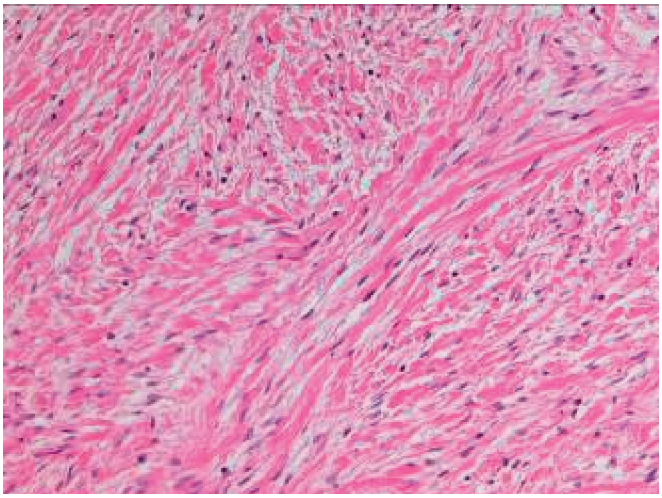

Figure 4. Cardiac fibroma showing uniform spindle-shaped, fibroblastic cells surrounded by a matrix of scarce fibrous connective tissue.

from fibroblasts and myofibroblasts that blends in with or infiltrates the myocardium of the ventricular septum or free wall of the right or left ventricle. Nevertheless, the etiology and the biological behavior of cardiac fibromas have not yet been fully elucidated.

Belying the axiom that benign neoplasms have normal (ie, diploid) karyotypes, improvements in cytogenetic methodologies and more frequent karyotyping have revealed that even benign tumors can harbor chromosomal abnormalities.

Recent molecular genetic investigations of Gorlin-Goltz syndrome, an autosomal dominant disorder characterized by body overgrowth, skeletal abnormalities, and benign and malignant neoplasms (ie, cardiac fibromas), have revealed a mutation in the PTC gene [Hahn 1996]. Apart from its function as a tumor suppressor gene, the PTC gene plays an important role in development by encoding a transmembrane protein that represses the Hedgehog signaling pathway, which controls cell fate, growth, and development [Ingham 2000].

Furthermore, there are no data on what triggers this rapid tumor growth. The fact that most cardiac fibromas present with congestive heart failure or life-threatening arrhythmias caused by undiagnosed formation of an obliterating solid tumor in the interventricular septum or ventricular free wall suggests a rapid, mostly postnatal tumor growth. In our case, we resected 44 g of tumor from the heart, the total mass of which is estimated to be 29 g for a 5-month-old infant.

The therapeutic strategies for cardiac fibroma should be individualized, with the options varying from observation to partial resection to heart transplantation. Because cardiac fibromas do not regress spontaneously and malignant transformation has not been reported, one can advocate for a watchful waiting policy in asymptomatic patients [Cope 1994]. A total resection should be attempted when the disease becomes symptomatic [Cope 1994], but one should keep in mind that total resection is not the only therapeutic end point. The most important consideration is the restoration of heart function. Large fibromas are often treated by subtotal resection or primary transplantation after biopsy. If debulking restores normal hemodynamic and respiratory function, the long-term outcome has proved to be favorable [Cope 1994]. Several studies have shown no further growth following partial resection [Gowdamarajan 2000].

Cardiac transplantation can be considered whenever hypoplastic heart syndrome or very large tumors are detected and when there is risk of destroying a vital part of the conducting system and subsequently jeopardizing normal cardiac function [Burke 1994].

Although Isaacs and colleagues reported a survival rate for cardiac fibroma in neonates of only 23% [Isaacs 2004], recent studies have indicated excellent mid-term results after partial resection of such fibromas, with no recurrence of disease [Gowdamarajan 2000].

CONCLUSION

We have reported the case of a 5-month-old infant with cardiac decompensation caused by an obliterating septal cardiac fibroma. Early follow-up indicates that partial resection can be a successful surgical treatment for cardiac fibroma.

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