Cardiac myxoma metastasizing to the brain

Case report

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A 55-year-old man, who had previously had a left atrial myxoma excised, developed recurrent hematomas of the left occipital lobe. Microscopic examination revealed the presence of metastatic myxoma.

KEY WORDS • cardiac myxoma • metastasis • brain neoplasm

Cardiac myxomas are the most common primary tumors affecting the heart and are thought to be derived from the multipotential mesenchymal cells of the subendocardium. There has been some controversy concerning the nature of these lesions. A number of investigators have considered them to originate from organizing mural thrombi but the prevalent opinion is that they are true neoplasms. There are well-documented cases of local recurrence and invasion, invasion of chest wall, and distant metastases. In the brain, occlusion of cerebral vessels by myxoma emboli with resultant aneurysmal dilatation has been reported. In our literature review, there have been only two cases of atrial myxoma metastasizing to the central nervous system, one having been reported three times. We report a case of an atrial myxoma producing metastatic lesions in the brain following total excision of the cardiac tumor.

Case Report

This 54-year-old male laborer presented to the neurosurgical service in July, 1987, for evaluation of blurred vision. Four months previously, he had undergone total excision of a left atrial myxoma in another hospital. Details of that hospital admission were not made available to us.

Examination. On examination, the patient was fully conscious with a normal cardiovascular system and no limb deficits. A right homonymous hemianopia and impaired visual acuity (20/50 bilaterally) were confirmed. Fundi were normal. A computerized tomography (CT) scan of the brain showed a left hyperdense space-occupying lesion in the occipital region (Fig. 1 left).

Operations. Craniotomy and debulking of the lesion suggested that this was an intracerebral hematoma rather than a tumor. Multiple biopsies of the cavity were negative for tumor. The patient made an expeditious recovery with a diagnosis of spontaneous intracerebral hematoma. Six months later, the patient experienced a right-sided focal epileptic seizure and persistent right leg weakness; however, he was still mobile with assistance. A CT scan showed a space-occupying lesion (Fig. 1 center) similar to the one demonstrated in the previous scan. Left occipital lobectomy was performed and a globular tumor 2 cm in diameter was included in the resection specimen. The patient was discharged home after 1 month of intensive rehabilitation.

One year after his second cranial operation, the patient was admitted with a 2-week history of progressive right hemiparesis and aphasia. A CT scan revealed a large tumor anterior to the left occipital lobectomy cavity with gross peritumoral edema (Fig. 1 right). Treatment with steroids improved his deficits dramatically. Complete excision of the solid tumor was achieved; no postoperative radiotherapy was given. Three months postoperatively, the patient was mobile on a walking frame. He was finally discharged to a nursing home, moderately disabled.

Pathological Examination. Histological materials from the original cardiac tumor were obtained; these
showed the typical appearance of an atrial myxoma, with no sarcomatous elements present. Tissues from the first craniotomy showed only old and recent hemorrhages; no metastatic tumor tissue was seen. In the specimen from the second operation, a discrete, well-circumscribed nodule with a gelatinous consistency and measuring about 2 cm in diameter could be identified mixed with semiorganized blood clots (Fig. 2 left). Microscopic examination of both the second and the third operative specimens revealed myxoma tissues admixed with recent and old hemorrhages, hemosiderin deposits, and fibrosis. Tumor tissue consisted of islands of stellate and polygonal cells with scant eosinophilic cytoplasm (Fig. 2 right). These cells were scattered in a matrix rich in acid mucopolysaccharides, which stained for Alcian blue at pH 2.5, and formed vessel-like channels that simulated the appearance of primitive capillaries. Islands of tumor cells also infiltrated haphazardly into the dense fibrous tissue, which apparently was derived from organization of old hemorrhages. Extramedullary hematopoiesis was not present and no sarcomatous elements could be identified.

Discussion

Cardiac myxomas are regarded by most authorities as genuine neoplastic lesions. They often present with the classic triad of embolic manifestations; obstructive symptoms with cardiac failure; and systemic disorders including fever, weight loss, and malaise. Al-
Cardiac myxoma metastatic to brain

### References


Table 1: Clinical summary of patients with atrial myxoma metastasizing to the brain

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Location of Metastasis</th>
<th>Presentation</th>
<th>Interval: Primary Resection to Metastasis</th>
<th>Outcome</th>
<th>Other Metastases</th>
</tr>
</thead>
<tbody>
<tr>
<td>DeSousa, et al., 1978</td>
<td>44, F</td>
<td>choroid plexus</td>
<td>headaches, previous &quot;strokes&quot;</td>
<td>8 yrs</td>
<td>survived 6 more years</td>
<td>multiple bone lesions</td>
</tr>
<tr>
<td>Rankin &amp; DeSousa, 1978</td>
<td>52, F</td>
<td>parietal lobe, dura, &amp; cerebellum</td>
<td>incidental autopsy finding</td>
<td>primary not resected</td>
<td>not relevant</td>
<td>none at autopsy</td>
</tr>
<tr>
<td>Budzilovich, et al., 1979</td>
<td>55, M</td>
<td>occipital lobe</td>
<td>recurrent cerebral hematomas</td>
<td>1 yr</td>
<td>moderately disabled 2 years after initial recurrence</td>
<td>none</td>
</tr>
<tr>
<td>Ng &amp; Poon, 1990</td>
<td>44, F</td>
<td>choroid plexus</td>
<td>headaches, previous &quot;strokes&quot;</td>
<td>8 yrs</td>
<td>survived 6 more years</td>
<td>multiple bone lesions</td>
</tr>
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</table>

Though most patients are cured by adequate excision, rare cases have been documented of recurrence, local invasion, and metastases, to lung, long bones, skull, and skin. Recently, cytogenetic studies revealed chromosomally abnormal clones from cultured myxoma cells, giving further support to the neoplastic nature of these tumors. However, the histological specimen has not always been carefully reviewed to exclude the presence of a primary cardiac sarcoma, which may undergo myxoid degeneration and assume the appearance of myxoma on cursory examination. In McAulister and Fenoglio, in their monograph on cardiac tumors, reported that they had never seen an example of a truly malignant myxoma in cases where the histology was reviewed. Not surprisingly, the histology of malignant cardiac myxoma often revealed sarcomatous tissues in the original tumors.

Although it is not uncommon for embolic lesions from a cardiac myxoma to occur in the brain, only two previous cases of metastatic tumors have been recorded in the central nervous system (Table 1). Careful review of the literature shows that the first case has been reported three times. It concerned a 44-year-old woman who initially had recurrent cerebrovascular accidents thought to be due to embolic phenomena from a cardiac myxoma. Ultimately, she had a mass lesion of metastatic myxoma resected from the choroid plexus 8 years later. She survived 6 more years at which time an indolent metastatic bone lesion in the scapula was removed. The second case was that of a 52-year-old woman who died of an apparent stroke. An autopsy revealed metastatic hemorrhagic nodules in the cerebrum and cerebellum from a cardiac myxoma. In both instances, review of the histology showed the typical appearance of a cardiac myxoma.

This case differs from previously reported cases of metastases from atrial myxomas in that the metastatic lesions developed after a relatively short interval following primary excision of the cardiac tumor. Long intervals have been documented before metastases developed in the brain or other sites. In spite of multiple cerebral recurrences, the tumor in our case appeared to be only of low-grade malignancy. Seo, et al., reported a case illustrating that repeated tumor recurrence is not incompatible with prolonged survival. The indolent nature of the metastases apparently applies to other metastatic sites as well. The neurological presentation of our patient was not unlike that of the previously reported two cases in that the patient had a stroke-like syndrome both clinically and radiologically. In the second case, metastasis from cardiac myxoma was only established by microscopic examination at autopsy. In the first operation of our patient, the hemorrhage evacuated did not yield any definite myxoma tissue. Although the hemorrhage might represent an embolic manifestation of cardiac myxoma, the rapidly recurrent nature of the lesion and the identification of tumor tissue from subsequent operative specimens make it more likely that hemorrhage had occurred in the first instance as a result of tumor erosion into blood vessels.

The mechanism of tumor metastases in this case is speculative. In general, tumor emboli from cardiac myxoma are noninvasive in metastatic sites. They survive locally as occluding luminal masses for varying unpredictable durations. Tumor tissues may occasionally grow into the walls of the vessels which subsequently undergo aneurysmal dilatation. In some cases, the surviving tumor in the embolic site focally disrupts the internal elastic lamina. Similar tumor erosion into pulmonary arteries has also been reported by Heath and Mackinnon. It is conceivable that, in our case, embolic myxoma tissue invaded the vessel walls, providing a nidus for the cerebral hemorrhage and subsequent growth of extravascular metastatic tumor tissue. However, this must be regarded as a rare phenomenon, as the majority of emboli from cardiac myxomas do not result in metastasis.

### References


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