Myxoma of the Left Atrium

Hemodynamic and Phonocardiographic Consequences of Sudden Tumor Movement

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SUMMARY

Two patients with myxoma of the left atrium were studied by left heart catheterization and cineangiography, and the diagnosis was confirmed at operation in both cases. An electrocardiographic timing signal on the cineradiographs permitted correlation of heart sounds and pressure waves with movement of the tumor between the left atrium and the left ventricle. In early systole, the tumor suddenly moved from the left ventricle to the left atrium, and a notch in the rising left ventricular pressure, a prominent c wave, and loud, late elements of the first sound were noted. In early diastole, the tumor moved rapidly through the mitral valve, causing an abrupt diminution in the left atrial volume, thus causing a rapid y descent despite severe obstruction of the mitral valve. An early diastolic sound, thought to be an opening snap, appeared to be related to the checking of the tumor in the left ventricle ("tumor plop").

The unusual left atrial pressure pulse seen in these two cases resembles data from other cases of myxoma reported in the literature, and recognition of these unusual pressures may permit accurate preoperative diagnosis in other patients. Diagnostic changes may be present on apexcardiograms.

Additional Indexing Words:
Myxoma Ball valve thrombus Left heart catheterization Cineangiography Phonocardiography Cardiac surgery Apexcardiography

Myxoma of the left atrium is known to mimic the clinical and hemodynamic features of mitral valvular disease. In some cases, cardiac catheterization has shown a mitral valve gradient, and only at the time of attempted valvotomy has the correct diagnosis of myxoma been made. In these cases where tumor was unsuspected, the operation has carried a high risk. The purpose of this communication is to describe several unique hemodynamic features of left atrial myxoma, based on a correlation of hemodynamic and cineangiographic studies, which may permit the diagnosis to be made at the time of cardiac catheterization.

Report of Cases

Case 1 (K.L.)

A 42-year-old housewife with no past history of heart disease or rheumatic fever was in good health until 6 months prior to admission when she developed increasing dyspnea on exertion, fatigue, and intermittent ankle edema. She was found to be febrile on admission to another hospital, and a cardiac murmur was heard for the first time. She was thought to have subacute bacterial endocarditis, and a course of antibiotic therapy was given. She became afebrile and...
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was transferred to Medizinische Klinik der Universität Kiel for diagnostic studies on August 19, 1965.

On admission, she was not in acute discomfort; her temperature was 37.7 °C, her blood pressure was normal, the pulse rate was 88, and she was in normal sinus rhythm. The jugular venous pressure was slightly elevated, and there was no ankle edema. The cardiac apex was palpated in the fifth left intercostal space at the midclavicular line, and a right ventricular lift was felt at the left sternal border. Auscultation at the apex revealed a loud first heart sound and a very soft holosystolic murmur. An opening snap was present 0.13 second after the second sound and was followed by a short mid-diastolic murmur without presystolic accentuation. The second sound was split normally, and the pulmonary element was accentuated. There were no basal murmurs. The chest was clear to auscultation.

The transverse diameter of the heart was normal on x-ray, but the left atrium appeared to be enlarged with a prominent appendage. The electrocardiogram showed sinus rhythm, right axis deviation, and right ventricular hypertrophy. The hemoglobin was 11.2 g% and the erythrocyte sedimentation rate was 26 mm in 1 hour. The white cell count was 9,600, with a normal differential. Serum protein electrophoresis was within normal limits.

The results of cardiac catheterization are given in table 1. The right ventricular pressure was markedly elevated as was left atrial pressure, obtained by transseptal catheterization (fig. 1A). A left atrial cineangiogram revealed a globular filling defect in the left atrium which entered the left ventricle during each diastolic interval.

Figure 1

Pressures obtained by transseptal left atrial catheterization in cases 1 and 2. The pressure contours are similar and exhibit small a waves, prominent c and v waves, and a rapid fall between v and y ("y descent"). Lead II of the electrocardiogram is displayed at the top of each tracing (LA = left atrial pressure; time lines 0.04 second).

<table>
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<tr>
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At subsequent surgery under cardiopulmonary bypass, Professor B. Lohr removed a tumor from the left atrium which arose from the region of the fossa ovalis and measured approximately 7 cm in diameter. The evening after surgery the patient developed left heart failure with pulmonary edema. She responded readily to therapy. During convalescence, however, she had an episode of bronchopneumonia, again followed by cardiac failure, and she eventually died 3 weeks after surgery.

At postmortem examination the heart weighed 400 g. Both ventricles and the left atrium were dilated. The wall of the left ventricle measured 1.1 cm in thickness. There were only very small pinhead-sized mural thrombi detectable at the former attachment zone of the tumor, close to the foramen ovale. On histological examination, the heart showed evidence of chronic rheumatic myocarditis, with perivascular fibrosis, small scars, and perivascular aggregations of inflammatory cells consisting of histiocytes, lymphocytes, and typical Aschoff bodies. There was no evidence of valvular endocarditis. Emboli, apparently originating from the left atrium, were found within the right brachial artery as well as the left and right femoral arteries. Besides pulmonary edema, widespread bronchopneumonia had developed within both lower lobes. The liver, spleen, and kidneys showed signs of chronic congestion compatible with right-sided heart failure. Death was thought due to heart failure and widespread bronchopneumonia.

Macroscopically, the tumor had a glossy gelatinous appearance, was dark brown-red in color, was extremely friable, and the surface was shiny. On histological examination, the tumor showed a characteristic picture of so-called "myxoma": the base of the tumor was continuous with the connective tissue of the atrial endocardium. The bulk of the tumor consisted of strands and nests of homogenized thrombocytes and fibrin, intermingled with erythrocytes. Part of the thrombotic material was organized, as indicated by a rich vascular supply, capillaries, and medium-sized thin-walled vessels running through the tumor. The surface of the tumor was covered with endothelium. Large areas of the tumor contained macrophages with hemosiderin, fibroblasts, and delicate collagen fibers. The connective tissue within these areas had undergone mucoid changes. There were manystellate cells with long extensions, and the ground substance between the cells was mucoid in appearance.

**Case 2 (D.D.)**

A 24-year-old medical secretary gave no history of rheumatic fever or a cardiac murmur and was in excellent health until 3 months prior to admission. Her first symptom was a nonproductive cough which led her to consult a physician, who heard an opening snap and diastolic rumble typical of mitral stenosis. Mild anemia was present, and a course of oral iron therapy was prescribed. Although she continued to have good exercise tolerance, over the next 10 weeks she had two dramatic episodes of severe breathlessness which occurred while she was supine. The first lasted for 3 hours and resolved spontaneously. The second episode was more severe and led to admission to The John Hopkins Hospital on the evening of January 28, 1966, with frank pulmonary edema, which responded promptly to digitalization and diuretics.

On admission, her blood pressure was 115/60, the pulse rate was 116 and regular, the jugular venous pressure was not elevated, and the apical impulse was palpated in the fifth left intercostal space at the midclavicular line. There was a slight right ventricular lift at the left sternal border. After the pulmonary edema cleared, auscultation at the apex revealed a loud first heart sound and a normally split second sound with accentuation of the pulmonary component. An opening snap was present, followed by a mid-diastolic murmur without presystolic accentuation. There was no systolic murmur, and there were no murmurs at the base. No significant alterations in the auscultatory findings occurred with alteration in the patient's position. The electrocardiogram was normal except for sinus tachycardia. A chest x-ray revealed marked pulmonary venous congestion; this cleared by the time a second x-ray was taken, 48 hours later. The transverse diameter of the heart was normal, but the left atrium was enlarged. A phonocardiogram confirmed the presence of a loud first sound, and the second-sound opening-snap interval was 0.08 to 0.10 second.

Further laboratory data obtained the day after admission revealed a hematocrit of 37% and an erythrocyte sedimentation rate of 24 mm in 1 hour. The white cell count was 8,000, with a normal differential. Serum protein electrophoresis was within normal limits.

Five days after admission, right and left heart cardiac catheterization was performed percutaneously from the femoral vessels (table 1). The pulmonary artery pressure was moderately elevated. Simultaneous transseptal left atrial pressure and retrograde left ventricular pressure revealed an elevated left atrial pressure (fig. 1 right) and a significant diastolic pressure gradient (fig. 2 left). A left atrial cineangiogram showed a

*This pathological description was kindly furnished by Priv. Doz. Dr. Caesar, Pathologisches Institut der Universität Kiel, West Germany.
large globular filling defect in the left atrium that entered the left ventricle with each diastole (fig. 3), establishing the diagnosis of left atrial myxoma. A retrograde left ventricular cineangiogram revealed no mitral incompetence. Two days after catheterization, the patient underwent cardiac surgery under cardiopulmonary bypass. The mitral valve appeared normal on direct inspection. Dr. Vincent Gott excised a left atrial tumor, measuring 3 by 3 by 7 cm, which arose from the region of the foramen ovale by a pedicle 3 mm in length and 1 cm in diameter. The patient's post-operative course was uneventful, and she has remained asymptomatic. She later stated that her current exercise tolerance made her realize that she had actually been limited previously.

The surgical specimen weighed 34 g and consisted of a dark red, soft, smooth, and glistening tumor. On cut section, the mass was dark red and flecked with irregular patches of white material. On histological examination, the surface of the tumor was covered with endothelium, and there were many scattered, large, stellate cells interspersed in the ground substance which had undergone mucoid changes. In addition,
there were a number of fine vascular channels and small areas of hemorrhage.*

**Methods**

A roentgenopaque marking flag, triggered by the R wave of the electrocardiogram permitted correlation of hemodynamic and cineangiographic events by methods previously described.\(^3\) Pressures were recorded through polyethylene cardiac catheters containing an end hole and paired side holes, by means of Statham P-23 DB strain gauges and photographic-oscillographic recorders. The cineangiograms were made at 80 frames per second in case 1 and 60 frames per second in case 2, and permitted timing of events with an accuracy of 12.5 and 16.6 msec, respectively. Contrast medium was injected with a mechanical syringe for cineangiography.

Cardiac output determinations were made by the dye-dilution technique. Chest wall phonocardiograms were obtained during the hemodynamic study and were related to pressure events recorded simultaneously (fig. 2 left).

**Results**

Simultaneous left atrial and ventricular pressures were recorded in case 2, whereas only left atrial pressures were obtained in case 1. Left atrial pressures were elevated in both patients (fig. 1), and a significant pressure gradient was recorded in case 2. In neither case was there a pressure gradient recorded within the atrium.

The left atrial pressure contour was similar in both cases, in that there were a small a wave, prominent c and v waves, and a rapid y descent (fig. 1). These pressures differed from the characteristic left atrial pressure pulse in mitral valve stenosis, which exhibits a tall a wave and a gradual y descent (fig. 2 right).

The simultaneous pressures recorded in case 2 (fig. 2) revealed a prominent notch on the upstroke of the left ventricular pulse, higher than, but synchronous with, the prominent c wave in the left atrium. There was a prominent left atrial v wave, reaching a peak at the time of the second heart sound. The rapid y descent resulted in equilibration of pressures in the left atrium and left ventricle in the first 0.8 second of diastole, but the pressure gradient increased abruptly thereafter and persisted throughout diastole. This early diastolic dip followed by a rise was noted in the left atrial pressure curves from both patients (fig. 1).

Phonocardiograms confirmed the presence of a loud first heart sound and an opening snap (fig. 2 left). In both patients, the Q to first-sound interval was 0.08 second, but the first sound was prolonged by loud vibrations persisting another 0.04 second. These later vibrations occurred at the time of the c wave of the left atrial pressures in both cases and the notch in the left ventricular upstroke of case 2. In case 1, the second-sound opening snap (S2-OS) interval was 0.12 second. In case 2, the S2-OS interval was 0.10 second, and the opening snap was recorded 0.06 second after...

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*This pathological description was kindly furnished by Dr. W. C. Roberts, National Heart Institute, Bethesda, Maryland.
mitral valve opening was indicated by intersection of left atrial and ventricular pressure waves.

The cineangiograms in both cases revealed that the myxoma passed into the ventricle with each diastole and returned to the left atrium with the onset of ventricular systole (fig. 3). During passage through the mitral valve the tumor appeared to occupy most of the circumference of the mitral orifice and actually became deformed as it passed through the valve. The left ventriculogram in case 2 revealed no mitral valvular incompetence, and there was no indication of incompetence in the left atrioogram in case 1.

Correlation of the hemodynamic findings with the cineangiograms in each case revealed that the tumor had profound hemodynamic effects (fig. 4). The tumor obviously obstructed the mitral valve, but tumor movement within the left heart caused abrupt and paradoxical alterations in the pressure in the left atrium and ventricle. The passage of a large portion of the tumor (it was estimated from the surgical specimens that the tumor displaced 110 ml of blood in case 1 and 40 ml in case 2) from atrium to ventricle in early diastole produced a sudden decompression of the left atrium, resulting in a rapid y descent. In both cases, this rapid y descent was present despite angiographic evidence of nearly complete obstruction of the mitral valve orifice as the tumor passed through like a piston or plunger. In mid and late diastole, the left atrial pressure increased, as did the gradient across the mitral valve as the tumor came to rest in the left ventricle. Thus, the initial rapid fall in pressure (y descent) resulted largely from tumor, rather than blood, leaving the atrium. Egress of blood from the left atrium was then blocked by the tumor in the mitral orifice, and the atrial pressure rose in late diastole.

With the onset of the isometric contraction phase of ventricular systole, the tumor abruptly returned to the atrium. The sudden increase in left atrial volume was associated with a prominent c wave in the left atrial pressure pulse. In case 2 there was a notch in the left ventricular pressure pulse at the time of the c wave, apparently resulting from the sudden loss of left ventricular volume and release of tension on the contracting left ventricular muscle fibers as the tumor was suddenly expelled from the ventricular cavity. During systole the tumor remained in the left atrium, and the addition of the mass to the atrial volume undoubtedly contributed to the tall v wave.

The abrupt cessation of the tumor movement from left atrium to left ventricle in early diastole occurred at the time the opening snap was recorded on the phonocardiogram. This auscultatory event occurred after the mitral valve was seen to be fully open on the cineangiograms. The late loud vibrations of the first heart sound occurred after mitral valve closure, at a time when the tumor came to rest in the left atrium.
Correlation of hemodynamic, phonocardiographic and cineangiographic events in case 2. A schematic phonocardiogram has been inserted above high-speed (200 mm/sec) recording of left heart pressures and the motion of the tumor between atrium and ventricle is plotted below. The timing of the three cineangiographic frames shown in figure 3 is indicated, and line drawings of these frames are reproduced. With the onset of left ventricular isometric contraction, the tumor moves through the mitral valve into the left atrium (A) where it remains until the end of ventricular isometric relaxation. As the myxoma leaves the ventricle and moves through the mitral valve (shaded zone on "tumor movement" graph), there is a prominent c wave. In diastole, the myxoma passes through the mitral valve (B) and comes to rest in the left ventricle (C) at the time of the phonocardiographic inscription of the opening snap.

Discussion

The clinical features suggestive of left atrial myxoma* in case 1 were fever, late opening snap, and elevated sedimentation rate. In case 2, the two episodes of pulmonary edema while supine in a patient with apparently unlimited exercise tolerance aroused the suspicion of the House Officer who saw the patient in the Emergency Room, but other physicians (including the authors) who saw the patient thought that the auscultatory findings were typical of rheumatic mitral stenosis, and the diagnosis of myxoma was considered unlikely.

Although no complications resulted from the left heart catheterizations, the transseptal punctures would not have been undertaken in either case if a strong suspicion of myxoma had been entertained. The proximity of the tumor stalk to the usual site of transseptal puncture of the interatrial septum.

*It is not the purpose of this paper to debate the distinction between the so-called "myxoma" and "ball-valve thrombus". The term "myxoma" is used in the generic sense, that is, a predunculated tumor arising from the interatrial septum.
puncture makes this procedure potentially hazardous. In cases of suspected myxoma or left atrial thrombus, it is our usual policy to perform a pulmonary arteriogram and examine the levoangiogram for filling defects before a transseptal puncture is done.

The presence of a loud first heart sound or opening snap, or both, in patients with left atrial myxoma has been well documented in previous reports. The loud late elements of the first sound in our two patients were possibly contributed to by tumor movement, either through checking of the tumor by the stalk or perhaps striking of the atrial wall or mitral valve by the tumor, as has been suggested by Wasserman et al. It has been suggested that the loudness of the first sound is a result of the large atrioventricular pressure difference at the time of ventricular isometric contraction.

The opening snap was relatively late in the present cases and was recorded after the mitral valve was seen to be fully opened on the cineangiograms and pressure records. Abbott suggested that the early diastolic sound resulted from the myxoma hitting the cardiac wall ("tumor plop"). Sudden tensing of the tumor stalk at the end of the excursion of the tumor provides an alternative explanation.

The cause of the two dramatic episodes of left heart failure experienced by the second patient while in the supine position is a subject for speculation. Impaction of the tumor in the mitral orifice seemed unlikely to us, since the tumor was seen to move through and obstruct the mitral orifice with each cardiac cycle during the cineangiogram and caused the patient no discomfort at the time. An alternative explanation for her distress is that the supine position may have permitted the tumor to fall posteriorly and obstruct pulmonary venous inflow into the left atrium.

The unusual pressure phenomena recorded at left heart catheterization (figs. 1 and 2 left) were quite different from those seen in typical rheumatic mitral stenosis (fig. 2 right). A tall v wave and rapid y descent in the left atrial or pulmonary wedge capillary pressures of patients with left atrial myxoma have been previously described. These findings, usually attributed to mitral incompetence, are noted in this condition in the absence of clinical or angiographic evidence of mitral incompetence. A notch on the left ventricular upstroke similar to the one seen in case 2 has been previously demonstrated, although no explanation for the phenomenon was offered. It is assumed that the ventricle, having initiated contraction with a relatively large end-diastolic volume (tumor and blood), undergoes a sudden release of tension before aortic valve opening as the tumor is propelled into the left atrium. This release of tension results in a notch and change in slope of the left ventricular pressure curve. We would postulate that this notch will occur only in those cases in which the tumor passes through the mitral valve. The presence of a notch and change in up-slope on the left ventricular pressure curve, therefore, may be diagnostic of left atrial tumor although their absence does not rule out the diagnosis. Although an apexcardiogram was not obtained in either of our cases, we have found two patients with left atrial tumor in whom a notch was seen on the apexcardiogram, possibly reflecting the hemodynamic event that we have described above (Ongley and associates and personal communication from C. M. Wender and R. B. Logue). The significance of the notch seen on apexcardiography must await further correlative studies.

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