Calcified Left Atrial Myxoma
Simulating Mitral Insufficiency

Hemodynamic and Phonocardiographic Effects
of Tumor Movement

By John L. Penny, M.D., John J. Gregory, M.D., Stephen M. Ayres, M.D.,
Stanley Giannelli, Jr., M.D., and Plinio Rossi, M.D.

SUMMARY
In the atypical case presented, a calcified myxoma of the left atrium simulated isolated mitral insufficiency. Unique hemodynamic and phonocardiographic events were recorded. A notch in the upstroke of the left ventricular pressure tracing during isometric contraction occurred simultaneously with an ejection sound, which fused with the first heart sound, and with an abrupt decrease in ventricular volume as the tumor was ejected from the ventricle. A similar notch in the downstroke of the ventricular pressure tracing was most likely due to the abrupt increase in ventricular volume as the tumor descended into the left ventricle. A third heart sound was present which was identified as a ventricular diastolic gallop and was differentiated from a mitral opening snap by simultaneous pressure tracings.

Additional Indexing Words:
Ejection sound Ventricular diastolic gallop Ventricular pressure tracing

Atrial Myxomas are relatively rare lesions, but antemortem diagnosis has been made with increasing frequency in the past 10 years. Left atrial myxomas usually present as mitral stenosis with obstructive symptoms, and occasionally as systemic emboli, subacute bacterial endocarditis, or nonspecific constitutional symptoms. Mitral insufficiency as the sole feature of left atrial myxoma is very infrequent, and only two such cases have been documented in the literature.1,2 Calcification in the region of the mitral valve also reduces the likelihood of myxoma.3 The purpose of this report is to describe an unusual case of a successfully excised, calcified left atrial myxoma without constitutional or embolic symptoms, which simulated a case of isolated mitral insufficiency. The clinical manifestations and unusual hemodynamic and phonocardiographic events will be discussed.

Report of Case
A 48-year-old white woman was admitted to the hospital on March 15, 1966, because of dyspnea on exertion. Though she was in excellent health, a heart murmur had been noted 5 years previously. Three years before admission she developed congestive heart failure and was admitted to another hospital where the diagnosis of rheumatic mitral insufficiency was made. She improved after digitalization and was maintained on digoxin and penicillin therapy. Symptoms gradually returned but were unrelated to change in position.

On physical examination, the blood pressure was 120/70 mm Hg, the pulse was regular

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Simultaneous pulmonary capillary wedge, left ventricular pressure, and phonocardiogram recorded at the apex. The notch on the upstroke of the ventricular pressure is due to the sudden movement of the myxoma into left atrium during isometric contraction. This notch coincides with the tumor ejection sound (ES) which is fused with the first heart sound (S1). A similar notch is recorded on the downstroke of the left ventricular pressure and corresponds to opening of the mitral valve and the sudden increase in ventricular volume as the tumor enters the ventricle. A ventricular diastolic gallop (VDG) is inscribed 0.12 sec after the beginning of the second sound (S2) and occurs at the nadir of the Y descent of the pulmonary wedge pressure. The shaded area shows the small transmitral gradient.

with a rate of 88/min, respirations were 22/min, and oral temperature was 98.6 F. Pertinent findings were limited to the heart. There was a mild left parasternal heave, and the apical impulse was just outside the midclavicular line. The first sound was loud and was fused with an early systolic ejection sound. The second sound was normal. There was a grade II/VI blowing, high-pitched holosystolic murmur at the apex. A third heart sound was heard at the apex in early diastole. No diastolic murmur was heard, and the auscultatory findings did not vary with change in position.

The electrocardiogram showed sinus rhythm and digitalis effect but was otherwise normal. Fluoroscopy showed evidence of slight enlargement of the left atrium, right ventricle, and possibly the left ventricle. There was calcification in the area of the mitral valve. Cardiac catheterization data are presented in table 1, and the phonocardiogram and left ventricular and pulmonary “capillary” wedge pressures are shown in figure 1. Cineangiocardiography with injection of contrast material into the left ventricle demonstrated a large, round, lobulated filling defect which passed from the left ventricle to the left atrium with each systole, and back into the left ventricle in diastole (fig. 2). There was moderate regurgitation of contrast material into the left atrium.

At open heart surgery, the left atrium contained a 4 by 3 cm yellow, gelatinous, lobulated tumor attached to the lower interatrial septum just above the mitral annulus approximately 2 cm below the fossa ovalis by a base, 1 cm in diameter, which was partially calcified. The tumor was excised completely. The mitral valve was normal except for slight dilatation of the
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Table 1

<table>
<thead>
<tr>
<th>Study</th>
<th>At rest</th>
<th>During exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pressures (mm Hg)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>(3)*</td>
<td></td>
</tr>
<tr>
<td>Right ventricle</td>
<td>48/6*</td>
<td>60/8</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>48/24 (35)</td>
<td>60/29 (41)</td>
</tr>
<tr>
<td>Pulmonary “capillary” wedge</td>
<td>(20)</td>
<td></td>
</tr>
<tr>
<td>Left ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ascending aorta</td>
<td>105/6</td>
<td>105/65 (79)</td>
</tr>
<tr>
<td>Other data</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Arterial oxyhemoglobin saturation (%)</td>
<td>87.7</td>
<td></td>
</tr>
<tr>
<td>Arteriovenous oxygen difference (vol %)</td>
<td>4.09</td>
<td></td>
</tr>
<tr>
<td>Oxygen consumption (ml/min/m²)</td>
<td>102</td>
<td></td>
</tr>
<tr>
<td>Cardiac index (L/min/m²)</td>
<td>2.5</td>
<td></td>
</tr>
<tr>
<td>Pulmonary vascular resistance (dyne sec cm⁻⁵)</td>
<td>267</td>
<td></td>
</tr>
<tr>
<td>Mean mitral gradient (mm Hg)</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>End-diastolic mitral gradient (mm Hg)</td>
<td>4</td>
<td></td>
</tr>
</tbody>
</table>

*Pressures (mm Hg): systolic/diastolic; mean in parentheses.

Figure 2

A 35-mm cineangiogram showing a large lobulated filling defect first in the left ventricle (LV) in A, traversing the mitral valve in B, and in the left atrium (LA) in C. The ascending aorta (A) is seen above.

annulus, and there was no valvular calcification or fibrosis. The postoperative course was uneventful, and the patient has returned to full-time work without medication.

Discussion

The clinical diagnosis of left atrial myxoma is difficult, and many unsuspected myxomas have been found during mitral valve surgery and angiocardiography. Approximately 75% are located in the left atrium, originating from the atrial septum in the region of the fossa ovalis. The exact origin of myxomas is still controversial, but recently, authors agree that they are neoplasms rather than organized thrombi. Histochemical studies show similar staining properties with cutaneous myxomas and umbilical cord tissue. Thrombi lack these staining characteristics.

Goodwin, and Campeau and David in excellent reviews have classified the clinical features of left atrial myxoma under these headings: (1) obstructive, (2) embolic and,
(3) constitutional. The obstructive symptoms such as syncope, vertigo, and dyspnea are caused by varying degrees of occlusion of the mitral orifice by the tumor. In the case presented here, the only feature of obstruction was dyspnea on exertion which clinically was attributed to mitral regurgitation. Goodwin\textsuperscript{5} believes that too much emphasis has been placed on syncope as a symptom, for it has been present in only 8 to 20\% of cases in two large series. Although a small degree of mitral valve obstruction was manifested by the end-diastolic transmural pressure gradient of 4 mm Hg, the predominant hemodynamic lesion in this case was mitral regurgitation. This was due to dilatation of the mitral ring and stretching of the valve leaflets by the large myxoma traversing the mitral valve with each systole and diastole.

Clinical evidence of embolic phenomenon was completely absent in our case. This is unusual since most series\textsuperscript{6,7} report a 40 to 60\% incidence of systemic embolization. In several cases,\textsuperscript{6,7} the diagnosis of myxoma has been made on microscopic examination of tissue removed at embolectomy. Pulmonary emboli, which originate from the pelvic or leg veins, or thromboses in situ are also reported in approximately 25\% of left atrial myxomas. Constitutional features such as fever, anemia,\textsuperscript{8} loss of weight, and abnormal serum proteins were absent in our case.

Auscultation of patients with left atrial myxomas usually reveals findings similar to those of mitral stenosis accompanied by an early diastolic third heart sound which has been called an “opening snap” by some authors\textsuperscript{9,10} and a “ventricular gallop” by others.\textsuperscript{1,2,7,11} The first sound is usually accentuated and slightly delayed. Systolic murmurs have been recorded and have been attributed to either associated mitral or tricuspid insufficiency.

The phonocardiogram (fig. 1) in this case showed a high-pitched, holosystolic, apical murmur of moderate amplitude. The prominent first heart sound was preceded by, and fused with, a loud ejection sound which was most likely due to the forceful ejection of the tumor from the left ventricle back into the left atrium, striking the mitral orifice or the atrial wall. Since the mitral valve could not close until the tumor was back in the atrium, the latter part of this loud sound can be attributed to the loud closure of the mitral valve from a descended position. The ejection sound occurred at the end of isometric contraction simultaneously with a notch in the upstroke of the ventricular pressure tracing. This sudden brief change in the slope of the ventricular pressure may have been caused by an abrupt decrease in ventricular volume as the tumor is ejected from the ventricle.\textsuperscript{12} A similar notch occurring on the down slope of ventricular pressure tracing is attributable to a sudden increase in ventricular volume as the tumor entered the left ventricle. The second heart sound was normally split, was of low intensity, and was followed by a third heart sound which we interpret as a ventricular gallop. The simultaneous phonocardiogram and pressure tracings verify this interpretation. The pulmonary wedge pressure, equivalent to the left atrial pressure, is usually delayed by 0.04 to 0.08 sec.\textsuperscript{13} If the pulmonary wedge pressure in figure 1 is moved to the left 0.04 sec, the ventricular gallop sound falls at the nadir of the Y descent. The opening snap should occur at the beginning of the Y descent. Two alternate mechanisms of this third sound are: (1) the diastolic sound of mitral insufficiency due to increased flow or (2) the sound of the tumor hitting the mitral valve or the left ventricular wall on its descent in early diastole.

In retrospect, the only significant clinical clue to the diagnosis of left atrial myxoma in this case was the first heart sound which fused with an ejection sound of the tumor moving from the ventricle to the atrium. In the absence of other causes of an ejection sound, atrial myxoma should be considered and appropriate diagnostic procedures carried out to exclude or discover this curable lesion.

References

The Primary Physician
Foresight in Medical Education

We are engaged in a renegotiation of the contract between our profession and the society it serves. As far as society is concerned the renegotiation involves issues of comprehensiveness, availability, individualized adaptation and personalized application of medical care. However from the standpoint of the medical profession or the health professions generally, and particularly the teachers in these areas, the central issue is a matter of division of professional labor. Most of the discussions we are having in medical schools these days center on these issues. We are trying to decide, for example, what kind of doctors we need and what kind of training they need.

The division of labor in modern medicine is characterized by development of ever greater expertise in ever more narrow segments of medicine. . .

However this division of labor within medicine is ill suited for dealing with the primary data. It “prefers” its “clinical material” pre-sorted or pre-processed, after which the specialty apparatus works best. But to provide the comprehensive individually adapted care which the public wants and needs, the vertically oriented compartmentalized specialties of medicine require a horizontal integrative component. . . . What we lack in our medical apparatus today is a sufficient number of graduates whose professional commitment is to the unprocessed, unrefined patient and whose clinical material is centered in the primary data.—Richard M. Magraw: Medicine’s Primary Data and the Primary Physician. Med Bull Univ Minnesota 38: 274-275, 1967.
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