Fibroma of the Right Ventricle Producing Severe Tricuspid Stenosis

By Luke G. Van der Hauwaert, M.D., Lucien Corbeel, M.D., and Paul Maldague, M.D.

The great majority of benign tumors of the heart are myxomas. They usually affect adults, are localized in the atria, and may produce intermittent obstruction of the atrioventricular valves. Much rarer are fibromas, also designated by the term fibromyoma, fibromyxoma, fibroelastic hamartoma, or hamartoma. According to Freeman et al. only 27 cases had been reported up to 1962. Two more case reports have been published since.

Whereas previously most cardiac tumors were incidental autopsy findings, in recent years an increasing number has been diagnosed in life, and a few have been operated upon successfully.

The present case seemed worth recording in view of the following features: (1) unusual presenting symptoms, typical of severe tricuspid stenosis, (2) unexplained eosinophilia, (3) correct hemodynamic and angiocardio- graphic diagnosis of space-occupying lesion of the right ventricle, (4) attempt of surgical treatment, which however failed because of the extensiveness of the tumor.

Case Report

The patient, a 16-month-old boy had been perfectly well until a few weeks before admission when he became listless and uncooperative. The mother had noted puffiness of the face and associated this with a sudden weight gain of 800 Gm. within 2 weeks. A heart murmur had never been heard at the Infant Welfare Clinic where the boy had been examined during his first year of life.

On admission the child was restless and apparently in distress. His cheeks and extremities were slightly cyanotic. His face looked swollen but no frank edema was noted. The liver, felt 5 cm. below the costal margin, was strikingly pulsatile. The external recording of the hepatic pulsations, with the Elema plethysmograph, showed giant a waves (fig. 1). The spleen was not enlarged, and there were no adenopathies.

Figure 1

External recording of hepatic pulsations simultaneous with the phonocardiogram at the fourth left intercostal space. A giant a wave, starting 0.06 sec. after the onset of the P wave, is coinciding with the diastolic murmur recorded at the fourth left intercostal space.
pation the heart was quiet. A short grade II/VI ejection murmur was heard at the pulmonary area. The dominant auscultatory finding was a loud diastolic rumble ending in a presystolic accentuation, maximal at the fourth left intercostal space. The second sound was single.

The phonocardiogram, taken at the fourth left intercostal space (fig. 2A) demonstrated a diastolic murmur starting 0.06 sec. after the beginning of the P wave, its maximal intensity coinciding with the summit of a wave recorded over the hepatic surface (fig. 1). The murmur spilled over into early systole, at least if one assumed that the first sound recorded at the apex and coinciding with the S wave of the electrocardiogram (fig. 2B) represented the mitral closure sound and indicated the beginning of systole. At the fourth left intercostal space an extra sound, 0.06 sec. after the second sound, was registered. It may represent an opening snap of either tricuspid or mitral valve. Its origin could not be further elucidated.

The electrocardiogram (fig. 3) showed a frontal QRS axis of +90°. The P waves were peaked in II (4 mm.) and V2 (5 mm.). A QR pattern was seen in V3R. The amplitude of the R wave was abnormally high in the left precordial leads, being 34 mm. in V6. The tracing was interpreted as indicative of right atrial and biventricular hypertrophy.

On the chest x-ray film a grossly dilated heart was seen. The border of the right atrium was bulging. The conus of the right ventricle and the main branches of the pulmonary artery could not be identified.

The erythrocyte sedimentation rate was normal (1 mm./1 hr.). There was moderate anemia (hemoglobin 9.9 Gm. per cent, red cells 3,012,000 per mm.3), a reticulocytosis of 9 per cent and a leukocytosis of 80,000. The white cells were differentiated as follows: mature eosinophils 67 per cent, lymphocytes 24 per cent, neutrophils 5 per cent, basophils 2 per cent, and monocytes 2 per cent.

The sternal marrow contained predominantly precursors of the eosinophils: myeloblasts 2, promyelocytes type I 16, neutrophil promyelocytes type II 7, eosinophil promyelocytes type II 10, neutrophil myelocytes 1, eosinophil myelocytes 14, neutrophil metamyelocytes 5, eosinophil metamyelocytes 17, mature neutrophils 11, mature eosinophils 17, lymphocytes 9, proerythroblasts 1, basophilic normoblasts 3, polychromatic normoblasts 9, pyknotic normoblasts 8. Biochemical re-

Figure 2

Phonocardiogram registered at the fourth left intercostal space (A) and at the apex (B). At the fourth left intercostal space a murmur is recorded which is mainly diastolic but continues into early systole. The sound, which at the apex coincides with the S wave of the electrocardiogram, is considered to be the mitral closure sound and to indicate the beginning of systole.
FIBROMA OF RIGHT VENTRICLE

Figure 3

Electrocardiogram. Note the peaked P waves in II, V1, and V2, the QR pattern in V3R and the high R waves in V4 and V5 (one half of normal standardization).

Results, including transaminases, were all within normal limits. No parasites were found in fecal specimens.

Cardiac catheterization (table 1) was performed from the right saphenous vein. The catheter course was normal. Giant a waves of 18 to 20 mm. Hg were recorded in the right atrium. On the withdrawal tracing from the pulmonary artery to the right ventricle no systolic gradient was registered, while a considerable diastolic gra-

Table 1

Results of Right Heart Catheterization

<table>
<thead>
<tr>
<th>Site</th>
<th>Pressure, mm. Hg</th>
<th>O2 saturation, %</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>S</td>
<td>D</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>a = 18-20</td>
<td>x = 4-5</td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>a = 18-20</td>
<td>x = 4-5</td>
</tr>
<tr>
<td>Right atrium</td>
<td>16-20</td>
<td>6-5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>16-20</td>
<td>7-8</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>16-20</td>
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showed the latter to be considerably dilated. The inflow tract and the body of the right ventricle failed to fill. Through a narrow irregularly delineated channel the contrast material reached the normally developed infundibulum and pulmonary artery. Emptying of the right atrium was slow and with each atrial contraction a large amount of dye was regurgitating into the caval veins.

In view of the severity of the right ventricular inflow obstruction it seemed unlikely that the child would much longer survive unless the space-occupying lesion in the right ventricle were removed.

A medial sternotomy was performed and the pericardium incised longitudinally. The right atrium was greatly dilated and contracted forcefully. Only the infundibulum of the right ventricle showed normal contractions. On palpation the anterior surface of the right ventricle was immobile and of solid consistency. After having made a purse-string suture, a finger was introduced into the right atrium. The orifice of the tricuspid valve was felt to be almost completely occluded by a solid mass which was adherent to the tricuspid leaflets and firmly embedded in the myocardium of the right ventricle. Its removal was obviously impossible, even with the aid of extracorporeal circulation. Some tissue of the right ventricular wall was removed for biopsy. The postoperative course was complicated by tachycardia and tachypnea, which did not respond to digitalis. The child died 2 days after the operation.

Necropsy. The heart was greatly enlarged, weighing 130 Gm. The right atrium was enormously dilated and its trabeculation heavily developed. With the exception of the infundibulum, the cavity of the right ventricle was completely occupied by a firm, nonencapsulated, pink-white mass, which strikingly resembled a uterine myoma. No demarcation was seen between the tumor and the interventricular septum or the free wall of the right ventricle from which it seemed to have grown. The mass extended upwards into the right ventricular cavity, producing severe tricuspid stenosis. It saved only a narrow opening which allowed communication between the right atrium and the normal infundibulum. The pulmonary valve was normal. The endocardial surface of the tumor looked pearly white and impinged upon the tricuspid leaflets, which were thickened and distorted. The left atrium was normal. The left ventricle was slightly hypertrophied. Mitral and aortic valves were normal. The atrial septum was closed.

The liver showed changes of chronic passive congestion. The spleen was equally congested. Its reticulum was hyperplastic and its pulp engorged by mature eosinophils. Nowhere could leukemic infiltrations or metastases be found.

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Microscopically the cardiac tumor was composed of healthy looking myocardial fibers interlaced with dense bundles of connective tissue. In its subendocardial layers, where the tumor adhered to the tricuspid valve, it consisted almost exclusively of fibrocytes embedded in a loose collagenous framework.

Discussion

Although the tumor just described consisted mainly of myocardial tissue and should therefore be called myoma, it seemed justified to apply the generally adopted term of "fibroma." Indeed most tumors reported under this heading contained fibrous, collagenous, and myocardial elements in varying amounts. An identical problem of semantics arises with the uterine fibromas which, according to most pathologists, resemble the cardiac fibromas both macroscopically and microscopically. The term hamartoma was thought to be inappropriate in our case and should be reserved for nonproliferating embryonic malformations containing muscle, fibrous tissue, nerves, blood vessels, and fat. The histology of cardiac fibromas and related tumors has been reviewed by Bigelow et al.4 Adding two cases recently reported2,8 to the fibromas listed by Freeman et al.,1 a total of 29 cases may be collected from the literature. No sex predisposition is noted. The majority occurred in children. Twelve were found in infants below 1 year of age, 11 between 1 and 10 years, and six in older patients. Fibromas are more common in the left ventricle (22 cases) and in the interventricular septum (five cases) than in the right ventricle (two cases).

Sudden death, unexplained congestive heart failure, rhythm disturbances or a combination of these, are the most common presenting symptoms. Clinical data are scanty, as most cases were found at autopsy, reported by pathologists. A systolic heart murmur was noted by several authors.3-6 The electrocardiogram usually shows left ventricular hypertrophy, reflecting the common involvement of this ventricle. Right ventricular hypertrophy was found in one case in which the tumor caused pulmonary stenosis.8 Finally paroxysmal tachycardia of supraventricular origin7 and ventricular origin8 has been reported.

Radiologic examination may reveal gross, diffuse cardiomegaly or specific enlargement of the left ventricle. In two instances calcification within the bulging mass of the left ventricle was noted.7,8

Hemodynamic results were reported in one case.3 They were typical of infundibular pulmonary stenosis, the neoplastic nature of which was only discovered at operation.

The recognition of cardiac fibromas is obviously difficult in life, most clinical signs being nonspecific. Prior to the present report the preoperative diagnosis of cardiac tumor was made in only two of the 29 cases of fibroma found in the literature. In one6 the angiocardiogram showed leftward displacement of the left atrium and left ventricle. In the other2 it demonstrated a filling defect of the left ventricle. In two other cases6,9 the tumor was an operative finding. From the clinical and pathologic reports it appears that obstructing fibromas may mimic aortic stenosis5,7,10 or pulmonic stenosis.3 Myxomas, which very exceptionally arise from the right ventricle, may equally obstruct its outflow tract.11,12

As far as we know no previous case was described in which an intraventricular fibroma or myxoma produced tricuspid stenosis. Right ventricular inflow obstruction has of course been repeatedly reported in myxoma of the right atrium.

Marked eosinophilia, as found in the present case, has not been associated with any form of cardiac tumor. It should be mentioned that eosinophilic leukemia in its terminal phase, may involve the myocardium and produce large mural thrombi, leading to obstruction of the cardiac valves.13 This possibility, however, was ruled out by the sternal marrow smear and the autopsy findings.

Early recognition of benign cardiac tumors is no longer of purely academic interest. Since the introduction of extracorporeal circulation many atrial myxomas have been surgically removed. Successful excision of intraventricular myxomas has recently been reported.11,12,14 Furthermore, an intracavitary hamartoma was removed from the left ventricle in a 2-year-old child2 and from the
right ventricle in a 20-year-old girl. Only selective angiocardiography allows a correct diagnosis of intracardiac space-occupying lesion. It should be employed whenever a cardiac tumor is suspected.

**Summary**

A case of fibroma of the right ventricle in a child of 16 months is reported. The clinical and hemodynamic findings were those of severe tricuspid stenosis. Angiocardiography proved the obstruction to be due to a cardiac neoplasm, localized within the right ventricle. An attempt at surgical removal failed because the tumor, almost completely filling the cavity of the right ventricle, was too extensive and could not be distinctly delineated from the healthy myocardium.

**References**


**The Courage We Bring to Bear**

I am more and more convinced that our happiness or unhappiness depends far more on the way we meet the events of life than on the nature of those events themselves.—KARL VON HUMBOLDT.
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