Intracardiac Myxomas with Report of Two Unusual Cases and Successful Removal

By John T. Differding, M.D., Richard E. Gardner, M.D., and Benson B. Roe, M.D.

Until 1951 no intracardiac myxomas had been diagnosed before death. Since then at least 43 cases have been diagnosed during life; 27 of these were discovered during closed operations for suspected mitral or tricuspid stenosis. From March 1955, when Crafoord first reported the successful removal of an intracardiac myxoma, to December 1960 at least 43 successful surgical removals have been reported. This report adds two additional cases, both with unusual features.

In the first case the patient underwent successful removal of a myxoma by a closed method following accidental dislodgment of the tumor during exploration of the mitral valve. This is the second reported complete removal of such a tumor without the aid of extracorporeal circulation or inflow occlusion.

The second case is that of a 12-year-old boy who had a femoral artery embolus that on microscopic examination proved to be myxoma. The intracardiac tumor was successfully removed. This is the second patient successfully operated upon in whom a tissue diagnosis was established preoperatively. He is also the second youngest patient to survive removal of the tumor.

Case Reports

Case 1

A 55-year-old white woman had a 1-year history of repeated respiratory infections and progressive dyspnea. She had been treated for one episode of acute pulmonary edema and had responded to digitalization and diuretics.

Physical examination revealed slight cardiomegaly, normal sinus rhythm at 84, and blood pressure of 130/80. The mitral first sound was slightly accentuated, but the pulmonic second sound was normal. A grade-I diastolic murmur was heard when the patient was on her right side. No opening snap was heard. The lungs were clear.

Right-sided heart catheterization revealed a pulmonary artery wedge pressure of 29 mm. Hg and a pulmonary artery pressure of 64/30. The calculated mitral valve area was 1.2 cm².

A left posterolateral thoracotomy was performed on February 18, 1960, after a preoperative diagnosis of mitral stenosis had been made. When a finger was introduced through the left atrial appendage a firm, smooth, egg-sized tumor was encountered. The mitral valve was normal in structure and function except for partial occlusion by the mass. It was planned to withdraw and approach this lesion under direct vision with extracorporeal circulation. When the tumor was palpated to determine the size and location of its attachment, however, it was inadvertently dislodged; immediate removal became mandatory because of the ball-valve action that totally obstructed the circulation when the operator's finger did not protect the valve.

The left-sided operative approach and the need to maintain a finger in the atrium made it impossible to prepare for inflow occlusion. Despite the hazard created by the tumor's size and the inevitability of a significant atrial tear, the mass was forcibly extruded through the atriotomy. Massive bleeding occurred followed by ventricular fibrillation. The rent was rapidly sutured closed, the blood was replaced, and the circulation was restored by manual systole. Defibrillation was successful after 30 minutes. The patient had an uneventful recovery without evidence of neurologic or renal damage, and her symptoms have been relieved.

From the Departments of Pathology and Surgery of the University of California School of Medicine, San Francisco, California.
Pathologic examination of the tumor showed a white and light yellow mass, 5 by 4 by 3 cm., which weighed 38 Gm. (fig. 1). A 2 by 2 cm. roughened area was present at one end where the tumor was attached to the atrial wall. The rest of the surface was smooth and glistening.

On microscopic study the central part of the tumor was composed of finely vaculated eosinophilic material that contained numerous vascular channels and tiny islands of spindle-shaped and multinucleated cells. No mitoses were seen. The walls of the vascular channels consisted of spindle cells, frequently several cells thick, which were continuous with endothelial cells lining the lumen. The spindle cells were separated by vaculated pale basophilic material (figs. 2 and 3). The stroma contained a moderate number of lymphocytes and plasma cells, lesser amounts of neutrophils and macrophages, numerous foci of calcification, some recent hemorrhage, and small clumps of golden brown pigment. A thin rim of dense eosinophilic material on the surface of the tumor contained a few flattened endothelial cells and overlay a thicker layer of vacuolated light basophilic material.

Case 2

A 12-year-old boy was hospitalized in November 1959 following an episode of coma and shock, but he recovered and was discharged from the hospital without a specific diagnosis. In March 1960 he developed sudden severe cramping pains and pallor of both legs. Embolectomies were performed on the left popliteal and right common femoral arteries. The removed tissue was diagnosed as myxoma.

The patient was transferred to the University of California Medical Center with a diagnosis of intracardiac myxoma. On physical examination his blood pressure was 120/80 mm. Hg. Ventricular extrasystoles were fairly frequent, and a variable systolic and a short diastolic murmur were heard along the left sternal edge with splitting of the first and second sounds.

On March 25, 1960, a cardiotomy was performed through a right anterolateral thoracotomy with use of extracorporeal circulation. Before an incision was made in the left atrium, ventricular fibrillation was intentionally induced to prevent further embolization. When the atrium was opened, a villous gelatinous tumor, which apparently occupied nearly the entire atrium, immediately bulged through the incision. The tumor was extremely friable, had a jelly-like consistency, and fell apart as it was removed from its origin near the closed fossa ovalis. After closure of the atrium, the heart was initially defibrillated with ease, but reverted to ventricular fibrillation that resisted defibrillation. The electrocardiogram showed an ischemic pattern. The patient's condition gradually improved and regular rhythm occurred spontaneously after 76 minutes.

The postoperative electrocardiogram showed changes indicating myocardial injury. The patient recovered completely except for persistent electrocardiographic evidence of myocardial damage.

The emboli consisted of two white, partially translucent masses that measured 17 by 10 by 10 mm. and 14 by 11 by 7 mm. and contained small...
INTRACARDIAC MYXOMAS

Figure 3
Vascular channels showing the thick wall of vacuolated material and stromal cells which are continuous with the endothelial cells lining the lumen. There are neutrophils and erythrocytes in the lumina, and plasma cells and lymphocytes in the stroma. Hematoxylin-eosin stain.

tan and brown areas. The specimen removed from the heart was composed of multiple pieces of lobulated, smooth, glistening, light yellow, partially translucent tissue that weighed 19.2 Gm. (fig. 4).

Microscopic sections showed masses of vacuolated light basophilic or, occasionally, light eosinophilic material whose papillary external surfaces were partially covered by numerous round and flattened cells (figs. 5 and 6). Within this loose stroma were scattered spindle-shaped, round and stellate cells that had round or oval nuclei and a small amount of eosinophilic cytoplasm (fig. 7). Many of the cells were multinucleated. No mitoses were seen. Also within the stroma were large areas of hemorrhage, masses of homogeneous eosinophilic material, and a few clumps of golden brown pigment. The few vascular channels in the stroma were lined by plump endothelial cells, occasionally several layers thick (fig. 8). The embolus contained small focal collections of neutrophils and the cardiac specimen contained small numbers of plasma cells and lymphocytes.

Incidence

Myxoma, the most frequent primary tumor of the heart, is a benign growth constituting nearly 50 per cent of all primary cardiac tumors. In 1951, 128 myxomas were reviewed by Prichard, and, in 1959, 90 additional cases were reviewed by Padhi and associates. If Mahaim's case of a 3-month-old infant, who probably did not have a myxoma, is excluded, the youngest reported cases are in

3-year-old children. McAllen reported two patients, aged 72 and 73, and stated that Curtis had reported the oldest case, that of an 83-year-old. The sex distribution has been reported both as being equal in men and women and as being nearly three times more common in women.

Etiology

The impression that myxomas are really thrombi seems to have arisen in several ways: (1) the failure to establish any criteria for myxoma or to distinguish between a myxoma and an obvious polypoid thrombus; (2) the rarity of the tumors and the inability of any one observer to amass a large experience with them; and (3) the presence in many myxomas of plump fibroblasts, hemosiderin, and large numbers of capillaries. The last point deserves explanation. Myxoma is a soft tumor subjected to considerable mechanical stress; hemorrhages and hemosiderin deposition may be expected to occur from capillaries in such a structure. Likoff and associates, in 1954, reported a pedunculated mass that arose near the foramen ovale and microscopically resembled a myxoma, but which he considered to be an organized thrombus.
### Table 1

**Successfully Removed Intracardiac Mymomas**

<table>
<thead>
<tr>
<th>Reported by</th>
<th>Year reported and date of operation</th>
<th>Age and sex</th>
<th>Chamber</th>
<th>Tumor size</th>
<th>Surgical method</th>
<th>How diagnosed and known time of survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Crafoord</td>
<td>1955</td>
<td>45</td>
<td>Left</td>
<td>?</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 1 year later</td>
</tr>
<tr>
<td>2. Bigelow</td>
<td>1955</td>
<td>50</td>
<td>Left</td>
<td>8 × 5 × 3½ cm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; well 3 years later</td>
</tr>
<tr>
<td>3. Scannell</td>
<td>1956</td>
<td>33</td>
<td>Left</td>
<td>8½ × 3½ cm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; well 5 months later</td>
</tr>
<tr>
<td>4. Bahnson</td>
<td>1957</td>
<td>57</td>
<td>Left</td>
<td>6 × 5 × 4 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 6 months later</td>
</tr>
<tr>
<td>5. Robertson</td>
<td>1957</td>
<td>38</td>
<td>Left</td>
<td>6 × 6 × 6 cm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; well 3 months later</td>
</tr>
<tr>
<td>6. Chin &amp; Ross</td>
<td>12/15/56</td>
<td>25</td>
<td>Left</td>
<td>8 × 7 cm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; survived operation</td>
</tr>
<tr>
<td>7. Hanlon</td>
<td>1957</td>
<td>61</td>
<td>Right</td>
<td>?</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 2 weeks later</td>
</tr>
<tr>
<td>8. Marions</td>
<td>1957</td>
<td>?</td>
<td>Left</td>
<td>4 × 4 × 4 cm.</td>
<td>?</td>
<td>Angiocardiography; survived operation</td>
</tr>
<tr>
<td>9. Fatti</td>
<td>11/7/55</td>
<td>43</td>
<td>Left</td>
<td>6 × 5 × 4 cm.</td>
<td>Closed</td>
<td>Operation for mitral stenosis; well 18 months later</td>
</tr>
<tr>
<td>10. Contes</td>
<td>1958</td>
<td>50</td>
<td>Right</td>
<td>?</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 6 months later</td>
</tr>
<tr>
<td>11. Gerbode</td>
<td>1958</td>
<td>51</td>
<td>Left</td>
<td>42 Gm.</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; survived operation</td>
</tr>
<tr>
<td>12. Becher</td>
<td>6/5/57</td>
<td>52</td>
<td>Left</td>
<td>?</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; survived operation</td>
</tr>
<tr>
<td>13. Krelikova</td>
<td>7/19/57</td>
<td>14</td>
<td>Right</td>
<td>5½ × 5 × 4 cm.</td>
<td>Hypothermia</td>
<td>Fluoroscopy; well 4 months later</td>
</tr>
<tr>
<td>14. Krelikova</td>
<td>3/4/58</td>
<td>51</td>
<td>Left</td>
<td>8 × 5 × 2½ cm.</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; survived operation</td>
</tr>
<tr>
<td>15. Ellis</td>
<td>1958</td>
<td>45</td>
<td>Left</td>
<td>5 × 4 × 4 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 5 months later</td>
</tr>
<tr>
<td>16. Ellis</td>
<td>1958</td>
<td>48</td>
<td>Right</td>
<td>10 × 10 × 10 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for tricuspid stenosis; well 4½ months later</td>
</tr>
<tr>
<td>17. Malm</td>
<td>1958</td>
<td>?</td>
<td>Left</td>
<td>8 × 6 × 5 cm.</td>
<td>Extracorporeal circulation</td>
<td>Not reported</td>
</tr>
<tr>
<td>No.</td>
<td>Year</td>
<td>Sex</td>
<td>Side</td>
<td>Size</td>
<td>Procedure</td>
<td>Notes</td>
</tr>
<tr>
<td>-----</td>
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</tr>
<tr>
<td>19.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>5 x 5 x 5 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 1 year later</td>
</tr>
<tr>
<td>20.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>†</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 18 days later</td>
</tr>
<tr>
<td>21.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>158 Gm.</td>
<td>Hypothermia</td>
<td>Operation for mitral insufficiency; survived operation</td>
</tr>
<tr>
<td>22.</td>
<td>1959</td>
<td>M</td>
<td>Right</td>
<td>†</td>
<td>Extracorporeal circulation</td>
<td>Cardiac catheterization; well 6 months later</td>
</tr>
<tr>
<td>23.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>7 x 6 x 3 1/2 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 6 months later</td>
</tr>
<tr>
<td>24.</td>
<td>1959</td>
<td>M</td>
<td>Right</td>
<td>9 x 5 cm.</td>
<td>Hypothermia</td>
<td>Angiocardiography; well 6 weeks later</td>
</tr>
<tr>
<td>25.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>100 Gm.</td>
<td>Extracorporeal circulation</td>
<td>Cardiac exploration for emboli; well 2 months later</td>
</tr>
<tr>
<td>26.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>6 x 3 x 3 cm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; well 4 1/2 months later</td>
</tr>
<tr>
<td>27.</td>
<td>1959</td>
<td>M</td>
<td>Left</td>
<td>†</td>
<td>Hypothermia</td>
<td>Angiocardiography; survived operation</td>
</tr>
<tr>
<td>28.</td>
<td>1960</td>
<td>F</td>
<td>Right</td>
<td>7 x 6 cm.</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 8 weeks later</td>
</tr>
<tr>
<td>29.</td>
<td>1960</td>
<td>F</td>
<td>Left</td>
<td>28 Gm.</td>
<td>Hypothermia</td>
<td>Operation for mitral stenosis; survived operation</td>
</tr>
<tr>
<td>30.</td>
<td>1960</td>
<td>F</td>
<td>Right</td>
<td>6 x 6 x 6 cm.</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 1 1/2 months later</td>
</tr>
<tr>
<td>31.</td>
<td>1960</td>
<td>F</td>
<td>Left</td>
<td>7 x 4 1/2 x 2 1/2 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 3 weeks later</td>
</tr>
<tr>
<td>32.</td>
<td>1960</td>
<td>F</td>
<td>Right</td>
<td>10 x 8 x 4 cm.</td>
<td>Extracorporeal circulation</td>
<td>Operation for tricuspid obstruction; well 6 months later</td>
</tr>
<tr>
<td>33.</td>
<td>1960</td>
<td>F</td>
<td>Left</td>
<td>†</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; well 6 months later</td>
</tr>
<tr>
<td>34.</td>
<td>1960</td>
<td>F</td>
<td>Left</td>
<td>3 x 3 x 2 cm.</td>
<td>Hypothermia</td>
<td>Embolectomy; well 8 months later</td>
</tr>
<tr>
<td>35.</td>
<td>1960</td>
<td>M</td>
<td>Left</td>
<td>7 1/2 x 5 cm.</td>
<td>Extracorporeal circulation</td>
<td>Angiocardiography; well 3 weeks later</td>
</tr>
<tr>
<td>36.</td>
<td>1960</td>
<td>F</td>
<td>Left</td>
<td>†</td>
<td>Extracorporeal circulation</td>
<td>Operation for mitral stenosis; survived operation</td>
</tr>
<tr>
<td>37.</td>
<td>1960</td>
<td>F</td>
<td>Right</td>
<td>5 x 4 x 3 cm.</td>
<td>Closed</td>
<td>Operation for mitral stenosis; well 3 1/2 months later</td>
</tr>
<tr>
<td>38.</td>
<td>1960</td>
<td>M</td>
<td>atrium</td>
<td>19.2 Gm.</td>
<td>Extracorporeal circulation</td>
<td>Embolectomy; well 2 months later</td>
</tr>
</tbody>
</table>

Percentage of females, 69 per cent; males, 31 per cent. Average age: females, 45 years; males, 35 years.

Percentage in left atrium, 71 per cent.

Seven additional cases have been reported.81, 84-87
There are many sound reasons for considering the myxomas as true neoplasms. Most tumors are much larger than thrombi and are far less cellular. They do not retract in the fashion of thrombi elsewhere. They occur for the most part in hearts free from other disease. There is usually no evidence of damage to account for thrombosis. Almost all myxomas are found in the atria, especially the left, while thrombi occur with approximately equal frequency in the atria and ventricles. Most atrial thrombi are found in the atrial appendages but no myxomas have been found in this location. There are no convincing intermediate stages between a frank thrombus and a myxoma, even in the pedunculated and ball-valve thrombi found in the atria. There is no evidence of stratification in myxomas, although almost all thrombi are stratified. The position of elastic fibers along the peripheral free margin of these tumors is not observed in thrombi. The presence of acid mucopolysaccharide within either thrombi or myxomas is not specific but indicates a particular type of mesenchymal tissue. Tissue cultures of myxomas grow both endothelial cells and fibroblasts.

Ribbert considered that the tumors arose from remnants of the myxoid tissue that compose the embryonic endocardium. In his experience the frequent persistence of such tissue around the fossa ovalis furnished an explanation for the peculiar localization of these tumors. It was apparent that there was more connective tissue within the interatrial septum than in any other part of the heart. More of this was in the flat left portion of the septum than in the indented right side. In a few cases small irregular areas of loose fibrillar connective tissue were present between the flaps of the foramen. In four cases the endocardium showed minute areas in which there were lacunae of capillary size lined by plump endothelial cells. Orr suggested that the neoplastic part of the myxoma is the endothelial element and that the myxoid stroma is a degenerative phenomenon.

The mechanical stresses to which a soft tumor in a cardiac chamber is subjected are probably responsible for the tendency of these tumors to be myxoid. In two cases reported by Prichard the myxoid areas were most prominent at the base and edges of the tumor, where motion is greatest. Although myxomas have been named for this myxoid tendency, the evidence indicates that it is of minor importance. Myxomas frequently contain areas in which the solid groups and cords of spindle-shaped and multinucleated cells in the stroma are continuous with similar cells lining vascular channels and with endothelial cells over the surface of the tumor (figs. 3, 6, and 8). Raeburn regarded

Figure 5
Characteristic myxoid stroma containing stellate and multinucleated cells. Plump endothelial cells cover the surface. Hematoxylin-eosin stain.

Figure 6
Surface endothelial cells continuous with stellate and multinucleated cells in an abundant myxoid matrix. Hematoxylin-eosin stain.
the stroma cells as derivatives of fibroblasts and as precursors of endothelial cells, and stated that the tumors are essentially angiomaticous.

These tumors should be distinguished from the so-called myxomas of the valves of the heart. Small collections of myxoid tissue are occasionally found in cardiac valves, especially in infants. These probably represent persistence of the embryonic stage when the valves were composed entirely of myxoid tissue. Such focal collections of myxomatous or loose connective tissue, in some instances referred to as LambI's excrecences, were believed by Craig\textsuperscript{15} to represent a form of endocardial fibroelastosis. Raeburn\textsuperscript{16} reviewed these lesions and considered them fibroelastic hamartomas. Some lesions consist of villous processes covered by endothelium and arise from the valve cusps, generally the ventricular surface of the aortic and pulmonary valves and the atrial surface of the mitral and tricuspid valves.\textsuperscript{16} The stroma is composed of collagen fibers and is surrounded by looser connective tissue that often appears myxomatous. These lesions may be distinguished from atrial myxomas by their location, small size (up to 1.5 cm.), lack of clinical significance, characteristic papillary structure, and often by their lack of vessels.\textsuperscript{17} In addition these lesions fail to exhibit reactions for acid mucopolysaccharide, reactions commonly found in myxoma.\textsuperscript{10} In the myxoma of a heart valve in a stillborn child reported by Reddy and co-workers,\textsuperscript{18} the lesion was "the size of a pepper seed." The pseudomyxoma of the pulmonary valve in a newborn reported by Helwig\textsuperscript{19} "showed a loose edematous stroma with very many young connective tissue and myxomatous cells buried in it."

A case reported by Fayen and Baglio,\textsuperscript{20} which might be included in this group, was that of a patient with a polypoid mass, measuring 6 by 4 by 4 mm., found on the posterior papillary muscle of the left ventricle. The lesion was unusually cellular and similar cells were present in the adjacent myocardium. Abundant myxoid stroma, multinucleated cells, and plasma cells were absent. Another case, which probably does not belong in the myxoma group, was a 3-month-old girl reported by Mahaim.\textsuperscript{4} A microscopic focus of myxomatous tissue was found in the membranous septum partially interrupting the bundle of His. Its microscopic size, location, intramural position, and lack of vascularity are not typical of myxomas.

**Gross Pathology**

Myxomas are found almost exclusively in the atria; roughly 75 per cent occur in the left atrium. Their localization is even more specific in that almost all of them attach to, or overlie, the fossa ovalis or its rim. The tumors are generally solitary but cases of multicentric myxoma have been reported.\textsuperscript{21-25} In one case,\textsuperscript{25} multiple myxomatous nodules lay within the wall of the pulmonary artery. Three cases of myxomas arising in the left ventricle have been reported.\textsuperscript{20, 24, 25} Most myxomas nearly filled the chamber and produced some degree of obstruction. Some of the atrial tumors extended into the ventricles. One myxoma was microscopic in size,\textsuperscript{4} and a few have exceeded 10 cm. The tumors were entirely intracavitary except in one instance.\textsuperscript{4} They are usually polypoid (80 per cent), are frequently pedunculated, and are externally glistening, smooth or gently lobular, often with polypoid projections. Occasionally they are villous. Their consistency varies with

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*Figure 7*

Spindle-shaped, stellate, and multinucleated cells in a myxoid matrix. Hematoxylin-eosin stain.
their collagen content, which has been variously described as gelatinous, soft, rubbery, firm, or resilient. The tumors are white, yellowish-white, yellow, or light brown and frequently display areas of hemorrhage. Thrombi sometimes overlie the surface of the tumor. The external surface may be indented where the growth has prolapsed through a valve orifice. The weight of the heart is usually normal or slightly increased, although several tumor-bearing hearts have weighed 700 to 800 Gm.

**Microscopic Pathology**

The average tumor is described as being poorly cellular, covered by endothelium, and composed of loose tissue of varying vascularity. Stellate, spindle-shaped, and multinucleated cells are scattered throughout. Mitoses are not seen. Most of the blood vessels are of capillary size, and their lining cells are plump and often multilayered. Larger vessels with thick walls may be present and are occasionally thrombosed. Hemorrhages into the stroma are common and hemosiderin is found in varying amounts, both free and in macrophages. Lymphocytes and plasma cells are almost always present, with occasional focal collections. Mucin stains are often positive in all or part of the matrix. The matrix is vividly colored by the Alcian blue and the Rinehart-Abul-Haj stains. The Hale colloidal iron stain is also positive. These reactions, which indicate the acid mucopolysaccharide nature of the matrix, are best visualized in sections prepared from alcohol-fixed tissue. They are usually negative in Zenker-fixed tissue, and only faintly positive in formalin-fixed tissue. Calcification and bone formation are occasionally found in these tumors. Collagen deposition is variable. Stains for elastic tissue frequently reveal large numbers of elastic fibers focally or throughout the tumor. The elastic-fiber network of the endocardium has generally been described as intact, but in four cases reported by Raeburn and one by Fisher and Hellstrom, definite elastic laminae were demonstrated just beneath the surface of the tumor that were continuous with the atrial elastic fibers.

No evidence has been presented indicating that a significant number of these tumor-bearing hearts have been injured by a recognized process other than the primary one.

**Symptoms**

Symptoms result from cavity obliteration, occlusion of the valve orifice, or arterial embolism. Obstruction of blood flow in the right or left atrium may cause dyspnea, cough, retrosternal pain, weakness, palpitation, arrhythmia, hemoptysis, cardiomegaly, or congestive heart failure.

In many cases no features could be elicited that would distinguish mitral stenosis caused by rheumatic heart disease from these intracardiac tumors. The intermittent valvular obstruction occasioned by the prolapse of the pedunculated and mobile polyp may be manifested by intermittent attacks of syncope, as episodes of paroxysmal dyspnea or by arrhythmias, dyspnea, pain, syncope, or changing heart murmurs associated with positional change.

Recurrent emboli, abdominal pain, fever, and evidence of peripheral emboli may suggest subacute bacterial endocarditis. In many cases a significant elevation of the sedimentation rate occurs. Raynaud's phenomenon has been attributed to myxoma.
intracardiac myxomas

At autopsy bits of tumor emboli are often demonstrated in the heart, kidney, and spleen. Emboli have also been reported in the brain, lungs, aorta, renal arteries, central retinal artery, and the extremities. Sudden death from pulmonary embolism caused by a myxoma of the right atrium has been reported.

**Diagnosis**

By 1959 43 reports of confirmed diagnosis of myxoma during life were published, and in 27 of the 43 the tumor was encountered during surgical exploration. If myxomas are kept in mind when these symptoms are present, a positive diagnosis can often be made. Refractoriness of the congestive failure may suggest the diagnosis. A history of rheumatic fever is usually not present with myxomas simulating mitral stenosis but patients with myxoma and a history of rheumatic fever have been reported. Myxoma and mitral stenosis have been found in the same patient. An opening snap is usually absent but may occur. Phonocardiography will confirm its absence or presence. Mitral insufficiency has been diagnosed on the basis of right-sided and left-sided heart catheterization. A feature that may make the diagnosis possible is alteration of the symptoms when the patient changes his position. There may be roentgenographic evidence of interstitial pulmonary fibrosis or calcium within the tumor. Murmurs may rapidly appear or disappear, or change in quality or intensity. In cases simulating subacute bacterial endocarditis, splenomegaly, petechiae, and positive blood cultures will not be present. Microscopic examination of tumor emboli will often permit a diagnosis. Embolization in rheumatic heart disease is unusual without atrial fibrillation. Isolated tricuspid stenosis is rare in rheumatic heart disease and should suggest right atrial tumor. Right atrial myxomas may be confused with Ebstein's malformation of the tricuspid valve or constrictive pericarditis.

Once an intracardiac tumor is suspected, angiocardiography may confirm the diagnosis. Some tumors, however, are not demonstrated by this procedure. Myxomas must be differentiated from thrombi. Myxomas usually are not attached to the posterior wall of the left atrium and produce no filling defects in that region. The attachment of the myxoma near the fossa ovalis may be seen. Left atrial thrombosis is suggested by finding irregular margination of the junction of the left atrial appendage with the left atrium and sparse or no visualization of the left appendage. Van Buchem, using fluoroscopy, thought that a strong pulsatory movement of the esophagus at the level of the left atrium, synchronous with the left ventricular action, was of diagnostic significance.

**Treatment**

In 1942 Beck reported the first successful removal of a cardiac tumor. Other tumors that presented on the external surface of the heart subsequently have been partially or completely excised.

Attempts were made to remove cardiac tumors with the aid of hypothermia. These maneuvers were generally unsuccessful because the limitations of time, exposure, or ventricular fibrillation did not permit removal of the mass. In 1955 Bigelow successfully removed a left atrial myxoma using hypothermia. At least 12 additional cases in which hypothermia was used were managed successfully.

In 1954 the development of the pump-oxygenator led Clowes and associates to remove a left atrial tumor, but the patient died 6 hours postoperatively. In March 1955, Crafoord described the successful removal of a left atrial myxoma. In April 1956, Bahnson et al. reported that they had successfully removed a left atrial myxoma with the use of a pump-oxygenator system. Eleven reports have been published of successful removal of right atrial myxomas and one of a left ventricular myxoma.
In April 1955, Nichols\textsuperscript{64} became the first surgeon to remove an atrial myxoma using a closed method. He inserted a snare, with a net attached to it, through a purse-string atrial appendage. Because of the size of the tumor it could not be entirely removed during the first attempt, and the instrument broke before complete removal could be performed. In 1958 Fatti\textsuperscript{83} reported successful removal of a myxoma by a closed approach. He placed the index finger of one hand into a purse-string opening in the anterior wall of the left ventricle and the index finger of the opposite hand through a similar opening in the left atrium. He was able to deliver the tumor through the atrial opening.

Intracavitary myxomas should be treated by surgical removal, which can be carried out safely only with the aid of inflow occlusion. Closed methods are dangerous, since many of these tumors are very friable. In addition, blood loss is often considerable if the opening is large enough to permit delivery of the tumor, and it is not always possible to remove the tumor completely. It is important that the atrium and ventricle be searched carefully before normal circulation is resumed. Adequate time for this is assured by the use of extracorporeal circulation. When a myxoma is encountered during a proposed operation upon the mitral or tricuspid valve, it is seldom justified to attempt to remove it by a closed method. When circumstances make it necessary to do so, the hazards of embolization and exsanguination can be reduced by temporary, induced ventricular fibrillation to arrest the circulation.

**Discussion**

Both of the cases presented in this paper are good examples of how easily fragments of the tumor, or the entire tumor, may become detached, and why an open procedure is recommended. Fortunately the tumor was of a firm consistency in our first case so that embolization did not occur during its manipulation. Many other attempted removals without the aid of cardiac bypass or hypothermia have been unsuccessful and our first case is only the second successful complete removal by a closed method. Had it not been for the development of ventricular fibrillation, which arrested the circulation, it is probable that the massive hemorrhage could not have been controlled.

In our second case, the patient was the third child to have an intracardiac myxoma removed successfully, and the second successful case to be diagnosed by recovery of a peripheral embolus. Previously an 8-year-old girl had a myxoma successfully removed following recovery of a myxoma embolus.\textsuperscript{44} These cases demonstrate the importance of sectioning emboli routinely. We believe that the deliberately induced ventricular fibrillation prevented serious embolization of the huge, extremely friable, soft mass which was very difficult to evacuate from the atrium.

**Summary**

A comprehensive review of the various aspects of intracardiac myxomas is presented. Two unusual cases are illustrative of many of the problems encountered. Of particular interest are the successful surgical removals of the myxomas in both cases.

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INTRACARDIAC MYXOMAS


Every author contrives, I believe, to persuade himself that the work which consumes his own midnight oil, is precisely the one that is wanted. It is for the reader to determine whether I labour under the delusion common to my brethren.—J. Hope, M.D. Diseases of the Heart and Great Vessels. London, William Kidd, 1832, p. 10.
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