Multiple Cerebral Aneurysms and Brain Metastasis from 
Primary Cardiac Myxosarcoma: 
A Case Report and Literature Review

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Primary neoplasms of the heart are rare. Malignant cardiac myxoma, or so-called myxosarcoma, accounts for about 6% of primary malignant cardiac tumors. Cerebral metastasis of malignant cardiac myxoma is extremely rare; only three cases have been reported and two of them included an autopsy study. The case described herein is the first reported brain metastasis combined with multiple cerebral aneurysms originating from primary cardiac myxosarcoma. The true incidence of cerebral myxomatous aneurysm is unknown and the pathogenesis of myxomatous aneurysm formation has not been fully defined. The current hypothesis favors that tumor materials from cardiac myxomas embolize into the vasa vaso-rum of the peripheral arteries and subsequently prolifere in the vessel wall. This then leads to a weakening of subintimal tissue, such as the internal elastic lamina, with subsequent aneurysm formation. The prognosis of cardiac myxosarcoma is very poor. Although malignancies most likely develop from the mesenchymal cells, they are difficult to treat with any modality (operation, chemotherapy, radiotherapy or transplantation) because these tumors have usually undergone extensive spread by the time the diagnosis is made. (Chang Gung Med J 2011;34:315-9)

Key words: myxosarcoma, brain metastasis, aneurysm

A 33-year-old female aboriginal inhabitant of Taiwan was admitted in May 2006 with a 2-month history of intermittent productive cough, palpitation and dyspnea. The echocardiography revealed multiple tumors in the left atrium, measuring 2.9 x 3.0 cm, 2.0 x 3.0 cm, and 4.3 x 2.3 cm, respectively, at their greatest dimensions. Tumor excision was performed
through a Y sternotomy together with a cardiopulmonary bypass with bicaval and aortic cannulation. Microscopic examination revealed spindle cells set within a myxoid ground substance with transition to pleomorphic and hyperchromatic cells arranged in solid sheets. Immunohistochemical studies showed positive staining for smooth muscle actin and HHF-35, and focally for desmin. Stainings for myoglobin, myogenin, myo D1, S100, CD31, CD34, and calretinin were negative. The diagnosis was consistent with myxosarcoma (Fig. 1). Postoperatively, the patient received radiotherapy (4500 cGy/25 fractions) and adjuvant chemotherapy.

One year following the excision of the left atrial myxosarcoma, she presented at our emergency room with a two-week history of headache, dizziness, and mild weakness of the left upper extremity. Enhanced brain CT showed multiple mass-like lesions in the left frontoparietal, left occipital and right high parietal regions. Magnetic resonance imaging (MRI) and angiography (MRA) of the brain, which were performed 3 days after admission, revealed multiple intracranial tumors and aneurysms (Fig. 2). Unfortunately, the patient had a sudden onset of consciousness deterioration upon being sent for conventional cerebral angiography following brain MRI and MRA. Brain MRI was repeated immediately and showed a large hematoma in the left frontoparietal area, which was probably related to aneurysmal rupture of a left distal middle cerebral artery (MCA). An emergent left frontotemporoparietal craniectomy for removal of the hematoma was performed; an encapsulated yellowish solid tumor, measuring 20 x 15 x 10 millimeters in size, was also found intraoperatively. The left distal MCA aneurysm was left in place. Pathologically, the tumor showed brain tissues infiltrated with neoplastic spindle cells that were similar to those of the cardiac tumor. The immunohistochemical staining results were also the same. A postoperative non-enhanced brain CT revealed complete removal of the left frontoparietal hematoma, but brain swelling was present. Unfortunately the patient expired one week after admission due to brain stem failure.

**DISCUSSION**

The incidence of primary cardiac tumors was reported to be 0.0017%-0.33% in autopsy series. Thirty percent of all cardiac sarcomas have evidence of distant metastasis early in their courses, but brain metastasis is quite uncommon. To our knowledge, there have been only three previously reported cases of cerebral metastasis of primary cardiac myxosarcoma (Table 1). The case described herein is the first reported brain metastasis combined with multiple cerebral aneurysms originating from primary cardiac myxosarcoma.

The true incidence of cerebral myxomatous aneurysm is unknown and the pathogenesis of myxomatous aneurysm formation has not been fully defined. Stoane et al. were the first to describe the

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**Table 1. Summary of Cases of Primary Cardiac Myxosarcoma Metastasizing to Brain**

<table>
<thead>
<tr>
<th>Authors and Year</th>
<th>Age, sex</th>
<th>Interval to neurological symptom onset</th>
<th>Other metastatic sites</th>
<th>Surgery for brain lesions</th>
<th>Radiotherapy for cardiac myxosarcoma</th>
<th>Chemotherapy for cardiac myxosarcoma</th>
<th>Outcome</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kayama, 1978</td>
<td>32 y/o, F</td>
<td>2 months</td>
<td>–</td>
<td>+</td>
<td>–</td>
<td>–</td>
<td>DOD</td>
<td>4</td>
</tr>
<tr>
<td>Mahar, 1979</td>
<td>29 m/o, F</td>
<td>3 months</td>
<td>Multiple*</td>
<td>–</td>
<td>–</td>
<td>+</td>
<td>DOD</td>
<td>5</td>
</tr>
<tr>
<td>Roh, 2001</td>
<td>21 y/o, M</td>
<td>7 months</td>
<td>Lung, Skeletal muscle</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>AWD</td>
<td>6</td>
</tr>
<tr>
<td>Present case</td>
<td>33 y/o, F</td>
<td>12 months</td>
<td>–</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>DOD</td>
<td>This report</td>
</tr>
</tbody>
</table>

**Abbreviations:**  M: male; F: female; y/o: year-old; m/o: month-old; –: no; +: yes; DOD: dead of disease; AWD: alive with disease; *: lungs, ribs, thymus, liver, kidneys, and adrenal glands.
characteristic cerebral angiogram in two patients with left atrial myxomas.\(^{12}\) It was thought that embolization of large vessels leads to aneurysmal formation at the site of the occlusion.\(^{12,13}\) The current hypothesis favors that tumor materials from cardiac myxomas embolize into the vasa vasorum of the peripheral arteries and subsequently proliferate in the vessel wall, leading to a weakening of subintimal tissue, such as the internal elastic lamina, with subsequent aneurysm formation.\(^{11-16}\)

Pseudoaneurysms, or false aneurysms, are differentiated from true aneurysms by the involved layers of the artery. Pseudoaneurysms are the result of the rupture of all three layers of the artery: the tunica intima, media, and adventitia. The aneurysm is contained by an organized hematoma or neighboring connective tissue. With true aneurysms, the outer-

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**Fig. 1** The tumor contains an area similar to myxoma (A, H & E, X 13.2) with a transition to nodularly cellular (B, H & E, X 13.2) and diffusely cellular (C, H & E, X 13.2) areas. Higher magnification reveals pleomorphic and hyperchromatic nuclei with frequent mitotic activity and occasional multinucleation (D, E and F, H & E, X 66). Immunohistochemical staining shows diffuse positivity against smooth muscle actin (G, X 66), HHF-35 (H, X 66), and focally desmin (I, X 66).

**Fig. 2** Magnetic resonance angiography of the brain demonstrated multiple aneurysms (arrows).
most layer, the adventitia, remains intact. Rapid treatment is imperative because pseudoaneurysms generally grow faster and have a greater chance of rebleeding compared with true aneurysms.\textsuperscript{(17)}

The prognosis of cardiac myxosarcoma is very poor, with a mean survival of 10 months after operation, according to Basso et al.\textsuperscript{(18)} Although cardiac malignancies most likely develops from the mesenchymal cells, they are difficult to treat with any modality (operation, chemotherapy, radiotherapy or transplantation), because these tumors have usually undergone extensive spread by the time the diagnosis is made.\textsuperscript{(19)}

Dousbeck et al. reported that adjuvant chemotherapy and/or radiation therapy did not prevent local recurrence or visceral metastasis in a study of 24 primary cardiac sarcomas.\textsuperscript{(20)} However, Roh et al.\textsuperscript{(6)} considered that adjuvant chemotherapy is probably helpful, because one of their patient with left atrial myxosarcoma with widespread systemic metastasis was alive at 37 months after initial diagnosis. The exact evaluation of the clinical course and treatment modality of patients with cerebral metastasis has not been established due to the very small number of reported cases in cardiac myxosarcoma.

**Conclusion**

Cardiac myxosarcoma is exceptionally rare and is a distinct disease entity. There is no well established treatment for cerebral metastasis caused by cardiac myxosarcoma. Cardiac myxosarcoma patients who have cardiac surgery to remove the primary cardiac tumor should be followed up for neurological symptoms as well as for evidence of cardiac recurrence. The diagnosis of cerebral metastasis and myxomatous aneurysms should be considered in patients with delayed neurological symptoms or signs.

**REFERENCES**

原發性心臟黏液肉瘤的顱內轉移合併顱內多發性動脈瘤：
病例報告暨文獻回顧

李宗翰 黃純真 蘇宗明 楊嘉寅 鋒政玄

心臓的原發腫瘤相當時少。惡性黏液瘤，或稱作黏液肉瘤，約只占心臓原發惡性腫瘤的百分之六。原發性心臓黏液肉瘤的顱內轉移更少，目前文獻上只有三個病例報告，其中兩個病例是死後病理剖解發現。本篇病例報告則是第一個發表原發性心臓黏液肉瘤的顱內轉移合併顱內多發性動脈瘤。顱內黏液性動脈瘤的真正發生率仍是未知，而且黏液性動脈瘤的形成機轉尚未清楚。目前的假說傾向是因為心臓黏液瘤的腫瘤細胞在周遭的動脈栓塞並進而生長，導致血管內膜下的組織變得更脆弱，因此形成動脈瘤。原發性心臓黏液瘤的預後相當差，雖然黏液瘤部分是由間質細胞發展而來，但仍然很難以任何一種方法 (手術切除、化學治療、放射線治療或心臟移植) 來治療，因為通常在診斷出之前，腫瘤細胞就已經廣泛地擴散。

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關鍵詞：惡性黏液瘤，顱內轉移，動脈瘤

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