Intramural Ventricular Cardiac Fibroma

Successful Removal in Two Cases and Review of the Literature

By Alexander S. Geha, M.D., William H. Weidman, M.D., Edward H. Soule, M.D., and Dwight C. McGoon, M.D.

SUMMARY

Two cases in which large intramural ventricular cardiac fibromas were successfully removed from children are reported. In one the fibroma was principally septal, and in the other it lay in the free wall of the left ventricle. These are the second and third such operations reported. A review of these two cases plus 34 other reported cases of cardiac fibroma indicates that a primary mural tumor of the heart should be suspected in patients, particularly children, with unexplained cardiac failure, unexplained cardiac dysrhythmia, intracardiac calcifications, irregular shadows on roentgenograms, or unexplained newly developing cardiac symptoms and murmurs. Angiocardiography is the best way of confirming the diagnosis. In view of the potential hazard of these tumors, their surgical removal should be undertaken, and this experience indicates that even very large and extensive intramural ventricular tumors may be removed successfully.

Additional Indexing Words:
Angiocardiography Cardiac dysrhythmia Intracardiac calcification
Cardiac hamartoma Cardiac tumors

The first successful surgical removal of a cardiac tumor, an intrapericardial teratoma, was reported by Beck. However, until Crafoord successfully removed a myxoma in 1954, the diagnosis of primary tumors of the heart was not followed by successful therapy. The development of cardiac surgery has prompted greater recognition of the problem, reflected in a shifting of interest in the subject from the pathological to the surgical literature during the last decade.

Although de Senar’s statement in 1783 that “the heart is an organ too noble to be attacked by a primary tumor” was proved wrong long ago, primary tumors of the heart are rare, and particularly uncommon among them is the intramural fibroma, of which only 34 cases have been reported. This tumor often leads to sudden, unexpected death or produces clinically obscure heart disease simulating particularly endocardial fibroelastosis, subaortic stenosis, or coronary artery disease. In four cases of intramural fibroma the diagnosis of ventricular tumor was made before autopsy or thoracotomy, and there is only one reported case of successful surgical removal of the tumor.

Mahaim attributed the first recognition of a cardiac fibroma to Colombo in 1559, but the first documented case is that of Luschka (1855). Because of the rarity of these tumors, Freeman and associates suggested that recording their occurrence is desirable in order to assist in early clinical recognition and possible surgical treatment of future cases and in study of their biological behavior. We have recently excised successfully a fibroma of the interventricular septum in a 27-month-old boy and another of the left ventricular wall in a 38-month-old boy and have observed the patients postoperatively for 29 and 10 months, respectively. In both cases the diagnosis was made preoperatively.

From the Mayo Clinic and the Mayo Foundation, Rochester, Minnesota.

Circulation, Volume XXXVI, September 1967 427
Report of Cases

Case 1

A 27-month-old boy was referred for evaluation of a heart tumor in June 1964. He was the product of a full-term, uncomplicated pregnancy and an uneventful delivery. At the age of 1 month he had a syncopal attack lasting for a few minutes and subsiding spontaneously. He was completely asymptomatic and thrived normally for the next year. At the age of 13 months he had another similar syncopal episode, and, at 19 months severe cardiac failure developed. An electrocardiogram at that time revealed paroxysmal atrial tachycardia with ventricular conduction delay. He was hospitalized and digitalized, with reversal of the dysrhythmia and cardiac failure. The medication was discontinued at 31 months of age. A month later a thoracic roentgenogram showed an enlarged heart with calcification in the region of the left ventricle. The Casoni test was negative. A clinical diagnosis of hamartoma of the heart was made, but a right ventricular angiocardiogram did not localize the lesion well. The child had been well since then except for a third attack of syncope, in March 1964, which was similar to the previous ones. Both parents and his two brothers, 14 and 12 years old, have always been in excellent general health.

On physical examination at the Mayo Clinic, the patient appeared to be small but looked healthy and well nourished. The pulse was 88 beats/min, the blood pressure was 110 mm Hg systolic and 70 diastolic, and the lungs were clear. The cardiac findings were normal. No murmurs were heard, and the heart sounds were normal. The results of hematological examination were within normal limits. A thoracic roentgenogram (fig. 1 left) revealed an enlarged, globular heart (cardiothoracic ratio, 0.66) with calcification in the left side of the cardiac silhouette. The electrocardiogram (fig. 2 left) showed ectopic atrial rhythm with delayed ventricular conduction (QRS interval, 0.24 second). Biplane right and left ventricular angiocardiograms demonstrated a large, partially calcified, intramyocardial tumor occupying the anterior and basal portions of the ventricular septum and the left aspect of the crista supraventricularis and extending downward over the anterolateral aspect of the left and right ventricles. In the anteroposterior projection, it formed most of the left aspect of the cardiac silhouette (fig. 3).

In July 1964 the patient was operated on.*

*We are indebted to Dr. Enrique S. Ballina of LaPlata, Buenos Aires, Argentina, who referred this patient to us, having already made the correct diagnosis.

*A motion picture of this operation was presented at the “Spectacular Film Series,” meeting of the American College of Surgeons, Atlantic City, New Jersey, October 1965.

Figure 1

Case 1. Thoracic roentgenograms. (Left) Preoperatively, showing cardiomegaly and calcification in left side of cardiac silhouette. (Right) Postoperatively, heart is still enlarged but there is no calcification.
through a median sternotomy, with extracorporeal circulation at 30°C. A large, firm mass was noted within the myocardium in the region of the interventricular groove. It was covered with thickened white epicardium in the region of the left anterior descending coronary artery. It extended from the apex to just caudal to the takeoff of the left anterior descending coronary artery, nearly reaching the pulmonary valve, and was embedded in the anterior half of the interventricular septum with a thin layer of uninvoluted muscle between it and each ventricular cavity (fig. 4). It had no true capsule but was surrounded by compressed muscle. A longitudinal superficial incision was made just anterior to the anterior descending coronary vessels which were retracted. The incision was deepened to the tumor, and dissection was tediously continued about it, leaving a very narrow rim of muscle attached to the tumor. During the dissection, two small perforations into the left ventricular cavity occurred, and the right ventricular cavity was entered rather extensively along the anterior attachment of the septum. After removal of the tumor, the attenuated layers of the interventricular septum were approximated, thus restoring the integrity of the septum. The ventricular fibrillation which had developed during perfusion reverted spontaneously to a sinus rhythm on rewarming. The anterior wall of the right ventricle was reattached to the anterior edge of the septum, and the anterior descending vessels were replaced in their natural positions. No difficulty was encountered in discontinuing the extracorporeal circulation, and the operation was completed without incident. The postoperative course was completely benign, the electrocardiogram showing a normal sinus rhythm with a P-R interval of 0.10 second and a QRS duration of 0.12 second (fig. 2 right), but a harsh, high-pitched, grade III (on the basis of I to VI) systolic murmur was noted along the left sternal border and was thought to be due to a small and probably hemodynamically insignificant interventricular communication. A roentgenogram of the thorax (fig. 1 right) showed the heart to be of the same size as it was preoperatively, but calcification was no longer evident and the pulmonary vascular markings were normal.

In the 29 months since the operation the patient has continued to have no symptoms and to enjoy normal activities. There have been no episodes of dysrhythmia, and an electrocardiogram made in March 1965 was normal and identical to the one made in the early postoperative period. The systolic murmur is now grade I.

The final diagnosis was fibroma of the interventricular septum. The tumor was firm, white, and fibrous; it was 6 cm in diameter and weighed 72 g (fig. 5). It resisted cutting, and calcified regions were noted along the cut surface. Microscopically, the tumor was composed of dense fibrous tissue, incorporating strands of cardiac muscle in its periphery (fig. 6 left).

Case 2

A 38-month-old Caucasian boy was referred for cardiac evaluation in January 1966. The child had been examined regularly by a pediatrician since birth and had been entirely well. A 4½-year-old sister died suddenly in August 1965, and an autopsy revealed the presence of a coarctation of the aorta and a patent ductus arteriosus. This prompted a cardiac examination of the patient and his siblings. An 8½-year-old sister and a 6-year-old brother were found to be normal, but the patient was noted to have a systolic murmur as well as cardiomegaly and calcification in the region of the left ventricle roentgenographically.

On physical examination at the Mayo Clinic, the patient looked healthy and did not have any
Figure 3

Case 1. Right ventricular angiograms, showing partially calcified tumor of septum causing filling defect in right ventricular outflow tract in region of left aspect of crista supraventricularis. (Left) Anteroposterior view. (Right) Lateral view.

Figure 4

Case 1. Artist’s illustration of tumor of upper interventricular septum.
Figure 5
Case 1. Gross appearance of tumor; weight, 72 g. Tumor resembled a desmoid tumor grossly and exhibited small areas of dystrophic calcification on the cut surface.

Figure 6
(Left) Case 1, showing cellular fibroblastic nature of tumor with an area of calcification. Hematoxylin and eosin; ×100. (Right) Case 2, showing similar fibrous pattern with some pigmented myocardial fibers entrapped at periphery of tumor. Hematoxylin and eosin; ×200.

obvious manifestations of a cardiovascular disorder. The pulse was 110 beats/min, the blood pressure was 100/60 mm Hg in the right arm, the peripheral pulses were normal, and the lungs were clear. A left ventricular thrust was felt in the sixth intercostal space at the anterior axillary line; the second heart sound was physiologically split with slight accentuation of the pulmonary component. A grade III (on the basis of I to VI), systolic, crescendo-decrescendo murmur was heard maximally at the left lower sternal border, extending to the left upper sternal border and the apex. A grade I diastolic murmur was present at the middle and bottom of the left sternal border and over the apex. Thoracic roentgenograms (fig. 7 left) revealed a left aortic arch, cardiac enlargement (cardiothoracic ratio, 0.74), and an area of irregular calcification in the region of the left ventricle (on cardiac fluoroscopy this was localized to the posterior wall of that chamber). The electrocardiogram (fig. 8 left) showed a posterolateral “silent area” (abnormal Q waves in leads I, II, III, and aV5) as well as a deep Q wave and inverted T wave in lead

Circulation, Volume XXXVI, September 1967
V₆ compatible with an ischemic pattern. There was an associated ventricular excitation aberration (QRS, 0.24 second). Biplane angiocardiograms (fig. 9) revealed a large calcified mass in the posterior aspect of the left ventricular wall; it did not involve the interventricular septal region. The coronary arteries were normal in origin and distribution, and the mass did not appear to receive any significant vascular supply from the coronary system.

In February 1966 the patient was operated on through a median sternotomy. The anterior surface of the heart looked normal. Extracorporeal circulation was instituted at 30 C and, after the heart was lifted out of the pericardium, a tumor more than twice the size of the left ventricle was seen. It had a hard, rubbery consistency and a white surface; it extended from the anterior descending to the posterior descending coronary artery and from beyond the apex of the left ventricle to the coronary sinus (which was stretched across its surface) superiorly. The adjacent myocardium was compressed against the margin of its outer surface, and the endocardium was stretched across its inner surface. The posterior papillary muscle originated from the area of junction of the compressed myocardium with the inner surface of the tumor. The mass was removed through an elliptic incision. Only a narrow rim of muscle was included with the tumor in order not to disengage the papillary muscle. The coronary sinus was entered at one point superiorly and was repaired with continuous arterial silk; the left ventricular cavity was entered at multiple points even up to the attachment of the posterior leaflet of the mitral valve. However, the posterior leaflet and its chordae tendineae remained intact. The ventricular wall was closed with three layers of silk. Cardiac action was maintained throughout the procedure, and no difficulty was encountered in discontinuing the extracorporeal circulation. The patient’s postoperative course was uneventful. On dismissal 10 days later, he had a grade III apical diastolic murmur with presystolic accentuation, and the first heart sound was slightly accentuated, suggesting some mitral stenosis. There were no symptoms to suggest that this was of hemodynamic significance. The thoracic roentgenogram (fig. 7 right) showed decrease in cardiac size (cardiothoracic ratio, 0.60). The electrocardiogram (fig. 8 right) demonstrated a slight change in ventricular excitation pattern and continued to show evidence of damage to the posterolateral part of the left ventricular wall.

Case 2. Thoracic roentgenograms. (Left) Preoperatively, showing cardiomegaly and irregular calcifications in region of left ventricle. (Right) Postoperatively, heart is considerably smaller and no calcification can be seen.
VENTRICULAR FIBROMA

Figure 8

Case 2. Electrocardiograms. (Left) Preoperatively, showing Q waves in I, II, and III, and aVp, and a deep Q wave and inverted T wave in V6 (QRS, 0.24 second). (Right) Postoperatively there is still evidence of damage to posterolateral part of left ventricle.

In the 10 months since the operation, this patient has had no cardiac symptoms and has been very active. A thoracic roentgenogram made 5 months postoperatively was similar to the one made at dismissal; an electrocardiogram showed a QRS axis of -45° in the frontal plane with a QRS interval of 0.08 second. There was return of ventricular depolarization and repolarization toward normal.

The final diagnosis was fibroma of the left ventricle. The firm tumor measured 8.5 by 5 by 4.5 cm and weighed 118 g (fig. 10). It resisted cutting, and multiple areas of calcification were noted along the cut surface. Microscopically, the tumor was composed largely of dense fibrous tissue resembling a desmoid; about the periphery of the tumor, strands of cardiac muscle were entrapped in the proliferating fibrous tissue (fig. 6 right).

Discussion

Tumors of the heart have received increasing attention in the literature in recent years, although they are a relatively infrequent cause of cardiac dysfunction. Prichard, in an excellent and comprehensive review of the subject in 1951, noted that metastatic tumors of the heart occur approximately 20 to 40 times as frequently as do primary ones. He reported that autopsy disclosed a metastatic tumor of the heart in 326 (3.9%) of 8,414 cases in which death was due to cancer. Reports of the incidence of primary cardiac tumors in autopsy experience have given widely divergent observations, varying from none among 30,000 autopsies to three among 1,200 autopsies, an incidence of 0.25%. Lymburner found four cases among 8,550 autopsies at the Mayo Clinic between 1915 and 1931, and another 30 were encountered among 26,000 autopsies between 1932 and 1965 (J. L. Titus, personal communication), bringing the total to 34 cases, an incidence of about 1 per 1,000 autopsies. In addition, 18 patients were diagnosed as having a primary tumor of the heart and were operated on successfully since 1956. The two cases presently reported were the only instances of ventricular fibroma encountered at this institution.

The great majority of primary cardiac neoplasms are benign clinically and histologically. Regardless of type, however, the presence of a neoplasm in the heart frequently results in death due to functional impairment.

Classification

Location and histology have been commonly used as bases for classification.

Pericardial Tumors

Benign pericardial tumors are about 25 to 50% as frequent as benign tumors of the heart per se and, although a hemopericardium is as common in benign as in malignant pericardial tumors, the latter follow a more rapid course and are often associated with pain. Symptoms result from either inflow blockage or pericardial effusion.

Intracavitary Tumors

Endocardial myxomas are the most common type of primary heart tumor. They
Figure 9

Case 2. Anteroposterior (left) and lateral (right) views of left ventricular angiogram, showing large calcified mass in posterior aspect of left ventricular wall impinging on ventricular cavity. Coronary arteries are normal in origin and distribution.

Figure 10

Case 2. Gross appearance of tumor; weight, 118 g. Note similarity to figure 5.

originate almost exclusively in the atria, more than 75% occurring in the left atrium, and may interfere with valvular function.

Mural Tumors

The benign and malignant mesenchymal neoplasms of the heart wall are similar to those of any other striated muscle or connective tissue in variety of tumor types. Sarcomas are most common and usually involve the wall of the right atrium. Dysrhythmia, hemopericardium, and superior vena caval obstruction are the more common complications. These tumors have given rise to distant metastasis in 30% of the cases.

Benign mural tumors are less frequent and include angiomata, lipomas, rhabdomyomas (commonly associated with tuberous sclerosis), and fibromas. The latter constitute about 5% of primary cardiac tumors. To date, we have been able to find 34 published cases of intramural fibroma of the heart (table 1), excluding those occurring in the atrium.

Pathological Findings

Some controversy has arisen regarding the pathogenesis of primary benign cardiac fibromas, leading some pathologists to group them with other lesions and classify them as hamartomas. We prefer to reserve the
term "hamartoma" for nonproliferating embryonic malformations containing muscle, fibrous tissue, nerves, blood vessels, and fat, as in the cases recently reported by Grüber and Samuel,47 Nicks,48 and Dahlgren and Nordenström.49 The fibromas are histologically benign proliferations of fibrous connective tissue and are not encapsulated; grossly, they bear a remarkable resemblance to desmoid tumors (figs. 5 and 10).

Because of this confusion in terms, the pathological designations of the tumor discussed in this report, although mainly fibroma, have also been rhabdomyoma,13 fibrosarcoma,15 mesoblastoma,18 mesenchymoma,25 and hamartoma.16, 21, 24, 31, 33 The pathological appearance of these tumors was very similar to that of the two described in this report, consisting of interlaced collagenous fibers admixed with fibroblasts and occasional entrapped cardiac muscle cells (fig. 6). The tumor designated as rhabdomyoma by Brown and Gray13 contained a large number of cardiac muscle cells, but the basic structure fit the pattern of an intramural fibroma, while the tumor designated as fibroma by Van der Hauwaert and associates50 consisted predominantly of myocardial tissue and fit the pattern of a rhabdomyoma. The tumor considered to be fibrosarcoma by Fidler and co-workers15 contained a large number of spindle cells of uniform size surrounded by less dense fibrous tissue; cardiac muscle cells were found within the tumor at its periphery, and there was no evidence of anaplasia, mitotic figures, giant cells, or metastasis. The tumor resembled closely a benign intramural fibroma and has been designated as such by several authors.4, 22, 29, 32, 34, 38

In their discussion of the pathogenesis of intramural fibroma, Bigelow and associates4 concluded that it probably originates from the primitive cardiac mesenchyme which differentiates into the various elements contained in the tumor, the predominant one being fibrous tissue with abundant collagen and young fibroblasts. Freeman and co-workers34 suggested that this tumor should be in the same category as fibromas in general inasmuch as it has similar histological features and growth characteristics, and Ts'o and Teoh85 described a fibroma of the left ventricle associated with multiple fibromas in other parts of the body in a 2%-day-old boy.

Clinical Data

The ages in the 36 patients with intramural ventricular cardiac fibroma, including the two presently reported on, ranged from 42 hours32 to 65 years14; 31 cases occurred in children (86%). There were 15 females and 21 males.

The pertinent clinical features of the cases are summarized in table 1. Symptoms and signs of heart disease were present in 17 of the 34 patients previously reported on. Approximately 30% of the fibromas have been listed as a cause of sudden death, presumably by involvement of the interventricular conduction apparatus. Edlund and Holmødal’s38 case was the first to be diagnosed preoperatively after an angiocardiogram demonstrated displacement of the left ventricular cavity by a mass in the apex. The patient was operated on, but the mass was not removed and no biopsy was attempted. The patient was alive and well 1 year after the operation. Since then, three other cases35, 37, 38 were diagnosed preoperatively, but only the patient described by Parks and associates51 underwent successful surgical removal of the tumor and was reported to be doing well 27 months after the operation. However, extensive fibrosis and an aneurysm of the left ventricle in the region of the operative site were suspected. Wilson and associates57 removed the lesion completely in a 17-month-old boy, but the patient died 13 hours after the operation. Hoen and Ellis38 found a left ventricular tumor in an 11-year-old boy, which was too extensive for resection, and only a biopsy was done. The child’s condition was unchanged a year and a half after the operation.

In all the other reported cases of intramural fibroma, the correct clinical diagnosis was not made. Coronary artery disease and left ventricular aneurysm were thought to be present in a 51-year-old man31; at operation,
<table>
<thead>
<tr>
<th>Author</th>
<th>Age</th>
<th>Sex</th>
<th>Presenting condition</th>
<th>Fibroma size and location</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Luschka (1855)</td>
<td>6</td>
<td>M</td>
<td>Croup</td>
<td>4.3 x 3.5 cm; upper half of left ventricle</td>
<td>Incidental finding</td>
</tr>
<tr>
<td>Wagstaffe (1870-1871)</td>
<td>3</td>
<td>F</td>
<td>Sudden death</td>
<td>&quot;Hen's egg&quot;; interventricular septum</td>
<td></td>
</tr>
<tr>
<td>Zander (1880)</td>
<td>36</td>
<td>F</td>
<td>Pulmonic systolic murrum; edema; ascites</td>
<td>4.9 x 5.7 x 5.9 cm; interventricular septum &amp; right ventricle</td>
<td>Second fibroma in right atrium; ECG reported</td>
</tr>
<tr>
<td>Mönckeberg (1924)</td>
<td>Newborn</td>
<td>M</td>
<td>Sudden death</td>
<td>&quot;Hazelnut&quot;; apex</td>
<td></td>
</tr>
<tr>
<td>Teuscher (1927)</td>
<td>2½</td>
<td>F</td>
<td>Sudden death</td>
<td>3.2 x 3.0 cm; left ventricle</td>
<td></td>
</tr>
<tr>
<td>Brown and Gray (1930)</td>
<td>3</td>
<td>M</td>
<td>Sudden death</td>
<td>7.7 x 6.5 cm; lateral wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Macherey (1937)</td>
<td>65</td>
<td>M</td>
<td>Aortic stenosis; congestive heart failure</td>
<td>5.5 x 2.5 cm; anterior wall of left ventricle</td>
<td>Calcific aortic stenosis at autopsy</td>
</tr>
<tr>
<td>Fidler and associates (1937)</td>
<td>9</td>
<td>M</td>
<td>Cardiac arrest during operation for harelip</td>
<td>2.2 x 2.6 cm; apex &amp; interventricular septum</td>
<td></td>
</tr>
<tr>
<td>Symeonidis and Linzbach (1938)</td>
<td>15</td>
<td>M</td>
<td>R-sided paralyis (2 mo); diphtheria (2 wk)</td>
<td>&quot;Dove's egg&quot;; posterior wall of left ventricle</td>
<td>Incidental finding</td>
</tr>
<tr>
<td>Vukan (1940)</td>
<td>1</td>
<td>F</td>
<td>Sudden death after convulsion</td>
<td>5 x 7 cm; right ventricle</td>
<td></td>
</tr>
<tr>
<td>Schink (1940)</td>
<td>3</td>
<td>M</td>
<td>Prematurity</td>
<td>&quot;Bean size&quot;; right ventricle</td>
<td></td>
</tr>
<tr>
<td>Kulka (1949)</td>
<td>8</td>
<td>F</td>
<td>History of seizures; aspiration of vomiting</td>
<td>5 x 3.5 cm; anterior wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Bigelow and associates (1954)</td>
<td>3</td>
<td>F</td>
<td>Cardiomegaly; harsh murmur; heart failure</td>
<td>5 x 3 x 2.5 cm; lateral wall of left ventricle</td>
<td>Associated patent ductus arteriosus</td>
</tr>
<tr>
<td>Froboese (1955)</td>
<td>2</td>
<td>F</td>
<td>Shock for 2 days</td>
<td>5.4 x 4.1 cm; anterior wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Naeve (1955)</td>
<td>9</td>
<td>M</td>
<td>Dyspnea; tachycardia; systolic murrum</td>
<td>5.5 x 3.5 cm; lower half of left ventricle</td>
<td>ECG reported</td>
</tr>
<tr>
<td>McCue and associates (1955)</td>
<td>47</td>
<td>F</td>
<td>Subaortic stenosis</td>
<td>5 x 3 cm; left ventricle</td>
<td>ECG reported</td>
</tr>
<tr>
<td>James and Stanfield (1955)</td>
<td>4</td>
<td>F</td>
<td>Tachycardia; ectopic rhythm</td>
<td>6 x 5 x 4 cm; anterior aspect of left ventricle</td>
<td>ECG reported</td>
</tr>
<tr>
<td>Radnai (1956)</td>
<td>5</td>
<td>F</td>
<td>Shigellosis</td>
<td>1.6 x 3 cm; anterior wall of right ventricle</td>
<td></td>
</tr>
<tr>
<td>Conlon (1956)</td>
<td>7</td>
<td>M</td>
<td>Vomiting &amp; diarrhea</td>
<td>5 x 4 x 4 cm; posterolateral wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Edlund and Holmdahl (1957)</td>
<td>19</td>
<td>M</td>
<td>Systolic murrum; cardiac enlargement</td>
<td>&quot;Poorly defined&quot;; ventral surface of left ventricle</td>
<td>Gross diagnosis at surgery at age 32 mo; ECG reported</td>
</tr>
<tr>
<td>Author(s) and Dates</td>
<td>Age</td>
<td>Gender</td>
<td>Diagnosis</td>
<td>Pathology</td>
<td>Additional Information</td>
</tr>
<tr>
<td>--------------------</td>
<td>-----</td>
<td>--------</td>
<td>-----------</td>
<td>-----------</td>
<td>------------------------</td>
</tr>
<tr>
<td>Aurio and co-workers (1958)</td>
<td>43 yr</td>
<td>F</td>
<td>Epithelioma of scalp</td>
<td>6 × 4 cm; anterolateral myocardium</td>
<td>Incidental finding; ECG reported</td>
</tr>
<tr>
<td>Krueger and Knoll (1958)</td>
<td>4 yr</td>
<td>F</td>
<td>Sudden death</td>
<td>7.2 × 6 × 2.5 cm; wall of left ventricle</td>
<td>ECG reported</td>
</tr>
<tr>
<td>Jernstrom and Cremin (1959)</td>
<td>3½ yr</td>
<td>M</td>
<td>Subaortic stenosis</td>
<td>5.5 × 4.5 × 3 cm; anterior surface of left ventricle</td>
<td>Cardiac arrest during surgery; ECG reported</td>
</tr>
<tr>
<td>Valledor and associates (1960)</td>
<td>8 mo</td>
<td>F</td>
<td>Congestive failure; syncope</td>
<td>7-cm diam; interventricular septum</td>
<td>ECG reported; surgical diagnosis; partial excision; died postop. day 3</td>
</tr>
<tr>
<td>Švejda and Tomášek (1960)</td>
<td>51 yr</td>
<td>M</td>
<td>Anginal pain; syncope; picture of left ventricular aneurysm</td>
<td>4 × 5 × 5.2 cm; interventricular septum &amp; anterior wall of left ventricle</td>
<td>ECG reported; preop. diagnosis from angiocardiogram; tumor successfully removed</td>
</tr>
<tr>
<td>Boyette and Foushee (1960)</td>
<td>42 hr</td>
<td>M</td>
<td>Dyspnea; cyanosis</td>
<td>4.5 × 4 cm; interventricular septum</td>
<td>3-mm nodule; posterior wall of left ventricle</td>
</tr>
<tr>
<td>Parks and associates (1962)</td>
<td>26 mo</td>
<td>M</td>
<td>Bronchopneumonia; picture of endocardial fibroelastosis</td>
<td>7.2 × 4.5 × 3.5 cm; posterolateral wall of left ventricle</td>
<td>Associated fibromas of larynx and tongue (solitary) and of lungs (multiple)</td>
</tr>
<tr>
<td>Freeman and associates (1963)</td>
<td>17 mo</td>
<td>F</td>
<td>Sudden death</td>
<td>8 × 8 × 5.5 cm; interventricular septum &amp; anterior wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Ts'o and Teoh (1963)</td>
<td>2½ da</td>
<td>M</td>
<td>Tachypnea; cyanosis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Palásthy and Tóth (1965)</td>
<td>7 da</td>
<td>M</td>
<td>Tachypnea; cyanosis; cardiomegaly; harsh murmur</td>
<td>4.5 × 3 cm; posterior wall of left ventricle &amp; posterior aspect of interventricular septum</td>
<td>Preop. diagnosis from angiocardiograms; tumor removed; died postop. day 1; ECG reported</td>
</tr>
<tr>
<td>Wilson and associates (1965)</td>
<td>17 mo</td>
<td>M</td>
<td>Pneumonia; cardiomegaly</td>
<td>“Large”; wall of left ventricle</td>
<td>Preop. diagnosis from angiocardiograms; ECG reported; at surgery tumor too extensive for resection; biopsied only; doing well 1½ year later</td>
</tr>
<tr>
<td></td>
<td>18 mo</td>
<td>M</td>
<td>Ebstein's anomaly; sudden death</td>
<td>“Large”; right ventricle</td>
<td>Preop. diagnosis; successfully removed</td>
</tr>
<tr>
<td>Hoen and Ellis (1966)</td>
<td>11 yr</td>
<td>M</td>
<td>Chest pain; cardiomegaly</td>
<td>6 × 4 cm; lateral wall of left ventricle</td>
<td></td>
</tr>
<tr>
<td>Geha and associates (present report)</td>
<td>27 mo</td>
<td>M</td>
<td>Syncopal attacks; arrhythmia</td>
<td>6-cm diam; interventricular septum</td>
<td>Preop. diagnosis from angiocardiograms; successfully removed</td>
</tr>
<tr>
<td></td>
<td>38 mo</td>
<td>M</td>
<td>Cardiomegaly; murmur</td>
<td>8.5 × 5 × 4.5 cm; posterolateral wall of left ventricle</td>
<td></td>
</tr>
</tbody>
</table>
a left ventricular intramural fibroma was found and partially removed, but the patient died on the third postoperative day. Endocardial fibroelastosis or anomalous coronary artery circulation was suspected in four cases, possibly because of diminution of effective ventricular ejection and electrocardiographic evidence of abnormal ventricular excitation. Intramural fibroma has been confused with hypertrophic subaortic stenosis because of narrowing of the left ventricular outflow tract. It may also mimic pulmonary stenosis. In a 36-year-old woman, pulmonary systolic murmur, edema, and ascites developed 6 months before her death; autopsy revealed a fibroma arising from the interventricular septum, which had almost obliterated the right ventricular outflow tract.

Most intramural fibromas are located in the interventricular septum or left ventricular wall, only four being recorded as only in the right ventricular wall. Congenital heart disease in association with this tumor has been reported only once, in a boy who had a large patent ductus arteriosus and who died 3 days after birth. No familial incidence of intramural fibroma has been reported.

Laboratory Data

Thoracic Roentgenogram

In most of the published cases, the roentgenogram showed generalized cardiac enlargement. Calcification within the intramural fibroma was described in only four cases. However, calcification is not uncommon in these tumors and, because the presence of calcium in the heart wall is often the one finding pointing toward the diagnosis, care should be taken to look for calcium in all cases of heart disease.

Angiocardiography

This was performed in five cases of intramural fibroma. In four of them a cardiac tumor was diagnosed and, in the fifth, a filling defect in the left ventricular cavity was seen in retrospect. Angiocardiography confirmed the diagnosis and outlined the extent of the tumor in both of our cases (figs. 3 and 9). This is the most valuable method of making the diagnosis and should be used whenever a cardiac tumor is suspected.

Electrocardiography

These results were reported in 13 of the 34 cases of intramural fibroma, and various abnormalities have been noted. Ectopic rhythms were found in three cases. Incomplete bundle-branch block with biventricular hypertrophy was present in one, right bundle-branch block in two, and an unidentified intraventricular conduction defect in two others. Left ventricular hypertrophy with strain was reported in five instances. In four cases there were nonspecific S-T segment and T-wave changes. In one case the tumor produced the pattern of an old myocardial infarction.

Surgical Treatment

Successful treatment of cardiac tumors depends on extirpative surgery. The excision of each type of tumor presents specific problems. In any case, enough time must be available to allow adequate exposure and careful dissection, thus requiring the use of a pump-oxygenator in the majority of instances.

With mural tumors, every effort should be made to preserve the atroventricular valves, papillary muscles, and functional myocardium. Parks and associates pointed out that the benign intramural tumors seem to enlarge by displacing the normal ventricular muscle and, hence, one is left with sufficient residual myocardium even after excision of a large tumor (as was the case in our two patients) to permit reconstruction of an adequate ventricle.

None of the three patients who have survived excision of a cardiac fibroma has been known to have a recurrence, but this experience is very limited. The possibility of recurrence is a definite concern because fibromas generally have this propensity and because the margin of myocardium excised with these tumors has necessarily been limited.
VENTRICULAR FIBROMA

Concluding Comment

A review of these two cases and the literature on cardiac fibroma indicates that a primary mural tumor of the heart should be suspected in patients, particularly in children, with unexplained cardiac failure, unexplained cardiac dysrhythmia, intracardiac calcifications, irregular shadows on roentgenograms, or unexplained newly developing cardiac symptoms and murmurs. Angiography is the best way of confirming the diagnosis. In view of the potential hazard of these tumors, their surgical removal should be undertaken, and this experience indicates that even very large and extensive intramural ventricular tumors may be removed successfully.

References


Intramural Ventricular Cardiac Fibroma: Successful Removal in Two Cases and Review of the Literature

ALEXANDER S. GEHA, WILLIAM H. WEIDMAN, EDWARD H. SOULE and DWIGHT C. MCGOON

Circulation. 1967;36:427-440
doi: 10.1161/01.CIR.36.3.427

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/36/3/427