Right Atrial Myxoma

By Martin S. Belle, M.D.

Atrial myxomas can be successfully removed.\(^1\)\(^-\)\(^3\) Apparently left atrial myxomas are infrequent and those in the right atrium are approximately 25 per cent as common.\(^6\) Bahnsen and Newman\(^7\) in 1953 reported the first removal of a right atrial myxoma and since that time 4 others have been operated upon. Ripstein\(^8\) in 1953 attempted unsuccessfully to remove a right atrial myxoma through the open right atrium under hypothermia; this tumor extended through the septum into the left atrium.

Recently Coates and Drake\(^5\) reported the successful removal of a right atrial myxoma under open-heart conditions. This patient had a variable right-to-left shunt through a patent foramen ovale. Lyons and his group\(^9\) were unsuccessful in removal of a similar tumor under open-heart surgery. They thought that death was due to a flabby myocardium and the production of total tricuspid insufficiency upon removal of the myxoma, which had previously caused tricuspid stenosis.

Since myxomas of the atria can be successfully removed, it is important to diagnose the lesion correctly so that an otherwise fatal condition may be corrected and the patient restored to health.

Case Report

A 43-year-old white woman gave a history of long-standing nervousness and unexplained infertility. Her diet had been poor in proteins and high in alcohol.

Her childhood history was negative for any stigmata of rheumatic fever but she was first thought to have a heart condition when she first entered school and was restrained from strenuous activities. Nevertheless, she pursued all normal activities and had no reason to see a physician for approximately 35 years.

In April 1955 she developed a swelling of her feet and ankles after acute emotional stress. An enlarged liver and a murmur of the heart were found, and she was told that she had heart failure. After several days of bed rest the edema cleared without drugs.

In July 1955 right heart catheterization was done by Dr. Frank Hernandez of the National Children's Cardiac Home. An intracavitary electrode was used to permit accurate localization of the catheter. A high right atrial pressure was found with a diastolic gradient between the right atrium and ventricle (fig. 1 and table 1). Pulmonary artery and capillary pressure were not elevated even after exercise. The right atrial pressure curve showed no evidence of tricuspid insufficiency. No definite evidence of a left-to-right shunt was found. A slight to moderate degree of oxygen unsaturation of the brachial artery was present which was thought to be due to lung disease or a patent foramen ovale with a right-to-left shunt. Because the oxygen saturation increased on exercise, chronic lung disease was thought more likely to be the cause of the oxygen unsaturation. A diagnosis of tricuspid stenosis was made and the patient sent to an eastern medical center for surgery. There, after careful clinical evaluation, a diagnosis of Ebstein's anomaly was made and she was advised not to have surgery.

When first seen by us on January 31, 1956, the patient appeared nervous, hyperactive, and chronically ill. She complained of extreme weakness and pain in both shoulders. The blood pressure was 90/72, and the pulse was 79 and regular. Cyanosis or clubbing was not evident. The neck veins were hyperactive but not distended and a prominent pulsation was present which was synchronous with a very loud presystolic murmur heard along the left sternal border. No chest deformity was present. A pulsation was noted over a small area to the left of the sternum in the third and fourth intercostal spaces which preceded the apex impulse. The heart was not enlarged to the left, but the right border of dullness extended 3 cm. to the right of the sternum in the third and fourth intercostal spaces.

Auscultation revealed increased and split S₁, but no systolic murmur. A loud presystolic murmur was heard along the left sternal border followed by a third heart sound. At the base there were no murmurs, and both aortic and pulmonary second sounds were diminished. Upon inspiration and with the Valsalva maneuver the presystolic murmur over the xiphisternal area increased. Positional changes had no effect on the murmur.

From the Department of Cardiology, Jackson Memorial Hospital, Mercy Hospital, and the University of Miami Medical School, Miami, Fla.
The lungs were clear and resonant throughout. The liver was felt 2 fingerbreadths below the costal margin with a smooth, nontender edge. There was no ascites or edema.

Blood counts, urinalysis, and serum proteins were normal. The cephalin flocculation test was 1+ and there was 13 per cent retention of bromsulphalein in 45 minutes.

X-ray of the chest (fig. 2) showed a globular cardiac silhouette with suggestive slight generalized enlargement and right atrial enlargement. The pulmonary artery and pulmonary vasculature were diminished. Slight shelving of the right ventricular outflow tract was present, suggesting right ventricular enlargement. Fluoroscopy confirmed these findings and showed no abnormal pulsation in any chamber.

The electrocardiogram (fig. 3) showed low voltage throughout, clockwise rotation, very low potential over the right precordial leads, and relatively prominent P waves. It suggested right atrial hypertrophy and very little right ventricular musculature.

Ventilatory studies showed marked restriction of the maximum breathing capacity and a fairly normal vital capacity.

On February 26, 1956, the patient had a sudden attack of fainting; 30 minutes later she was still semi-stuporous, but conscious and slightly cyanotic. There was no change in the cardiac findings except an irregular sinus rhythm that varied between 80 and 120 beats per minute (fig. 4). Within an hour from the original episode she was completely oriented.
TABLE 1.—Catheterization Data Showing a Small But Definite Diastolic Pressure Gradient between Right Atrium and Ventricle. The Cardiac Output Is Also Considerably Reduced

<table>
<thead>
<tr>
<th>Catheter position</th>
<th>Oxygen</th>
<th>Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Content vol.%</td>
<td>Saturation</td>
</tr>
<tr>
<td>At rest</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Superior vena cava</td>
<td>6.8</td>
<td>44.5</td>
</tr>
<tr>
<td>Inferior vena cava</td>
<td>8.3</td>
<td>54.3</td>
</tr>
<tr>
<td>Right atrium—high</td>
<td>7.0</td>
<td>45.8</td>
</tr>
<tr>
<td></td>
<td>low</td>
<td>8.3</td>
</tr>
<tr>
<td>Right ventricle—mid</td>
<td>7.0</td>
<td>45.8</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>7.5</td>
<td>49.2</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>7.52</td>
<td>49.2</td>
</tr>
<tr>
<td>Pulmonary capillary artery</td>
<td>13.28</td>
<td>86.6</td>
</tr>
<tr>
<td>Brachial artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Exercise</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right atrium</td>
<td>18</td>
<td>6</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>28</td>
<td>4</td>
</tr>
<tr>
<td>Right pulmonary artery</td>
<td>6.52</td>
<td>41.1</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>14.3</td>
<td>90.5</td>
</tr>
</tbody>
</table>

Since we thought that the catheter studies with the intracavitary electrode were against the diagnosis of Ebstein’s anomaly,12,13 angiocardiographic studies were done on March 2, 1956.

The angiocardiogram (fig. 5) showed an irregular filling defect approximately 7.5 by 6 cm.; it was interpreted as being a myxoma attached to the right atrial septum which prolapsed through the tricuspid valve and produced tricuspid stenosis with obstruction of the inflow tract of the right ventricle.

Surgery was undertaken with hypothermia and the patient was cooled to 31 C. The inflow tract was occluded for about 4 minutes. The tumor (fig. 6) was a polypoid pedunculated mass growing from the lower portion of the interatrial septum and obviously obstructing the inflow of blood to the right ventricle. No interatrial septal defect was present. The patient’s immediate postoperative condition was good. She responded somewhat about 6 hours after surgery with a few words but there was no motion of the right side of the body and impaired motion on the left. Her condition gradually deteriorated, shock supervened, and the heart failed. Death occurred approximately 24 hours after surgery.

The pathologic report (fig. 7) of the tumor was consistent with myxoma.

**DISCUSSION**

Of course the prime unequivocal diagnostic tool for atrial myxoma is angiocardiography. This must be done in any case in which a tumor is suggested prior to surgical intervention. However, with the experience in this case and a review of the other right atrial myxomas certain important clinical and physiologic findings may help in deciding when to do angiocardiography. Harvey12 wrote an excellent review of the findings in left atrial myxomas but we think that right atrial myxomas present a few different diagnostic aspects which should be emphasized.

Table 2 summarizes the findings in the reported cases of right atrial myxoma. The presenting symptoms in 3 of the 7 patients was right-sided failure and eventually this was present in a fourth patient. Two patients simulated the historical findings in subacute bacterial endocarditis. One was asymptomatic.

The heart may or may not be enlarged. The enlargement may be both right and left-sided or may be right ventricular and atrial...
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FIG. 3. Electrocardiogram showing low voltage throughout, clockwise rotation, low potential over right precordial leads, and relatively prominent P waves. (This tracing and the one from the patient of Coates and Drake could practically be superimposed.) (Also seen in Ebstein's anomaly.)

only. One patient had intracardiac calcification on the right side. The electrocardiogram presented findings of prominent P waves in the limb leads and right precordium in 3 cases with low voltage of QRS over the right precordium. This is not unlike the electrocardiogram seen in Ebstein's anomaly. Atrial fibrillation may be present.

In our case when the patient had an attack of syncope, the heart rate on the electrocardiogram varied between 72 and 120 (fig. 4) without relation to respiration or change in rhythm, and with no apparent physiologic reason. This fact may suggest intermittent obstruction to flow.

Catheterization may help in deciding that something other than valvular disease is present.
In 2 cases, our own and that of Coates and Drake, a diagnosis of Ebstein's anomaly was made prior to angiocardiology. Coates and Drake could not clearly define the atrioventricular junction. In our case an intracavitary electrode catheter was used, which obviated this difficulty. As shown by Hernandez and co-workers and Yim et al., the diagnosis of Ebstein's anomaly can be made with a greater degree of certainty with the use of an intracavitary electrode catheter along with the pressure curves. Also, as Bahnson (fig. 8) pointed out, the atrial pressure curve may suggest that something other than tricuspid stenosis is present. The atrial curve that he secured showed that the lowest point of pressure occurred just before ventricular contraction, which suggested to him that intermittent obstruction was present.

Two cases showed atrial curves consistent with tricuspid stenosis and insufficiency without symptoms. This alone should lead one to suspect other than valvular disease in spite of an adequate right atrioventricular diastolic gradient.

The atrial pressure (fig. 1) curve in our case also showed its lowest point of pressure in diastole just before atrial contraction, at which time with tricuspid valvular stenosis atrial pressure should be rising rather than decreasing. This should make it imperative that angiocardiography be done.
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Fig. 5 Left. Angiocardiogram showing tumor of the right atrium with prolapse into the right ventricle.

Fig. 6 Right. Photograph of polypoid myxoma which was attached to the lower portion of the interatrial septum on the right.

Fig. 7. Microscopic photograph of myxoma showing irregular and varied-size masses of amorphous, cellular, and slightly granular material.

Finally, if surgical intervention is contemplated open-heart operation with a pump oxygenator and artificially stopped heart seems to be the method of choice. To date, to our knowledge, this method was used in the only successful operation for a right atrial myxoma with long-term survival.

Summary

A case of myxoma of the right atrium with physiologic, surgical, and angiocardiographic findings has been presented together with a review of 6 other published cases.

Several findings have been emphasized that may call attention to the possible diagnosis of right atrial myxoma: right-sided heart failure; intracardiac calcification of the right heart; enlargement of the right ventricle and right atrium; electrocardiographic findings of a prominent P wave and low voltage over right precordium simulating that found in Ebstein's anomaly and marked variation in heart rate not related to any physiologic event or change in rhythm; use of intracavitary electrode catheter to rule out Ebstein's anomaly; and finally, analysis of the right atrial curve may lead one to suspect that other than valvular stenosis is present.

Angiocardiography gives a definitive diagnosis.

Cure may now be obtained with surgical removal of myxomas; therefore, it is important that they be detected during life.
Table 2.—Summary of the Clinical and Physiologic Findings of the Reported Cases of Right Atrial Myxoma

<table>
<thead>
<tr>
<th>Case</th>
<th>Presenting symptoms</th>
<th>Auscultatory findings</th>
<th>X-ray of chest</th>
<th>Electrocardiogram</th>
<th>Catheterization findings</th>
<th>Angiocardiographic findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Bahson and Newman</td>
<td>Right sided heart failure</td>
<td>Short soft systolic murmur and rumbling diastolic murmur along left sternal border</td>
<td>Normal</td>
<td>—</td>
<td>Right atrial curve suggestive of intermittent obstruction</td>
<td>—</td>
</tr>
<tr>
<td>2. Facquet et al.</td>
<td>Asymptomatic</td>
<td>Enlarged heart</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Filling defect right atrium and inflow tract right ventricle</td>
</tr>
<tr>
<td>3. Ripstein</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>4. Coates and Drake</td>
<td>Simulated subacute bacterial endocarditis</td>
<td>Apical and pulmonic systolic murmur with variable 3rd and 4th heart sound</td>
<td>Right sided cardiac enlargement</td>
<td>Prominent P with low voltage QRS</td>
<td>No right atrium right ventricle pressure gradient. No clear cut transition from atrium to ventricle</td>
<td>Lobulated filling defect in right atrium with apparent protusion through tricuspid valve into right ventricle</td>
</tr>
<tr>
<td>5. Lyons et al.</td>
<td>Right sided heart failure</td>
<td>Presystolic murmur loudest at tricuspid area</td>
<td>Enlarged right ventricular contour</td>
<td>Prominent P in II, III, and V leads with QRS in V1 and V5</td>
<td>Right atrial curve suggested tricuspid stenosis and insufficiency (No clinical signs of latter)</td>
<td>Large non-opacified mass in right atrium</td>
</tr>
<tr>
<td>6. Bayer von O. et al.</td>
<td>Fever and enlarged heart followed by right sided failure</td>
<td>Systolic murmur over lower sternal area</td>
<td>Heart enlarged to right and left. Intracardiac calcification right side of heart</td>
<td>Atrial fibrillation</td>
<td>Atrial curve showed tricuspid stenosis and insufficiency (No clinical signs of latter)</td>
<td>Right atrium markedly enlarged with roundish irregular filling defect in the region of A-V junction</td>
</tr>
<tr>
<td>7. Belle</td>
<td>Right sided heart failure</td>
<td>Presystolic murmur at left sternal border with variable 3rd heart sound</td>
<td>Enlarged right atrium and suggestive right ventricular enlargement</td>
<td>Prominent P with low voltage QRS. Marked variation in rate</td>
<td>Right atrial curve showed lowest point in diastole with abrupt rise with atrial contraction</td>
<td>Filling defect 7.5 x 6 cm right atrium which prolapsed through tricuspid valve into right ventricle</td>
</tr>
</tbody>
</table>

**Summario in Interlingua**

Es presentate un caso de myxoma del atrio dextere, con datos physiologic, chirurgic, e angiocardiographic. Ee etiam presente un revista de 6 altere casos in le litteratura.

Plure constatationes es sublineate proque illos suggere de possibile diagnose de myxoma dextero-atrial. Illos es (1) disfallimento cardiae dextero-lateral, (2) calcification intracardiae del corde dextere, (3) allargamento del ventriculo dextere e del atrio dextere, (4) in le electrocardiogramma, prominentia del unda P e basse voltage super le precordio dextere, simile al constatationes in casos del anomalia de Ebstein, e marcate variation del frequentia cardiae, non relateate a ulle evento physiologic o a ulle alteration del rhythmo, (5) le use del catheter a electrode intracavitari pro
excluder the presentia de anomalia de Ebstein, e (6) le analyse del curva dextero-atrial supporta frequentemente le suspension que un condition altere que stenosis valvular es presente.

Angiocardiographia produce le definitive diagnose.

Curation es nune effectuabile per le abalation chirurgic de myxomas. Ergo, le detection de myxoma dextero-atrial durante le vita del paciente es importante.

REFERENCES


Fig. 8. Pressure tracings of case reported by Bahnson showing elevated atrial pressure with the contour of the curve suggesting a valve mechanism which is released between atrial and ventricular contraction. The lowest point on the atrial curve is just before ventricular contraction. ( Courtesy and permission of the authors and Johns Hopkins Press.)
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