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Myxoma of the Left Atrium Simulating Mitral Stenosis  

By John W. Harrison, M.D., Lawrence J. McCormack, M.D., and A. Carlton Ernsten, M.D.

Myxoma of the left atrium, although of rare occurrence, is the most common form of primary tumor of the heart.\(^1\) In the past, its recognition was a matter of academic interest only, but with the development of intracardiac surgery, its detection has become of practical importance. In the first place, the tumor may cause auscultatory and hemodynamic changes that may lead to an erroneous diagnosis of mitral stenosis. Second, myxomas of the atrium usually are pedunculated, and their surgical removal should be possible. The present case is reported to emphasize these two points and to direct attention to certain clinical features of atrial tumors that may aid in their diagnosis.

Abstract of Case

A white, married woman, aged 50 years, was admitted to the Cleveland Clinic Hospital on March 14, 1953, because of dyspnea, orthopnea, cough, abdominal discomfort, nausea and occasional vomiting. Shortness of breath on exertion, fatigue and weakness had first been noted approximately nine months earlier. These symptoms had become progressively worse, and for three months prior to entering the hospital there had been orthopnea and a persistent cough productive at times of pink-tinged frothy sputum. On two occasions the patient had been awakened in the early morning hours by severe paroxysmal dyspnea. Nausea and occasional vomiting had developed in January, 1953, following treatment with digoxin and a low-sodium diet and, because of the persistence of these symptoms, all therapy had been discontinued two weeks before admission. Edema of the ankles first was noted two days before entering the hospital. There had been a gradual loss of 14 pounds in weight since the onset of the illness.

The past medical history did not contribute significant information. There had been no illness suggestive of rheumatic fever, and the patient never had been informed of the presence of a heart murmur.

Physical examination revealed a well-developed and fairly well-nourished woman in no respiratory distress when well propped up in bed. The temperature was normal, the heart rate 100 per minute, and the blood pressure 100 mm. Hg systolic and 70 mm. diastolic. The veins of the neck were moderately distended, but there was no cyanosis and no malar flush. The area of relative cardiac dullness extended 11 cm. from the midsternal line in the fifth intercostal space. The heart rhythm was regular except for occasional premature beats. The first sound at the apex and the second sound at the pulmonary area were accentuated. The initial examiner reported the presence of a grade 2 late diastolic apical murmur with a presystolic crescendo. The lungs were clear on percussion, but on auscultation a moderate number of medium moist rales could be heard over the right base posteriorly. The liver extended 4 cm. below the costal margin in the right midclavicular line and was slightly tender. There was no peripheral edema.

On the morning following admission, the patient was examined by several members of the medical staff, including the original examiner, and none could confirm the presence of a mitral diastolic murmur, even with the patient lying in the left lateral position. Four days later, however, one examiner again detected the murmur.

The urinalysis and blood count gave normal findings, and the Wassermann reaction of the blood was negative.

An electrocardiogram showed sinus tachycardia with a rate of 110 per minute and changes in the preordial leads indicative of right ventricular hypertrophy.

Fluoroscopic and roentgenographic studies of the thorax revealed a moderate increase in the size of the heart, the enlargement involving predominantly the outflow tract of the right ventricle (fig. 1). Studies in the oblique positions showed slight enlargement of the right atrium but only questionable enlargement of the left atrium. The left ventricle appeared to be of normal size, and no valvular calcifications could be seen. The hilar vessels were prominent bilaterally but did not pulsate.

Treatment with digitalis, a low-sodium diet, and mercurial diuretics resulted in prompt improvement,
and on the fourth day Dr. F. Mason Sones, Jr., performed cardiac catheterization. The results of the studies are presented in the table.

The patient continued to improve, but on several occasions she experienced sudden spells of nausea, colicky pain in the upper abdomen, and an abrupt return of severe dyspnea, tachypnea, and orthopnea.

**Dr. Harrison:** On the basis of the clinical, roentgenographic and electrocardiographic observations, a diagnosis of rheumatic heart disease with severe mitral stenosis appeared to be justified. The fact that a diastolic apical murmur was detected on only two or three occasions and never was heard simultaneously by two or more observers was a disturbing feature. The elevated pulmonary artery and pulmonary "capillary" pressure, however, indicated a high resistance to blood flow beyond the pulmonary arteriolar bed, and this was considered further evidence in favor of the presence of mitral stenosis. The pulmonary arteriolar resistance and the total pulmonary resistance at rest were about four times that normally encountered. It was presumed that the orifice of the mitral valve was so greatly reduced that a diastolic murmur either was not produced or occurred only inconstantly.

**Dr. Ernstene:** I believe that that was a reasonable assumption. The murmur of mitral stenosis is difficult to hear in certain patients, and the ease with which it is detected may vary from time to time. Differences in the heart rate are responsible for some of these fluctuations, but I suspect that variation in the auditory acuity of the examiner also is an important factor. At times the murmur of mitral stenosis may be detectable only after exercise or with the patient lying in the left lateral position, but neither of these maneuvers was of help in the present instance. It has been emphasized repeatedly, chiefly with reference to the murmur of mitral insufficiency and some of the congenital cardiac anomalies, that no parallelism exists between the intensity of a murmur and the size of the aperture through which the blood is flowing. Either a very small or a very large defect may fail to produce a bruit.

The first sound at the apex and the pulmonary second sound were accentuated. These findings are, of course, consistent with the presence of mitral stenosis. No "opening snap" of the mitral valve was heard at any time.

![Roentgenogram of the thorax demonstrating enlargement of the outflow tract of the right ventricle.](image)

**FIG. 1.** Roentgenogram of the thorax demonstrating enlargement of the outflow tract of the right ventricle.

### Table 1.—Results of Cardiac Catheterization Studies

<table>
<thead>
<tr>
<th></th>
<th>Rest</th>
<th>Exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oxygen capacity of blood</td>
<td>15.3 vol. %</td>
<td>13.3 vol. %</td>
</tr>
<tr>
<td>Oxygen content of arterial blood</td>
<td>13.6 vol. %</td>
<td>14.0 vol. %</td>
</tr>
<tr>
<td>Oxygen content of pulmonary &quot;capillary&quot; blood</td>
<td>14.8 vol. %</td>
<td>5.2 vol. %</td>
</tr>
<tr>
<td>Oxygen content of pulmonary artery blood</td>
<td>9.0 vol. %</td>
<td>3.82 L./min.</td>
</tr>
<tr>
<td>Cardiac output</td>
<td>5.33 L./min.</td>
<td>2.57 L./min./M.</td>
</tr>
<tr>
<td>Cardiac index</td>
<td>3.57 L./min./M.²</td>
<td>88/43 (63) mm. Hg</td>
</tr>
<tr>
<td>Mean pulmonary artery pressure</td>
<td>91/41 (62) mm. Hg</td>
<td></td>
</tr>
<tr>
<td>Mean pulmonary &quot;capillary&quot; pressure</td>
<td>26 mm. Hg</td>
<td></td>
</tr>
<tr>
<td>Right ventricular pressure</td>
<td>88/9 mm. Hg</td>
<td></td>
</tr>
<tr>
<td>Mean right auricular pressure</td>
<td>7 mm. Hg</td>
<td></td>
</tr>
<tr>
<td>Pulmonary arteriolar resistance</td>
<td>384 dynes/sec./cm.²</td>
<td></td>
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<tr>
<td>Total pulmonary resistance</td>
<td>920 dynes/sec./cm.²</td>
<td></td>
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One roentgenologic feature was present that should have called for caution in accepting the diagnosis of mitral stenosis, namely, the fact that only slight or questionable enlargement of the left atrium could be demonstrated. If severe mitral stenosis were responsible for the patient's congestive failure, the valvular lesion should have been present for a long time and ordinarily would have produced considerable enlargement of the atrium. I should like to ask Dr. Harrison how severe the congestive failure was and whether or not the possibility of primary pulmonary disease was considered as a cause of illness.

Dr. Harrison: At the time of the patient's admission, orthopnea was present; the jugular veins were engorged even with the patient sitting erect; there were rales over the base of the right lung; and the liver was large and tender. Furthermore, there was a loss of 16 pounds in weight during the first seven days in the hospital, after which the weight remained stationary.

With regard to the question of primary lung disease, roentgenographic examination of the chest showed no evidence of any such condition. Complete pulmonary function studies were made and revealed a 20 per cent reduction in the vital capacity of the lungs, a normal residual air space, moderate reduction in total lung volume, normal maximum breathing capacity, and hyperventilation at rest. These findings were interpreted as being consistent with mild pulmonary fibrosis or pulmonary congestion, and in view of the clinical response to treatment it was believed that the latter explanation was the correct one.

Dr. Ernstene: In the daily progress notes several episodes are described in which there was an abrupt return of dyspnea and orthopnea accompanied by nausea and abdominal pain. What was the cause of those spells?

Dr. Harrison: No adequate explanation was offered. It was believed that acute left ventricular failure could be excluded in view of the fact that the left ventricle was not enlarged and the attacks continued to recur even after the patient's condition otherwise was much improved. The paroxysms occurred without detectable precipitating factors and were not accompanied by an increase in the number of rales in the lungs or by more than a moderate rise in the heart rate. Because the patient invariably became upset and anxious, the possibility was considered that the respiratory distress was part of an emotional reaction to abdominal pain. However, roentgenographic studies of the gallbladder and upper gastrointestinal tract revealed no abnormalities, and repeated measurements of the serum amylase also were normal. The liver did not seem to increase in size or become more tender during the attacks.

Dr. Ernstene: Will you continue with the clinical history? It appears that everyone who examined the patient ultimately accepted the diagnosis of mitral stenosis.

Dr. Harrison: The diagnosis was considered established and mitral commissurotomy was advised. The patient, however, requested a temporary postponement, and because her condition had improved satisfactorily, she was allowed to return home for one month. Three weeks after leaving the hospital she was readmitted because of a sudden return of dyspnea and orthopnea, cough productive of blood-tinged sputum, pain in the right flank and right upper abdomen, and abdominal distention. There had been repeated vomiting. The body weight had not increased.

On examination, the patient appeared acutely ill. There was great respiratory distress, the neck veins were distended, and the skin was pale, cool, and clammy. Cyanosis was not noted. The heart rate was 104 per minute, and the blood pressure 70 mm. Hg systolic and 40 mm. diastolic. There were no cardiac murmurs. A few coarse moist rales were present over the base of the right lung. The liver extended 5 cm. below the costal margin in the right midclavicular line and was very tender. There was minimal edema about both ankles. A diagnosis of pulmonary infarction was made. On the following day, icterus was noted for the first time. The blood urea content was 87 mg. per 100 cc. Roentgenograms of the chest showed an irregular area of increased density in the lower lobe of the right lung, approximately 4 cm. in diameter.

The patient's condition deteriorated gradually but steadily. Rales appeared in increasing number; dyspnea and orthopnea persisted; abdominal distention increased; the edema of the legs became more marked; and there was progressively severe prostration. Periods of confusion and stupor developed and deepened gradually to terminal coma. The arms and legs remained cold and pale, and tachycardia and hypotension persisted. Death occurred on the ninth day after admission.
Dr. Ernstene: Was pulmonary infarction considered to be the principal factor responsible for the terminal course of events?

Dr. Harrison: No. The area of infarction did not seem large enough for that. The primary cause was considered to be a change in the condition of the heart. The appearance of the patient and the clinical course raised increasing doubt in the minds of several consultants that the primary cardiac problem was mitral stenosis. No positive diagnosis was offered, but in discussing the problem the possibilities of ball-valve thrombus and tumor of the left atrium were mentioned. The first of these conditions was not considered likely because occluding thrombi of the left atrium usually are associated with mitral stenosis and/or auricular fibrillation. In this case, the presence of mitral stenosis appeared increasingly doubtful, and the heart rhythm remained regular throughout the illness. The persistent hypotension, tachycardia, and coldness of the extremities were, of course, compatible with high-grade occlusion of the mitral orifice by a thrombus. The possibility of a left atrial tumor was not considered beyond the point of passing comment.

I should like to ask Dr. McCormack to present the postmortem findings.

Dr. McCormack: The pertinent necropsy findings were observed in the heart, lungs, liver, and adrenal glands.

The heart was symmetrically enlarged and weighed 350 Gm. No localized chamber enlargement was present. The epicardial surface was normal. The coronary arteries were normal. The myocardium was firm and reddish tan in color. The right ventricle measured 0.7 cm. and the left 1.2 cm. in thickness, signifying some muscular hypertrophy. The leaflets of all valves were entirely normal, and there were no scarring, thickening, or shortening of the chordae tendineae.

Within the left atrium, a rubbery, slightly lobulated mass was firmly attached by a short pedicle to the interatrial septum in the region of the posterior portion of the valve of the fossa ovalis (fig. 2). The tumor was mottled red, gray, and yellow and measured 5.0 cm. in length and 4.0 cm. in diameter. The pedicle was 1.5 cm. in diameter. On cross section, the mass was homogeneous, gray, and translucent.

Microscopically, the tumor possessed a uniform appearance and presented as its outstanding feature a large number of blood vessels of capillary size coursing through the lesion (fig. 3). Areas of focal thickening of the walls of these vessels by small spindle cells imparted a peritheliomatous pattern. The intervening tissue was loose, myxomatous, and relatively acellular. An occasional stellate reticular cell as well as a rare macrophage was seen. Reticulum stains demonstrated fine and broad fibers coursing through the tumor but only occasionally were they
arranged around vessels. Invasion of the underlying atrial wall was not present.

The right lung weighed 575 Gm. and the left, 350 Gm. The pleural surface of the right lung was covered by a fibrinous exudate. The lower lobe contained several wedge-shaped, purple, firm areas ranging in size from 1.0 to 3.0 cm. In the lowermost portion there was a similar area, 5 cm. in diameter, with a softened center and, at its apex an adherent intra-arterial, soft, friable thrombus that histologically was partially organized. The left lung presented no pleural reaction but contained several additional small areas of infarction.

Microscopically, the walls of the pulmonary arterioles were greatly thickened and hyalinized (fig. 4).

The liver was finely nodular, brown and small, weighing 1070 Gm. The cut surface had a "mimic" appearance characteristic of acute hemorrhagic central necrosis.

The adrenal glands were bilaterally enlarged, weighing together 45 Gm. Grossly and histologically the cortices were thickened. The various zones were distinct, and abundant lipid was present.

The anatomic diagnoses were: Myxoma of the left atrium; cardiac hypertrophy; pulmonary infarction (multiple); thrombosis (embolus?) of small pulmonary artery; extensive hyalinization of pulmonary arterioles; acute hemorrhagic central necrosis of the liver; hyperplasia of the adrenal glands ("stress phenomenon").

**Dr. Ernstene:** Were the changes in the pulmonary arterioles similar to those commonly present in patients who have mitral stenosis?

**Dr. McCormack:** The changes differed somewhat in that hyalinization was a much more prominent feature than is usual in association with mitral stenosis.

**Dr. Harrison:** Systemic arterial embolism has been reported in several instances of myxoma of the left atrium, the emboli consisting of fragments of neoplastic tissue or thrombotic material from the surface of the tumor. Was there any evidence of such emboli in this case?

**Dr. McCormack:** No. The endothelium was intact over the tumor, and there were no areas of ulceration or attached thrombi.

**Dr. Ernstene:** This, then is a case of myxoma of the left atrium in which a number of features originally suggested the presence of mitral stenosis. Should a correct antemortem diagnosis have been made? The highly questionable presence of a mitral diastolic murmur and the absence of appreciable enlargement of the left atrium undoubtedly should have suggested, during the first period of observation, some process other than long-standing disease of the mitral valve. It appears also that insufficient attention was paid to the inconsistency between the elevation of pulmonary artery and pulmonary "capillary" pressures, on the one hand, and the lack of positive evidence of mitral stenosis, left atrial enlargement, or primary pulmonary disease on the other. This suggests that the obstruction to blood flow may have occurred at the orifices of the pulmonary veins rather than at the mitral valve. Will Dr. McCormack tell us whether this was possible from his necropsy findings?

**Dr. McCormack:** Our findings do not appear to make an obstruction of that kind very likely. When the heart was reconstructed, the tumor mass prolapsed into the orifice of the mitral valve.

**Dr. Ernstene:** The paroxysms of dyspnea and orthopnea probably were due to temporary prolapse of that kind. But if chronic obstruction of the mitral valve orifice was the sole cause of the pulmonary hypertension that must have
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been present for a considerable time to produce the changes in the pulmonary arterioles, it is strange that more than minimal dilatation of the left atrium should not have resulted. Chronic obstruction of the orifices of the pulmonary veins would explain the findings very well.

The abdominal pain that accompanied the attacks of paroxysmal dyspnea probably was due to a rapid increase in venous congestion of the liver, although physical examination did not give demonstrable proof of such a change.

To summarize, if one considers all aspects of the case and especially the doubt as to the presence of a diastolic apical murmur, the absence of definite enlargement of the left atrium, and the recurrent, abrupt changes in the circulation, a diagnosis of atrial tumor would have been warranted.

Dr. HARRISON: It is of interest that at one time the decision to operate for the purpose of performing a mitral commissurotomy had been made, but circumstances forced a delay. Although the removal of tumors of this type appears feasible, it is doubtful that the lesion in the present case could have been extirpated through the customary approach by way of the left atrial appendage. An accurate diagnosis and the planning of an operation involving some such procedure as the "well" of Gross and his co-workers2 might have led to a favorable outcome.

SUMMARY

A case of myxoma of the left atrium originally diagnosed as mitral stenosis has been presented. Analysis of the clinical and roentgenologic features and the data obtained from cardiac catheterization indicate that a correct diagnosis probably could have been made. Certain of the findings suggest that although prolapse of the tumor into the orifice of the mitral valve was responsible for striking changes in the patient's condition, the site of the chronic obstruction responsible for the pulmonary hypertension and the changes in the pulmonary arterioles may have been at the atrial orifices of the pulmonary veins.

REFERENCES

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JOHN W. HARRISON, LAWRENCE J. MCCORMACK and A. CARLTON ERNSTENE

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