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Left Atrial Myxoma with an Atrial Septal Defect

Report of a case

Daniel T. Anbe, MD,* Edward Arciniegas, MD,** Henry Green, MD*** and Ellet H. Drake, MD*

The diagnosis of myxoma has been difficult in spite of increasing awareness and the utilization of newer diagnostic aids.* The clinical picture can mimic various acquired heart diseases, and even hematologic diseases.* The association of congenital heart diseases with myxomas has rarely been reported. This case is reported in detail because of its unusual features which include an association with an atrial septal defect.

Case Report

DM, a 59-year-old Caucasian housewife was admitted on 5/6/69 after a one-year illness which remained undiagnosed in spite of three admissions to another hospital. Her symptoms began in June, 1968, with weight loss and persistent fever from 101 to 102°F (38.3° to 38.9°C). Positive physical findings were confined to the cardiac examination and consisted of a probable opening snap followed by a short apical diastolic rumble and a Grade III/VI apical systolic murmur. The diastolic murmur was not present continuously. During the initial hospitalization in September, 1968, she was treated with antibiotics after complete evaluation failed to reveal a specific cause of the fever. The following studies were normal or negative: histoplasmin, coccidiodin, first and second strength PPD tests, multiple blood cultures, Coombs test, LDH, stool guaiac, ASO titer, latex fixation, serum calcium, Schilling test, liver scan, IVP and lymphangiography. Initially, penicillin and streptomycin were used for treatment but were discontinued when dermatitis and neuropathy developed. A two-week course of Vancomycin was given without any effect on the fever. Elevated eosinophil counts were
found on several occasions and the antinuclear factor was positive on one occasion and negative on another. The LE preps were negative. She had a hypochromic anemia, with serum iron of 46 mcg and the IBC 324. A subsequent hospitalization was required for an apparent drug reaction several weeks later. For several months afterwards, her temperature was 99° or less and she was relatively well. Pleuritic chest pain associated with blood streaked purulent sputum developed in late March, 1969. Then hospitalized for the third time, she was found to have bilateral pleural effusions and cardiomegaly. Repeated thoracentesis revealed bloody pleural effusions. Recurrent pulmonary embolization was suspected. A lung biopsy was consistent with pulmonary infarction. Keflin therapy did not affect the fever.

After she was transferred to the Henry Ford Hospital on 5/6/69, evaluation revealed an extremely cachectic and toxic patient. Blood pressure measured 110/70, right arm and 88/60, left arm. There were diminished breath sounds and vocal fremitus over both lung
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bases but significant rales were absent. A pleural friction rub was present over the lower lateral aspect of the right chest. The cardiac point of maximum impulse was 8 cm to the left of the midclavicular line over the 6th intercostal space. Cardiac rate was 100 per minute with a regular rhythm. S2P was accentuated but the other heart tones were normal. A gallop sound was not heard. A faint Grade II/VI apical ejection systolic murmur was heard and a Grade II/VI short diastolic rumble and opening snap were noted once. A pericardial friction rub was heard over the lower sternal area. The liver edge was palpable 13 cm below the right costal margin and was tender to palpation. The spleen was not felt. There was 1+ presacral edema and bilateral 3+ pedal edema. The peripheral pulses were normal.

Laboratory studies: Hemoglobin 11.0 gms, hematocrit 40%, WBC 14,700; segmented neutrophils-88%, bands-1%, eosinophils-2%, basophils-1%, monocytes-7%, leukocytoid lymphocytes-1%. Sedimentation rate (Westergren) 29 mm/hr; serum Na-124 mEq/L, K-3.8 mEq/L, Cl-91 mEq/L; CO2 -27 mEq/L; LDH 750 U; total protein-5.8 gm%, albumin-2.28 gm%, alpha-1 globulin-.61 gm%, alpha-2 globulin-.81 gm%, beta globulin-.77 gm%, gamma globulin-1.31 gm%; total bilirubin-1.28 gm; urine culture - numerous enterococci. PO2 -186 mm/Hg (with nasal oxygen); PCO2 -44.1 mm/Hg, pH-7.35; O2 saturation-96.4%; serum osmolality-265 mOsm; urine osmolality-373 mOsm; alkaline phosphatase-9.5 Bodansky units; SGOT 110; serum thyroxin 4.2 mcg% and 2.2 mcg%; serum iron 56 mcg; total iron binding capacity 210 mg; stool guaiac-positive twice and negative once. The following laboratory studies were negative: BUN, serum creatinine, LE prep, pleural fluid culture, sputum culture, ANF, PPD skin test.

Electrocardiogram - sinus tachycardia with a rate of 104. PR 0.17, QRS 0.08, Axis +80: nonspecific ST-T changes consisting of ST depression and T-wave inversion (Figure 1). The admission chest x-ray revealed large bilateral pleural effusions and lower lobe infiltrates (Figures 2A & 2B). The heart shadow was enlarged. A radioactive lung scan showed decreased activity in the left mid-lung field and superior posterior portion of the right lung field.

A right and retrograde left cardiac catheterization with angio-cardiography was performed six days after admission. This showed severe pulmonary hypertension and right heart failure (Table 1). Angiocardiography revealed a large right ventricle. There was considerable enlargement of the main pulmonary artery and its branches, associated with a slow flow of contrast media through

![Figure 2A](image1.png)

![Figure 2B](image2.png)

FIGURES 2A & 2B

PA and lateral x-rays of the chest: Bilateral pleural effusions and lower lobe infiltrates.
TABLE I
CARDIAC CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>LOCATION</th>
<th>PRESSURES mm./Hg</th>
<th>OXYGEN</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Syst/Diast</td>
<td>Mean</td>
</tr>
<tr>
<td>Superior Vena Cava</td>
<td>9</td>
<td>9.0</td>
</tr>
<tr>
<td>Inferior Vena Cava</td>
<td>8</td>
<td>9.7</td>
</tr>
<tr>
<td>Right Atrium</td>
<td>8</td>
<td></td>
</tr>
<tr>
<td>Right Ventricle</td>
<td>88/0-10-10</td>
<td>45</td>
</tr>
<tr>
<td>Main Pulmonary Artery</td>
<td>85/25</td>
<td>47</td>
</tr>
<tr>
<td>Right Main Pulmonary Artery</td>
<td>100/25</td>
<td>21</td>
</tr>
<tr>
<td>Right Inferior Wedge</td>
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<td></td>
</tr>
<tr>
<td>Right Inferior Wedge #2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right Inferior Wedge #3</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Femoral Artery</td>
<td>180/68</td>
<td>90</td>
</tr>
<tr>
<td>Brachial Artery</td>
<td>114/60</td>
<td>76</td>
</tr>
<tr>
<td>Central Aorta</td>
<td>112/0-8-11</td>
<td></td>
</tr>
</tbody>
</table>

**CO. = 4.1 L/Min**  
**CI = 2.4 L/Min/M²**

the basilar segments of the lung. No definite intraluminal defects were appreciated. The left atrium was mildly enlarged, and there was an irregular lobulated filling defect within the atrium measuring up to 7 cm in diameter. A portion of the mass extended through the mitral valve during diastole. The prolapsed portion measured 3 cm in diameter (Figures 3 and 4). There was mild enlargement of the left ventricle, but no angiographic evidence of a left to right intracardiac shunt.

Angioneurotic edema increased and dyspnea and hypotension (80/60) developed shortly after the contrast injection. Intravenous epinephrine was used successfully.

In a surgical operation on 5-15-69, a 10x8x6cm pedunculated, transluscent yellow-green and grape-like mass (Figure 5) was removed. This was attached to the left atrial wall over an area of one square cm between the upper and lower right pulmonary veins. The attachment was quite superficial and there was no actual invasion of the atrial wall. The endocardium at the point of insertion of the tumor was shaved off along with the mass. A 2 cm diameter atrial septal defect was present. This was closed with a running suture of 3-0 silk. The tumor specimen weighed 32 grams.

Specimen sections taken through the base, middle, and peripheral areas of the tumor showed a similar histologic appearance. The bulk of the specimen was a combination of homogenous eosin staining material intermixed with varying amounts of red blood cells, fibrin, and cellular elements. The latter were predominantly tumor cells consisting of single and small groups of uniform polygonal or spindle shaped cells having a moderate amount of eosin staining cytoplasm and uniform nuclei which were devoid of mitoses (Figures 6 & 7). The surface of the tumor often was covered by a single or multilayered tumor cell covering. Scattered histiocytes, some containing hemosiderin, were seen in the stroma. These features are consistent with previously reported myxomas.12,13

Immediately postoperatively, the patient required transfusions, digoxin, glucagon, isuprel and steroids. A tracheostomy was performed on the first postoperative day. The cardiac rhythm varied from sinus rhythm with frequent premature ventricular contractions to transient AV dissociation. Pulmonary congestion and pleural effusions required large amounts of diuretics and a thoracentesis. There was difficulty with fluid and electrolyte imbalance. A right lower lobe abscess developed on the third postoperative week.
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Figure 3
Left atrial and left ventricular visualization in systole: Note the myxoma (lucency, with arrows) in the left atrium.

Figure 4
Left atrial and left ventricular visualization in diastole: Note the myxoma (lucency, with arrows) prolapsing into the left ventricle. The clear arrow indicates the level of the mitral valve.

Figure 5
Gross specimen of the excised tumor. 10x8x6cm.
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There was also a partial collapse of the left lower lobe. The patient's course was further complicated by the development of thrombophlebitis, orthostatic hypotension and iron deficiency anemia. In spite of the stormy postoperative period, the patient recovered fully and remains well five years after the removal of the myxoma.

Discussion

Reports of left atrial myxoma with associated septal defects have been rare. Malm et al\textsuperscript{14} reported a 54-year-old man who was found to have a left atrial myxoma with a large left to right shunt at the atrial level. Their review of the literature at that time (1963) revealed no other report. One of Marpole's\textsuperscript{10} patients had a right atrial myxoma with a 1 cm secundum atrial septal defect with a variable right to left shunt. This shunt was through a foramen ovale which, presumably, was opened by the high right atrial pressure. Coates and Drake reported a case of a right atrial myxoma with a variable right to left shunt.\textsuperscript{16}

Oximetry during cardiac catheterization in this patient did not reveal a left to right shunt even though the pulmonary capillary (wedge) pressure (ie, indirect left atrial pressure) was elevated. A 2 cm diameter atrial defect or patent foramen ovale should be detected by oximetry. It is suggested that this myxoma may have been functioning as a ball valve in the septal defect. The absence of radiographic contrast media in the right atrium during the left atrial visualization supports this.

Figure 6
Low power (35x) section of the tumor. Note the lobulated nature of the tumor.
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Most left atrial myxomas originate on the atrial septum. However, the origin in this case was from the posterior wall.

Greenwood pointed out that it was an odd paradox that pulmonary arterial obstruction was more common in left atrial myxomas. The explanation for this phenomenon may be that many patients with left atrial myxomas are quite ill, have congestive heart failure, and require prolonged bed rest. These conditions predispose to thrombophlebitis and subsequent pulmonary embolism. The clinical course of our patient certainly is compatible with recurrent pulmonary embolism and infarction; however, it is felt that this patient probably had paradoxical embolization from the myxoma across the septal defect. In addition to the aforementioned unusual features, this case illustrates very typical features of left atrial myxoma, ie, transient diastolic murmurs, varied constitutional symptoms and non-specific hematological changes.

References


