Atrial Myxomas: Special Emphasis on Unusual Manifestations*

Panagiotis N. Symbas, M.D., F.C.C.P.;** Osler A. Abbott, M.D., F.C.C.P.;† William D. Logan, M.D., F.C.C.P.;** and Charles R. Hatcher, Jr., M.D., F.C.C.P.*

From January 1959 to January 1969, eight patients with atrial myxoma were seen at Emory University and Grady Memorial hospitals. Five patients had symptoms and signs compatible with mitral stenosis, three of whom underwent surgery with an incorrect preoperative diagnosis of mitral stenosis. Two myxomas were found at autopsy, one in a patient who died of acute myocardial infarction. One patient had findings suggestive of mitral stenosis and symptoms of subacute bacterial endocarditis. One patient was seen for recurrent dyspnea and a calcified right atrial mass and one for dyspnea and unexplained cyanosis. Myxoma of the heart is more common than generally realized and its clinical manifestations vary and can mimic other clinical entities. Therefore, a high index of suspicion concerning their diagnosis is needed. Early diagnosis should be established utilizing angiocardiology and immediate resection should be performed before catastrophic complications occur. The results of the surgical treatment of atrial myxoma are gratifying and long lasting.

Despite increased clinical awareness and improved diagnostic techniques, the diagnosis of atrial myxoma may still appear as a surprise at surgery or autopsy. This is due to the tumor's variable clinical presentations and its tendency to mimic other clinical entities.1-6

The purpose of this communication is to present ten years' experience with this cardiac tumor and emphasize certain details of three patients with unusual atrial myxoma manifestations.

**Observations**

During the last ten-year period, January 1959 to January 1969, eight patients (four men and four women) with atrial myxoma, were seen at Emory University and Grady Memorial hospitals, some of which have been previously reported7 (Table 1). Six of the tumors were located in the left atrium and two in the right atrium, one of which protruded into the left atrium through an atrial septal defect. Four of the patients had symptoms of impairment of blood flow at the left atrioventricular level and the preoperative diagnosis of mitral stenosis was made, but an unexpected atrial myxoma was found in three of them at the time of thoracotomy for mitral commissurotomy, and in one at autopsy. One patient complained of dyspnea and his chest roentgenogram showed a calcified mass in the right atrium. The diagnosis of atrial tumor, probably myxoma, was made which was confirmed after its successful surgical resection.

One patient was admitted with symptoms and electrocardiographic findings indicative of acute myocardial infarction and died shortly after admission. At autopsy, an unexpected left atrial myxoma was found in addition to the acute myocardial infarction. In another instance a patient presented

---

*From the Joseph B. Whitehead Department of Surgery, Thoracic and Cardiovascular Surgery Division, Emory University School of Medicine, Atlanta, Georgia.

**Associate Professor of Surgery, Thoracic and Cardiovascular Surgery Division, Emory University School of Medicine, Atlanta, Georgia.

†Professor of Surgery, Thoracic and Cardiovascular Surgery Division, Emory University School of Medicine, Atlanta, Georgia.

Supported in part by USPHS grant No. HE05861-02.


---

504
Table 1—History, Management and Results of Treated Patients.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>Sex</th>
<th>Symptoms and Signs</th>
<th>Cardiac Catheterization</th>
<th>Angiocardiography</th>
<th>Preoperative Diagnosis</th>
<th>Operative or Autopsy Findings</th>
<th>Motive of Death</th>
<th>Operative Procedure</th>
<th>Patient Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>65</td>
<td>M</td>
<td>Chest pain</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Myocardial infarction</td>
<td>Sudden death</td>
<td>None</td>
<td>Deceased</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Dyspnea</td>
<td>No</td>
<td>No</td>
<td>RA tumor</td>
<td>RA myxoma</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>26</td>
<td>M</td>
<td>Recurrent dyspnea</td>
<td>Mobile calcified RA mass</td>
<td>No</td>
<td>No</td>
<td>RA tumor</td>
<td>RA myxoma</td>
<td>Sudden death in myocardial infarction</td>
<td>Resection of RA myxoma</td>
</tr>
<tr>
<td>3</td>
<td>51</td>
<td>F</td>
<td>Dyspnea</td>
<td>Yes</td>
<td>No</td>
<td>Mitral stenosis</td>
<td>LA myxoma</td>
<td>Sudden death while awaiting mitral commissurotomy</td>
<td>None</td>
<td>Deceased</td>
</tr>
<tr>
<td>4</td>
<td>49</td>
<td>M</td>
<td>CHF</td>
<td>Yes</td>
<td>No</td>
<td>Mitral stenosis</td>
<td>LA myxoma</td>
<td>Sudden death while awaiting reoperation</td>
<td>Cardiotomy for mitral stenosis</td>
<td>Deceased</td>
</tr>
<tr>
<td>5</td>
<td>43</td>
<td>F</td>
<td>Transient CVA</td>
<td>No</td>
<td>No</td>
<td>Mitral stenosis</td>
<td>LA myxoma</td>
<td></td>
<td>Cardiotomy for mitral stenosis</td>
<td>Dealing well</td>
</tr>
<tr>
<td>6</td>
<td>43</td>
<td>F</td>
<td>Chest pain</td>
<td>Normal</td>
<td>Yes</td>
<td>LA tumor</td>
<td>LA myxoma</td>
<td></td>
<td>Resection of LA myxoma</td>
<td>Dealing well</td>
</tr>
<tr>
<td>7</td>
<td>59</td>
<td>M</td>
<td>Orthopnea</td>
<td>LA enlargement</td>
<td>No</td>
<td>No</td>
<td>Mitral stenosis</td>
<td>LA myxoma</td>
<td>Cardiotomy for mitral stenosis</td>
<td>Dealing well</td>
</tr>
<tr>
<td>8</td>
<td>49</td>
<td>F</td>
<td>Severe cyanosis</td>
<td>Normal</td>
<td>Yes</td>
<td>Yes</td>
<td>RA tumor</td>
<td>RA myxoma</td>
<td>Reoperation of RA myxoma</td>
<td>Dealing well</td>
</tr>
</tbody>
</table>

RA = Right atrium.  CHF = Congestive heart failure.
LA = Left atrium.  CVA = Cerebral vascular accident.

constitutitional symptoms, malaise, fever, weight loss, anemia, in addition to chest pain, dyspnea and the murmur of "mitral stenosis." After prolonged but unsuccessful treatment with massive antibiotics for suspected subacute bacterial endocarditis, atrial myxoma was considered, which was diagnosed by angiocardiography and proved on a subsequent histologic examination of the surgically removed left atrial tumor. Another patient was seen with dyspnea, severe cyanosis and a grade 2/6 systolic murmur. This clinical picture was initially considered to be the result of pulmonary hypertension. However, angiocardiography and subsequent microscopic study of the excised right atrial tumor revealed the cause to be atrial myxoma.

There was no operative mortality in the five patients who had resection of their atrial myxoma and all are doing well ten months to ten years after surgery. One patient died while awaiting reopera-

CHEST, VOL. 59, NO. 5, MAY 1971

Case Reports

Case 1

In January 1962, a 43-year-old woman had an episode of nocturnal chest pain, dyspnea and nausea and two years later developed arthralgia for the first time. By September 1965 weight loss, recurrent pleuritic chest pain, dyspnea on mild exertion and cough were noted. All of her symptoms were more intense when she was lying on the left side. In January 1968 she became febrile. Two months later she was admitted to the hospital complaining of malaise, weight loss,
The diagnosis pulmonary hypertension was made after exercise 50/20 mm Hg (mean 39). The pulmonary artery wedge pressure at rest was 19, v 23 (mean 19 mm Hg) and after exercise mean 30 mm Hg. The angiocardio-gram showed a filling defect in the left atrium (Fig 1). Soon thereafter, under total cardiopulmonary bypass, a left atrial myxoma attached to the margin of the fossa ovalis was removed (Fig 2). On histologic examination the findings of the resected specimen were compatible with myxoma. Subsequent to surgery she has been asymptomatic.

Case 8

A 49-year-old woman was seen as an outpatient in April 1967, because of vague upper abdominal pain. Physical and radiologic examinations were unremarkable. In June 1967 she developed weakness, dyspnea on exertion, hepatomegaly, pretrial edema. Hospital study revealed that the hemogram, urine analysis, serum albumin, total serum bilirubin, electrocardiogram, and chest roentgenogram were within normal limits. The serum alkaline phosphatase was 22.5 King Armstrong units, the serum amylase 223 units, the sulfobromophthalein (Bromsulphalein) retention at 45 minutes was 32 percent. The liver scan showed diffuse areas of decreased uptake. Percutaneous liver biopsy demonstrated nonspecific fatty infiltration and exploratory laparotomy, performed to confirm a clinical impression of pancreatic carcinoma, revealed only an enlarged liver. The liver biopsy obtained at the time of laparotomy showed "portal and periporal chronic inflammation with periporal scarring and marked sinusoidal dilation."

During this procedure, she suddenly developed intense cyanosis which gradually cleared over the next two days. Following this episode, she did well and in December 1967, she underwent an uneventful total abdominal hysterectomy for vaginal bleeding secondary to uterine fibromyoma. In April 1968, she noted cyanosis of the fingernails, ears and tongue and a progressive onset of dyspnea which by November 1968, markedly limited her activity. By this time she developed clubbing of the fingers. The arterial blood pO2 was 43 mm Hg, which became 52 mm Hg after the administration of 100 percent oxygen by mask, the PCO2 27 mm Hg and the pH 7.46.

In December 1968, she was hospitalized with a tentative diagnosis of "pulmonary hypertension." The only abnormal findings on admission were: symmetrical cyanosis with moderate clubbing of the fingers and toes, mild pitting edema of the ankles, a soft grade 2/6 middiastolic murmur audible along the left sternal border and an enlarged liver 5 cm below the right costal margin which was firm, smooth, slightly tender and nonpulsatile.

Right heart catheterization was performed, during which the catheter met resistance in the right atrium and could not be advanced from the superior to the inferior vena cava nor vice versa but easily crossed the atrial septum. A right-to-left shunt at the atrial level was demonstrated by indicator dilution curves, which was estimated by oximetry to be 3.2 L/min. Angiocardiography was performed via both superior and inferior vena cavae, demonstrating a large apparently immobile defect within the right atrial chamber and the interatrial shunt (Fig 3).

In January 1969, the patient was operated upon. The heart was exposed through a right anterolateral thoracotomy incision and the anterior pericardium was resected and preserved in sterile saline. On gentle palpation of the right atrium, a hard tumor occupying practically the entire chamber was felt. The tumor extended down to the inferior vena cava and
precluded cannulation of the inferior vena cava through the right atrium. The inferior vena cava, therefore, was cannulated through the femoral vein and the superior vena cava through the appendage of the right atrium. Following cannulation of the left femoral artery, the patient was placed on cardiopulmonary bypass. The inferior vena cava was dissected off the diaphragm and was occluded with a vascular clamp. The right atrium was opened close to the atrioventricular groove and a gray, greenish tumor was visualized which occupied the right atrial cavity and herniated into the left atrium through an atrial septal defect (Fig 4). The tumor was partially obstructing the orifice of the tricuspid valve. Because of its extensive attachment to the right atrial wall and the interatrial septum, the tumor with the right atrial wall and the interatrial septum were resected. After resection, the left atrium and left ventricle were thoroughly irrigated with saline. The interatrial septum was then reconstructed with a Teflon felt patch and the right atrial wall was reconstructed with the preserved excised pericardium (Fig 5). Pathologic examination revealed a gelatinous tumor which had the microscopic characteristics of a myxoma.

Postoperatively the patient was noted to have a left hemiparesis and to be partially aphasic. These neurological deficits were thought to be the result of tumor or air embolization. The aphasia cleared completely and there has been partial resolution of the hemiparesis. Her cyanosis disappeared and she is currently free of cardiopulmonary symptoms. In January 1970 right heart catheterization and angiocardiography were repeated and showed normal pressures, no evidence of shunt and normal appearing right atrium (Fig 6).

**Case 7**

A 59-year-old man, in 1967, noted onset of shortness of breath on exertion which progressed by 1968 to severe dyspnea. By that time he noted paroxysmal nocturnal dyspnea when sleeping on his back or left side but not while lying on his right side, and precordial chest pain occurring especially on exertion. He was then seen by his private physician and was started on digoxin and diuretics without improvement. In February 1969 he was admitted to the hospital at which time a loud S1 and P2, a soft presystolic rumble at the apex and an opening snap at 0.10 to 0.11 seconds at the apex and left sternal border were heard but no diastolic murmur was present. The electrocardiogram and the chest roentgenogram were considered compatible with mitral stenosis (Fig 7). Two days after admission he was operated upon for closed commissurotomy but when the finger was inserted into the left atrium a tumor was palpated. Cardiopulmonary bypass was instituted and a tumor attached to the margin of the fossa ovalis was removed (Fig 8). Pathologic examination showed a 5.5 × 4.5 × 4.0 tumor which had the typical histologic picture of an atrial myxoma. Subsequent to surgery the patient has been asymptomatic.
Myxoma of the heart is the most common of all benign cardiac tumors and predominantly involves the atria, the left one three times more frequently than the right. It may appear either as a semi-translucent, gelatinous sessile, lobulated, or as a round firm, green or pink mass, the latter is almost always attached with a pedicle to the interatrial septum at the margin of the fossa ovalis. Histologically, the bulk of the myxoma consists of an amorphous matrix which is considered to be predominantly an acid mucopolysaccharide related to chondroitin-6-sulfate.

The clinical manifestations of the myxoma are variable and are dependent, to a certain extent, on the location of the tumor. The myxomas can mimic several other clinical entities but basically present themselves in three ways: by embolization, by obstruction of intracardiac blood flow and by constitutional disturbances.

Virtually any artery in the body may be occluded by myxoma emboli. For this reason, the diagnosis of myxoma should be considered in a patient who presents signs and symptoms due to embolism of no apparent cause, and the embolectomy specimen should always be examined histologically because there are several myxomas diagnosed in this way.

The majority of patients with atrial myxoma present symptoms of stenosis of the atrioventricular valves and in many of them the diagnosis of left atrial myxoma will be made preoperatively.
atrial myxoma is made at the time of surgery for a planned closed mitral comissurotomy. Four of the reported cases herein had symptoms of mitral stenosis, in three of which the correct diagnosis was made at surgery. In one of the patients operated upon for mitral stenosis, the resection of the tumor was postponed for a later day because then the use of prime solutions other than blood and disposable oxygenators was not practiced. He died suddenly the next day as did an additional patient with similar symptoms while he was awaiting surgery, "mitral comissurotomy." These two cases emphasize the risk of postponing the resection of the myxoma. Resection of this tumor now is immediately performed if it is found unexpectedly at the time of closed comissurotomy, or as soon as possible if the diagnosis is made preoperatively. The variability of the symptoms and signs with the change in the patient's position and the odd crushing noises, which are the result of the tumor rubbing on the endocardial surface are rarely present, but are the most valuable of the diagnostic physical findings.

Fever, anorexia, malaise, weight loss, raised sedimentation rate, anemia and elevation of gamma globulin are the constitutional manifestations of atrial myxoma. Some of these patients are admitted and treated for subacute bacterial endocarditis. However, if the patient fails to respond to the proper treatment and blood cultures are persistently negative, as seen in case 1, the possibility of cardiac myxoma should be investigated.

The factors responsible for the constitutional manifestations are not clear. The presence of breakdown products released from the myxoma into the bloodstream have been suggested. An immune response to fragments of tumor released into the circulation has been suggested as a possible cause of the increase in gamma globulin. The possibility that an intracardiac myxoma initiates and maintains an autoimmune state analogous to that seen after cardiotomy or myocardial infarction has also been suggested.

Patients with myxoma of the right atrium and tricuspid valve obstruction may have high hemoglobin instead of anemia and arterial oxygen desat-
uration (as seen in case 2) due to the right-to-left shunt through a coexisting atrial septal defect or through a dilated foramen ovale.

The diagnosis of an intra-atrial mass, myxoma, is established in nearly 100 percent of the cases with angiocardiography.2,20 The radiopaque material is injected into the cavae for right atrial myxoma and into the pulmonary artery for those on the left side.21

The development of the cardiopulmonary bypass unit has made resection of the atrial myxoma possible and the treatment of this tumor gratifying. Resection of the atrial myxoma under total cardiopulmonary bypass should be performed shortly after the diagnosis is established. The excision of the area of the septum where the atrial myxoma is attached remains a controversial point.16,22 Most authors suggest this is not necessary,18 while others, having seen recurrence of the atrial myxoma at the site of the previously transected tumor pedicle, feel that resection of the cardiac wall at the tumor attachment is necessary.22 Two of the reported patients herein had resection of their tumor with its pedicle, two had, in addition, resection of the atrial septum where the myxoma was attached and one had resection of the intraatrial septum and right atrial wall in addition to the tumor. None of the resected myxomas recurred in the 13 months to ten years' follow-up period.

REFERENCES