Cardiac Myxomas with Systemic Embolization

Review of the Literature and Report of a Case

By Jay Silverman, M.D., John S. Olwin, M.D., and John S. Graettinger, M.D.

The purpose of this paper is to emphasize the occurrence of systemic embolism as a sign, often the presenting one, of cardiac myxomas. The literature is reviewed and a new case reported.

Primary cardiac tumors are rare. They have been found in 0.05 per cent of autopsied material; secondary metastatic cardiac tumors occur in 0.6 per cent of cases. The antemortem diagnosis of primary cardiac tumors is rarer still, but with the advent of angiocardiography and adequate techniques for their surgical removal, the consideration of this diagnosis has become critical. The most common primary tumor is the myxoma. This basic tumor has a broad spectrum of appearance, namely, fibromyxoma, myxohemangioma, myxohemangiendothelioma, etc. The myxoma accounts for 50 per cent of all primary tumors. Approximately 75 per cent of these occur in the left atrium, usually in the region of the rim of the fossa ovalis. Approximately 20 per cent occur in the right atrium; the rest may arise on a valve leaflet, from the ventricular endocardium, or from the wall of the pulmonary artery.

Cardiac myxomas have been reported in the age group of the newborn to 70 years. The greatest incidence is between 30 and 60 years. Thirty-two cases demonstrating embolization have been previously reported. Of these, 29 had their site of origin in the left atrium, one in the right atrium, one in the left ventricle, and one in the root of the pulmonary artery. The tumor arising in the root of the pulmonary artery was a fibromyxosarcoma. The remainder were myxomas. The sites of embolization are listed in tabular form in table 1.

Case Report

The patient was a 33-year-old Negro waitress who, 8 months prior to admission, developed a sudden onset of left-sided hemiplegia. She was admitted to another hospital where the diagnoses of rheumatic heart disease with mitral stenosis, presumed paroxysmal atrial fibrillation, and embolization to the right middle cerebral artery were made. Her hospital course and clinic follow-up were uneventful. The salient physical findings during her clinic visits were a residual left hemiparesis, normal sinus rhythm, and a mid-diastolic apical thrill and rumbling murmur. There was no evidence of decompression, and the patient was given no therapy. There was no past history of rheumatic fever.

On the day prior to admission, the patient noted sudden tingling and lightheadedness which lasted 5 minutes. Two hours later she suddenly lost consciousness and remained so for 30 minutes. There were no convulsive movements, tongue biting, or incontinence. Incoherent speech lasted several hours after this episode, and a frontal headache persisted to the time of admission.

On admission, the patient's temperature was 99.0 F., pulse 92 and regular, blood pressure 105/75. There was a residual left hemiparesis. The heart was not enlarged. There was a grade-V rumbling mid- and presystolic murmur and thrill at the apex, which radiated to the axilla. The first heart sound was accentuated at the apex. The second heart sound was loudest in the second right intercostal space. No third sound was evident. The remainder of the physical examination was not remarkable.

On admission, urinalysis, hemoglobin, leukocyte count and differential were normal. Blood cultures were negative. Fluoroscopy revealed moderate enlargement of the left atrium and outflow tract of the right ventricle. An electrocardiogram was within normal limits.

On the fourth hospital day, a grade-II, blowing, apical, pansystolic murmur appeared without change in the diastolic murmur. On the fifth day, the patient complained of coldness in her legs. The apical diastolic murmur had disappeared and
the systolic murmur had become very loud. The femoral pulses were weak, as were the popliteal, left posterior tibial, and dorsalis pedis pulses. Both legs were cool, the right more than the left, and a rather sharp line of temperature change occurred just above the right popliteal fossa. The patient was taken to surgery with a diagnosis of saddle embolus of the aortic bifurcation from a detached cardiac thrombus or rupture of a mitral cusp.

At operation, a mass of gelatinous, multiloculated, homogeneous, yellow embolus was removed from the bifurcation of the aorta. No clot was present above or below the embolus. A tentative diagnosis of a detached myxoma was made. The patient's blood pressure was unobtainable on several occasions during the procedure despite blood replacement and vasopressor agents.

Immediately postoperatively, the patient developed a transient right bundle-branch block and myocardial ischemia. The left leg had good pulsations throughout, whereas mummification devel-

Table 1

<table>
<thead>
<tr>
<th>Author</th>
<th>Reference number</th>
<th>Tumor</th>
<th>Origin</th>
<th>Eye</th>
<th>Brain</th>
<th>Coronary</th>
<th>Spleen</th>
<th>Kidney</th>
<th>Abdominal aorta</th>
<th>Iliofemoral or popliteal</th>
<th>Lung</th>
<th>Index factor</th>
</tr>
</thead>
<tbody>
<tr>
<td>Edwards</td>
<td>5</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Block</td>
<td>6</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Brewin</td>
<td>7</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Paquet</td>
<td>8</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ringertz</td>
<td>9</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Carter</td>
<td>10</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Manschot</td>
<td>11</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>+</td>
<td>+</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Reichling</td>
<td>12</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gleason</td>
<td>13</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>+</td>
<td>+</td>
<td>$+$</td>
<td>$+$</td>
<td>$+$</td>
<td>$+$</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Russell</td>
<td>14</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gorlitzer</td>
<td>15</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Kalbfleisch</td>
<td>16</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zuimeda</td>
<td>17</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zuimeda</td>
<td>17</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Silverman</td>
<td>17a</td>
<td>Myxohemangiendothelioma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Brown</td>
<td>18</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>?*</td>
<td>?*</td>
<td>$+$</td>
<td>?*</td>
<td>?*</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Differding</td>
<td>19</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Skause</td>
<td>20</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scannell</td>
<td>21</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Horlick</td>
<td>22</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Mills</td>
<td>23</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Thompson</td>
<td>24</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Beanlands</td>
<td>25</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Weinstein</td>
<td>26</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Gerbode</td>
<td>27</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>?§</td>
<td>?§</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Towers</td>
<td>28</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>?*</td>
<td>?*</td>
<td>$+$</td>
<td>?*</td>
<td>?*</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chin</td>
<td>29</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td>?§</td>
<td>?§</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Harvey</td>
<td>30</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td>$+$</td>
<td>$+$</td>
<td>$+$</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Castlemman</td>
<td>31</td>
<td>Myxoma</td>
<td>L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bland</td>
<td>32</td>
<td>Myxoma</td>
<td>F.L. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chiari</td>
<td>33</td>
<td>Myxoma</td>
<td>R. atrium</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Young</td>
<td>34</td>
<td>Myxoma</td>
<td>L. ventricle</td>
<td>?*</td>
<td>?*</td>
<td></td>
<td></td>
<td></td>
<td>$+$</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Haythorn</td>
<td>3</td>
<td>Fibromyxosarcoma</td>
<td>Root of pulm. artery</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Infarcts of these organs cited. No mention of tumor cells.
†Infarcts of these organs cited. Emboli found in these areas did not contain clear-cut neoplastic tissue.
‡History of seizures. Brain examination not performed.
§Emboli cited. No emboli tissue available for examination.
CARDIAC MYXOMAS

Morphologic Findings
At autopsy, a small tumor pedicle was found in the left atrium, arising from the posterior margin of the atrial septum (figure 1). Tumor emboli and infarcts were found in the liver, spleen, left kidney, and right middle cerebral artery. The histology of these peripheral emboli, the aortic embolus removed at surgery (figure 2) and the tumor mass in the left atrium (figures 3 and 4), was identical, and was that of a myxohemangiendothelioma: the residual left atrial tumor jutted out from a thickened endocardium. A portion of its surface was covered by flattened endothelial cells. At its point of origin from the endocardium there was a considerable degree of necrosis, hemorrhage, and round-cell infiltration. The nodule itself consisted of a myxomatous stroma in which can be seen scattered plasma cells and giant cells.

**Figure 1**
*Photomicrograph of left atrial myxoma, arising from the posterior margin of the atrial septum.*

**Figure 2**
*Photomicrograph of saddle embolus.*

**Figure 3**
*Photomicrograph of left atrial myxohemangiendothelioma.*

**Figure 4**
*Photomicrograph of left atrial myxohemangiendothelioma.*
The nuclei of the latter were round to slightly oval in shape. They were surrounded by an ill-defined, pale eosinophilic cytoplasm. In some fields they appeared to surround thin, slit-like spaces. Also present within the myxomatous stroma were small collections of red cells, occasional hemosiderin-laden macrophages, and moderate numbers of lymphocytes.

Summary

A case of left atrial myxohemangioendothelioma, diagnosed ante mortem by removing a tumor embolus to the bifurcation of the aorta, is reported. The patient had been misdiagnosed as having had rheumatic heart disease with paroxysmal atrial fibrillation and embolization to the right middle cerebral artery. In retrospect, because of the characteristics of the second heart sound and absence of an opening snap, a rheumatic etiology should have been questioned.

The literature regarding embolization of atrial myxomas has been reviewed.

References


The nature of our mind leads us to seek the essence or the why of things. Thus we aim beyond the goal that it is given us to reach; for experience soon teaches us that we cannot get beyond the how, i.e., beyond the immediate cause or the necessary conditions of phenomena. In this respect the limits of our knowledge are the same in biological as in physico-chemical sciences.

When, by successive analyses, we find the immediate cause determining the circumstances in which a phenomenon presents itself, we reach a scientific goal beyond which we cannot pass. When we know that water, with all its properties, results from combining oxygen and hydrogen in certain proportions, we know everything we can know about it; and that corresponds to the how and not to the why of things. We know how water can be made; but why does the combination of one volume of oxygen with two volumes of hydrogen produce water? We have no idea. In medicine it is equally absurd to concern one's self with the question "why." Yet physicians ask it often. It was probably to make fun of this tendency, which results from lack of the sense of limits to our learning, that Molière put the following answer into the mouth of his candidate for the medical degree. Asked why opium puts people to sleep, he answered: "Quia est in eo virtus dormitiva, eujus est natura sensus assoupire." This answer seems ludicrous and absurd; yet no other answer could be made. In the same way, if we wished to answer the question: "Why does hydrogen, in combining with oxygen, produce water?" We should have to answer: "Because hydrogen has the quality of being able to beget water." Only the question "why," then, is really absurd, because it necessarily involves a naive or ridiculous answer. So we had better recognize that we do not know; and that the limits of our knowledge are precisely here.—Claude Bernard, An Introduction to the Study of Experimental Medicine. New York, The Macmillan Company, 1927, p. 80.
Cardiac Myxomas with Systemic Embolization: Review of the Literature and Report of a Case
JAY SILVERMAN, JOHN S. OLWIN and JOHN S. GRAETTINGER

Circulation. 1962;26:99-103
doi: 10.1161/01.CIR.26.1.99
Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1962 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/26/1/99

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/