



## Intraluminal Ascending Aorta Fibroma

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Received 4 February 2010; Accepted 14 June 2010

### Abstract

*Primary cardiac tumors are quite rare, especially in the pediatric age group, and their atypical presentations often prevent a timely diagnosis. Most primary cardiac tumors in the pediatric age group are benign. Fibromas are generally reported as the second most common primary cardiac tumors in the pediatric age group. These neoplasms are often intramural and involve the left ventricular free wall or the interventricular septum. Although benign, fibromas may become life-threatening by causing arrhythmias or obstruction to the blood flow. A case of supravulvar intraluminal ascending aorta fibroma in a 23-month-old girl, presenting with syncope, is described here; the location is rare and the presentation atypical for this type of tumor. Transesophageal echocardiography helped us to evaluate the anatomic details of the tumor and plan surgery.*

*J Teh Univ Heart Ctr 2011;6(1):45-47*

**This paper should be cited as:** Aarabi Moghadam MY, Moradian M, Givtaj N, Mozaffari K. Intraluminal Ascending Aorta Fibroma. *J Teh Univ Heart Ctr 2011;6(1):45-47.*

**Keywords:** Heart neoplasms • Fibroma • Syncope

### Introduction

Primary cardiac tumors are quite rare, especially in the pediatric age group,<sup>1-3</sup> and their atypical presentations often prevent a timely diagnosis.<sup>4</sup> Most primary cardiac tumors in the pediatric age group are benign; autopsy studies in children have reported incidence rates ranging from 0.027% to 0.08%.<sup>4</sup> One echocardiography database has reported an incidence of 0.17%, which suggests that one or two new primary cardiac tumors will be detected for every 1000 first-time pediatric echocardiograms.<sup>4</sup>

Fibromas are generally reported as the second most common primary cardiac tumor in the pediatric age group.<sup>3</sup> However, in a more recent review of Boston Children's Hospital database from 1980 to 2005, fibromas were the third most common tumor. To date, no distinct genetic inheritance or familial predisposition has been associated with cardiac

fibromas. These neoplasms are often intramural and involve the left ventricular free wall or the interventricular septum. Less frequently, they can be multiple and invade the right ventricular free wall, the atrial septum, or its free wall. Although rare, intracavitary fibromas have also been reported.<sup>4,6</sup>

It is very rare for primary intracardiac tumors to occur in the supravulvar pulmonic or aortic positions.<sup>7</sup> A case of a seven-year-old boy with an intraluminal pulmonary artery fibroma was reported in the *Pediatric Cardiology Journal* (21: 480-482 2000) from Southwest Texas Methodist Hospital. We herein present the case of a girl with an intraluminal fibroma in her ascending aorta.

### Case Report

We present a 23-month-old girl admitted for the evaluation

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of a cardiac murmur with a history of one episode of syncope two months before admission. She had no positive history for syncope and no positive family history for congenital heart diseases or any kind of cardiac tumors.

On physical examination, her growth and development were within normal limits (height = 85 cm, weight = 11 kg). The central and peripheral pulses were normal and no cyanosis was detected. Her blood pressure was 90/60 mmHg. On heart auscultation, there was a grade 3/6 systolic ejection murmur at the second right inter-costal space with radiation to the neck. Electrocardiography revealed normal sinus rhythm with left ventricular hypertrophy. Her heart rate was 110 beats per minute. The chest X-ray showed mild cardiomegaly with normal vascular markings.

Transthoracic echocardiography demonstrated mild left ventricular hypertrophy and a large intraluminal mass in the ascending aorta, but its borders were not obvious. Doppler interrogation of the ascending aorta showed a turbulent, high-velocity ante grade flow with a 66 mmHg peak pressure gradient and a 46 mmHg mean pressure gradient. By color mapping, turbulency began at the level of the aortic valve.

In order to confirm the transthoracic echocardiography findings, transesophageal echocardiography was performed, which confirmed the presence of a large intraluminal mass in the ascending aorta. The mass was elongated and occupied about 60% of the ascending aorta's area (3 cm × 1 cm × 1 cm). It was heterogeneous, lobulated, and immobile. The mass had attachments to the anterior cusp of the bicuspid aortic valve. The turbulent antegrade flow around the margins of the mass was obvious and there was no more adherence. The distal end of the mass juxtaposed the initiation of the transverse aorta (Figure 1).

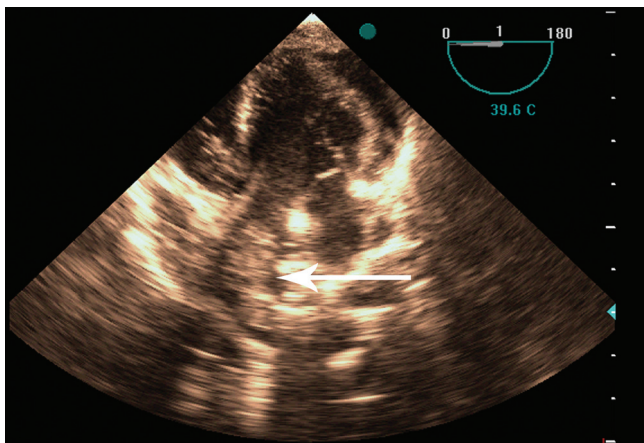


Figure 1. Transesophageal echocardiography. Deep transgastric long-axis view showing the tumor. At least 60% of the ascending aorta's circumference is occupied

The high likelihood of the mass being tumoral and its hazardous location precluded catheterization, and the child

underwent surgery for mass resection. Given the position of the mass, cannulation via the femoral artery, inferior vena cava, and superior vena cava was done. With cerebral protection during cardiopulmonary bypass, the arteriotomy of the ascending aorta was preformed. The mass was found to be entirely within the ascending aorta with firm adherence to the anterior leaflet of the bicuspid aortic valve (Figure 2).



Figure 2. The tumoral mass, sectioned for removal

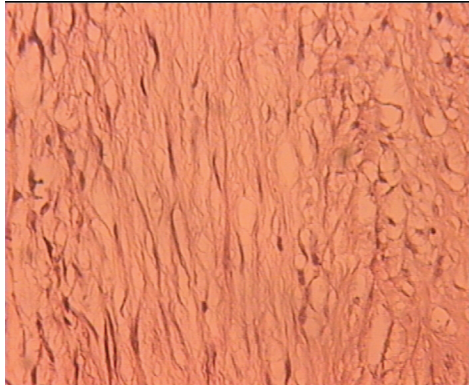
The tumor was excised subtotaly because it could not be detached from the aortic valve leaflet. The arteriotomy was closed, and the child was weaned from the cardiopulmonary bypass machine without any difficulty.

Repeat transthoracic echocardiograms following the operation did not reveal any evidence of residual supravalvular aortic stenosis and there was no aortic insufficiency.

In gross pathology description, the mass was solid, firm, and creamy with a nodular appearance (3 × 1 × 1 cm). Microscopic sections showed a benign neoplasm composed of fascicles of fibroblasts with variable amounts of collagen and a scanty number of lymphocytes arranged in a focally myxomatous stroma without any evidence of malignancy such as increased mitotic figures or areas of necrosis (Figures 3A & B).



A



B

Figure 3. A & B show fascicles of fibroblasts in a focally myxomatous stroma without any evidence of malignancy (H & E  $\times$  400)

## Discussion

The most common tumors in newborns and infants are rhabdomyomas, fibromas, and intrapericardial teratomas; whereas in older children and adolescents, myxomas, rhabdomyomas, and fibromas are prominent.

Rhabdomyomas constitute 45% to 80% of all primary cardiac tumors in the pediatric age group. These tumors can be diagnosed in the prenatal period but are most frequently diagnosed in the newborn infant. Although fibromas have recently been reported in utero and in patients younger than 1 month of age, they are found much less commonly than rhabdomyomas in this age group. In a recent review at Boston Children's Hospital, fibromas were the second most common tumors (17%) in patients diagnosed between 1 month and 1 year of age. These primary tumors are rarely seen in older children, adolescents, or young adults.<sup>4</sup> No known sex predilection is recognized, although the rarity of different benign cardiac tumors prevents an accurate determination of a male-to-female ratio.<sup>6</sup> On two-dimensional echocardiography, cardiac fibromas are seen as a single, bright, intramural, echogenic mass. CT scanning is often performed and might provide clues regarding tissue characterization, with central calcification suggestive of a cardiac fibroma.<sup>6</sup>

This case has two interesting aspects: presenting symptom of the tumor and its location.

Our patient was previously healthy with a normal development, and she sought medical attention after a syncopal event and a murmur was noted on the auscultation of the heart.

The presence of fibroma in the ascending aorta is extremely unusual. Fibromas are predominantly intramural tumors, and extensive intramural fibromas can encroach and obliterate the intracavitary space. A similar case has been previously

reported with an intraluminal fibroma in the proximal main pulmonary artery.<sup>8</sup>

Although benign, fibromas may become life-threatening by causing arrhythmias or obstruction to blood flow.<sup>8, 9</sup> Acquired supra-ventricular aortic stenosis and syncope due to the tumor were the presenting features of our patient.

Although complete resection is preferable, our patient's tumor was not amenable to complete resection due to its firm adherence to the anterior cusp of her bicuspid aortic valve.

## Conclusion

Syncope can be the presenting symptom of the tumors of the intraluminal ascending aorta as was the case in the patient described here with an unusual location of fibroma. Transesophageal echocardiography is useful for the evaluation of anatomic details and can be of assistance for surgical planning.

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