Right Ventricular Myxoma Simulating Pulmonic Stenosis

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MYXOMA of the heart is found in 0.02 to 0.1 per cent of autopsies, more frequently in the left than in the right atrium.1-3 Myxoma originating in the ventricles is extremely rare: six cases of left ventricular4-7 and three of right ventricular origin8-10 are recorded in the literature. In addition, two fibromas of the right and three thrombotic pseudotumors of the left ventricle have been described.8,11 The question whether myxomas are fundamentally different from organized and degenerated thrombi12,13 will not be discussed. One left ventricular tumor has been successfully removed with the aid of cardiopulmonary bypass.5

The following is a report of an intracavitary right ventricular myxoma; this appears to be the fourth case described and the first one treated surgically.

Case Report

J.E., a 16-year-old boy had had moderate dyspnea on exertion for a year on his first admission in February 1959. Physical examination revealed normal development and was unremarkable except for the heart. The point of maximal impulse was 1 cm. outside the midsclavicular line. A harsh systolic ejection murmur was audible with maximal intensity in the second left intercostal space. The blood pressure was 110/70 mm. Hg. An electrocardiogram revealed sinus rhythm with evidence of right ventricular hypertrophy. X-ray showed enlargement of the right ventricle and prominence of the pulmonary arch with normal vascular markings. Pulmonary stenosis was presumed but cardiac catheterization was deferred because of possible active infection.

Right heart catheterization performed in August 1959 (table 1) revealed a systolic gradient of 70 mm. Hg across the pulmonary valve with a transitional pressure zone between the pulmonary artery and the right ventricle. Lack of poststenotic dilatation also argued in favor of infundibular stenosis; final confirmation of the diagnosis by angiocardiography was prevented by iodine hypersensitivity of the patient.

Increased dyspnea and low-grade fever led to readmission 14 months later, November 1960. Slight cyanosis, palpatory and auscultatory signs of tricuspid insufficiency, and a systolic thrill accompanying the ejection murmur were registered. A reversed atrial shunt was considered possibly responsible for the cyanosis. Repeated catheterization failed to confirm this assumption; however, the catheter did not enter the pulmonary artery (table 1).

Evidence of congestive heart failure was present at the last admission in January 1961 and was not controlled by strophanthin completely, as shown by the elevated diastolic pressure of the right ventricle at the third cardiac catheterization (table 1). This was performed preliminary to surgical intervention necessitated by the rapid progression of the circulatory failure; in contrast to the former results, the withdrawal curve demonstrated a sharp change of systolic pressure localized at or near the pulmonary valve. Thus the important question about the type of the stenosis, whether valvular or infundibular, was not settled satisfactorily; therefore it was decided to operate with the aid of extracorporeal circulation and hypothermia. Some days prior to operation an attack of exertional syncope with apnea and cardiac arrest occurred.

Surgery was performed with the aid of the KayCross heart-lung machine. The outflow tract of the enlarged right ventricle was opened. A large tumor of myxomatous appearance was found obstructing almost completely the pulmonary orifice, protruding partly into the artery itself. The tumor was excised from its attachments to the ventricular wall and the semilunar valve and was entirely removed; it measured 6 by 4 by 3 cm. The ventricle was closed, and the extracorporeal circulation was discontinued after a total duration of 40 minutes. The patient awoke in the operating room and answered several questions; his mean arterial pressure was 75 mm. Hg, the electrocardiogram revealed atrioventricular dissociation. After 3 hours, sudden cardiac arrest occurred with ventricular fibrillation, which was irreversible in spite of thoracotomy, intubation, cardiac massage, and several trials of electric defibrillation. Extensive myocardial damage was considered re-
sponsible for the conduction disturbance and final asystole.

Microscopically, the tumor consisted of infiltrated connective tissue with regular fibroblasts and star-shaped pale cells (fig. 1). Small foci of ossification and striated muscle were visible in the tumor (fig. 2). The pathologic diagnosis of myxomatous hamartoma was made.

At autopsy, the heart weighed 410 Gm. Every cavity was dilated, especially the right ventricle, the wall of which was 13 mm. thick. The left ventricular wall was 12 mm. thick. The endocardium of the anterior wall of the outflow tract was covered by yellow-gray soft tissue; similar spots were disseminated on the endothelium of the pulmonary artery, immediately over the thickened semilunar cusps.

Discussion

Our patient offered the unmistakable signs of pulmonic stenosis; the etiology was revealed only by open-heart surgery. The clinical picture of right ventricular myxoma is rather unexplored. The physical examination, electrocardiogram, x-ray, and autopsy findings of the last published case\(^3\) are in marked agreement with our own; pulmonic stenosis may be considered a possible manifestation of this tumor. From the diagnostic point of view, three features seem to differentiate these cases from other congenital anomalies: (1) attacks of syncope, (2) unexplicable changes of the hemodynamic findings, (3) a rapid and irreversible worsening course.

Table 1

<table>
<thead>
<tr>
<th>Hamodynamic Findings of Three Right Heart Catheterizations</th>
<th>August 13, 1959</th>
<th>December 14, 1960</th>
<th>February 17, 1961</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium: Pressures (mm. Hg)</td>
<td>8/2 (5)</td>
<td>20/8 (14)</td>
<td>15/8 (11)</td>
</tr>
<tr>
<td>(O_2) content (vol. %)</td>
<td>11.3</td>
<td>8.6</td>
<td>10.4</td>
</tr>
<tr>
<td>Saturation (%)</td>
<td>64</td>
<td>42</td>
<td>55</td>
</tr>
<tr>
<td>Right ventricle: Pressures (mm. Hg)</td>
<td>100/0</td>
<td>100/5</td>
<td>110/10</td>
</tr>
<tr>
<td>(O_2) content (vol. %)</td>
<td>10.5</td>
<td>8.6</td>
<td>(\ldots)</td>
</tr>
<tr>
<td>Saturation (%)</td>
<td>59</td>
<td>42</td>
<td>(\ldots)</td>
</tr>
<tr>
<td>Pulmonary artery: Pressures (mm. Hg)</td>
<td>30/8 (19)</td>
<td>(\ldots)</td>
<td>18/4 (11)</td>
</tr>
<tr>
<td>(O_2) content (vol. %)</td>
<td>12.1</td>
<td>(\ldots)</td>
<td>(\ldots)</td>
</tr>
<tr>
<td>Saturation (%)</td>
<td>68</td>
<td>(\ldots)</td>
<td>(\ldots)</td>
</tr>
<tr>
<td>Femoral artery: Pressures (mm. Hg)</td>
<td>110/70 (83)</td>
<td>110/75 (87)</td>
<td>130/80 (97)</td>
</tr>
<tr>
<td>(O_2) content (vol. %)</td>
<td>17.2</td>
<td>16.8</td>
<td>17.1</td>
</tr>
<tr>
<td>Saturation (%)</td>
<td>97</td>
<td>82</td>
<td>93</td>
</tr>
</tbody>
</table>

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RIGHT VENTRICULAR MYXOMA

In Kishimoto and Sakaibori's case, numerous attacks of syncope at rest were observed; our patient had a single effort syncope of Adams-Stokes type in the last days of his life. Although the occurrence of fainting attacks precipitated especially by change of position is a well-known sign of intracavitary heart tumors, one episode of cardiac arrest cannot be considered pathognomonic; rheumatic fever or congenital heart block far more frequently cause Adams-Stokes attacks in young people. Loss of consciousness is rarely if ever observed, however, in cases of pulmonic stenosis and may be a useful clue pointing to the diagnosis of myxomatous obstruction.

The varying pattern of the withdrawal curve, i.e., intermediate pressure zone at the first catheterization and sharp transition from arterial to ventricular pressure at the third catheterization, reflects positional changes of the obstructing tumor. Rapid progression to irreversible heart failure is more characteristic of primary heart tumor of any kind than the generally well-tolerated pulmonary stenosis of adolescents. Angiocardiography is to be considered the most efficient diagnostic tool.

Summary

The fourth case of right ventricular myxoma is reported, the first one in which the tumor was removed surgically. The clinical picture was that of pulmonic stenosis with some unusual features: effort syncope, changing hemodynamic pattern, and a rapidly progressive clinical course of irreversible congestive heart failure. The importance of early diagnosis, the angiocardiographic findings, and the necessity of operation before development of serious myocardial damage are stressed.

References


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