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Case Report

Multiple craniotomies for the resection of symptomatic multifocal intracranial metastatic cardiac myxoma: A case report

V. Peter Abdow III[1], Jeffrey M. Breton[2], Vikram V. Nayar[2]

[1]School of Medicine, Georgetown University, [2]Department of Neurosurgery, MedStar Georgetown University Hospital, Washington, DC, United States.

E-mail: *V. Peter Abdow III - vpa3@georgetown.edu; Jeffrey M. Breton - vpa3@georgetown.edu; Vikram V. Nayar - vpa3@georgetown.edu

***Corresponding author:**
V. Peter Abdow III,
School of Medicine,
Georgetown University,
Washington, DC, United States.

vpa3@georgetown.edu

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ABSTRACT

Background: Myxomas, rare benign mesenchymal lesions, are the most common cardiac tumors. Patients may rarely develop hematogenous metastasis to the brain, which can present as new-onset neurological deficits that correlate with multifocal hemorrhagic lesions on imaging. Limited guidelines presently exist for the treatment of such lesions. This report outlines a unique case involving three craniotomies and failed radiation therapy in the treatment of metastatic cardiac myxoma.

Case Description: A 63-year-old woman presented with a right middle cerebral artery embolic stroke secondary to a left atrial myxoma and multifocal hemorrhagic lesions consistent with intracranial metastasis. She had a right frontal craniotomy for tumor resection, followed by stereotactic radiosurgery, though this did not arrest disease progression. She later had a left occipital craniotomy for a symptomatic lesion. More than two years after her initial presentation, she returned with acute-onset symptoms correlating to growth in a left frontal lesion requiring another resection. Following this third craniotomy, imaging has not revealed the progression of metastatic intracranial disease. She is pursuing further treatment through primary cardiac tumor resection.

Conclusion: Although rare, hematogenous seeding with subsequent formation of hemorrhagic metastasis is a possible complication of atrial myxoma. While surgical resection, radiation therapy, and chemotherapy have historically been used, no standard of care currently exists. This case demonstrates repeat tumor resection as effective for managing symptomatic intracranial metastatic myxoma in a patient with poor response to radiation therapy and multiple recurrences, with follow-up showing improvement in neurological symptoms and mass effect and absence of recurrence on imaging.

Keywords: Case report, Craniotomy, Metastatic myxoma, Recurrent myxoma

INTRODUCTION

Myxomas, rare benign lesions believed to derive from multipotent mesenchymal cells, are the most common cardiac tumors. Current estimates suggest that about 75% of myxomas develop in the left atrium, putting patients at greater risk for systemic thromboembolic events.[6] In addition, patients may rarely develop hematogenous metastasis to the brain. This may present as new-onset neurological deficits in a patient with known myxomatous disease, and multifocal hemorrhagic infarcts may be seen on imaging.[3] While surgical resection is the treatment of choice for atrial myxoma, limited data and treatment guidelines exist for intracranial myxomatous lesions.[8]

This report outlines a detailed description of a unique case involving a patient requiring three craniotomies for myxoma resection for symptomatic lesions after radiation therapy failed to halt disease progression.

CASE DESCRIPTION

A 63-year-old woman initially presented with acute onset of left-sided facial droop, numbness, and weakness concerning for right middle cerebral artery stroke. Computed tomography (CT) of the head was unremarkable, and CT angiography (CTA) showed occlusion of the distal right M1 segment. She received tissue plasminogen activator (tPA) and was transferred to our facility for endovascular thrombectomy with thrombolysis in cerebral infarction (TICI) 2a recanalization. The etiology was determined to be thromboembolic and a cardiac workup revealed a large left atrial mass attached to the interatrial septum, consistent with the left atrial myxoma. This was later confirmed histopathologically following urgent surgical resection of the myxoma and repair of an associated atrial septal defect.

Eight months later, she presented again after a fall and possible seizure. CT showed the right frontal intracranial hemorrhage (ICH) in her prior stroke bed, as well as small left frontal and occipital ICHs. Magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) confirmed multifocal hemorrhagic lesions in these areas concerning hemorrhagic metastatic lesions [Figure 1]. She underwent right frontal craniotomy for tumor resection and postoperative pathology revealed myxoid neoplasm, consistent with intracranial metastasis. She was also found on cardiac MRI to have a new right ventricular mass at that time.

She underwent stereotactic radiosurgery (SRS) to the postoperative right frontal region and left frontal and occipital lesions, though this did not arrest disease progression. She did not undergo chemotherapy given a paucity of data to guide oncological recommendations. Thirteen months after her initial presentation, she experienced blurry vision and a right visual field defect, with associated interval growth in the left occipital lesion and significant vasogenic edema, for which she underwent a left occipital craniotomy. One year later, she presented with delays in speech, cognition, and movement and was found to have interval growth in the left frontal lesion [Figure 2]. She was initially scheduled to have the right ventricular cardiac mass resected, though given these symptomatic lesions and the requirement for anticoagulation during cardiac surgery, this intervention was postponed. She underwent left frontal craniotomy, and pathology revealed radiation necrosis. From a neurologic perspective, she tolerated surgery well: she remained fully oriented, she was without cranial nerve deficits, her speech was mildly slowed without aphasia, and she followed commands symmetrically and with full strength.

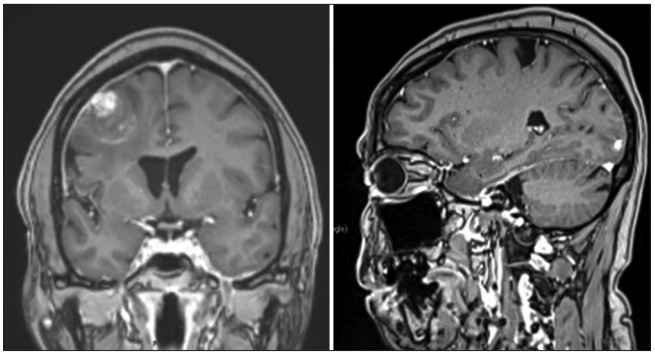

Figure 1: T1-weighted magnetic resonance imaging, post contrast, revealing multiple enhancing lesions concerning multifocal hemorrhagic metastases, including right frontal (left image, coronal view) and left occipital (right image, sagittal view).

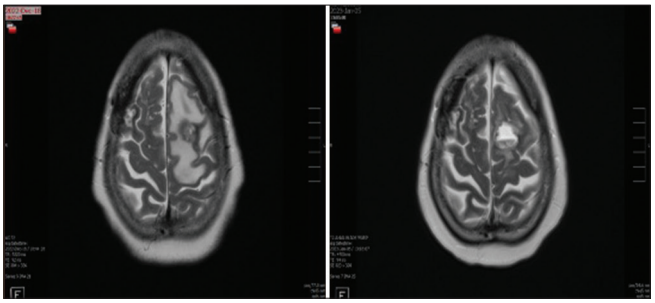

Figure 2: Axial T2-weighted magnetic resonance imaging demonstrating preoperative (left) and postoperative (right) views of a left frontal lesion with significant associated vasogenic cerebral edema, which improved after surgical resection.

MRI 4 months postoperatively showed no signs of disease. The right ventricular mass has been stable on repeat imaging and she has remained hemodynamically stable. Cardiac workup has not revealed cardiac or pulmonary right-to-left shunting, making paradoxical embolization less likely as an etiology for new metastatic lesions. She awaits further evaluation for repeat cardiac surgery.

CONCLUSION

Although rarely encountered, hematogenous seeding with subsequent formation of hemorrhagic metastasis is a possible complication of atrial myxoma, a classically benign lesion that should not be ignored. The potential for myxomatous embolism is thought to be due to the tumor's friable and villous qualities, as well as its tendency to degenerate, either through fibrosis, necrosis, thrombosis, calcification, or Gamna–Gandy body formation.[4,7] Tumor cells seeded within the embolus then proliferate to form a metastatic lesion.[7] Patient presentation may vary depending on the location of the metastasis, but typically resembles hemorrhagic infarction of the affected area.[3] It is usually recommended to remove the primary cardiac lesion before brain metastases unless there are urgent indications for surgery, such as hemorrhage with significant

mass effect or progressive neurologic deficits. In one review, myxoma metastasis to the brain occurred from two months to eight years following resection of the cardiac mass.[4] In patients who present with signs of myxoma metastasis, a transesophageal echocardiogram is often recommended to investigate the presence of a primary lesion.[4] Due to the rarity of metastatic myxomatous lesions, data describing their treatment is limited, and there are no established standards of care.

Surgical resection can be considered in patients with relatively few metastases, unstable conditions, or progressive neurologic deficits.[8] Radiation therapy has also been used in patients with multiple metastases, usually whole-brain radiation at 25–30 Gy.[5] Postoperative chemotherapy with either ifosfamide and doxorubicin or doxorubicin alone has also been attempted in patients with multifocal disease, though with mixed results.[7] One case report even documents the use of Gamma Knife radiosurgery to treat cerebral metastases, without recurrence of disease 2 years postoperatively.[4] Despite these interventions, recurrence of disease is common, even after surgical resection of the primary atrial tumor.[1,2] In this case, the patient required three separate craniotomies for tumor resection due to stroke-like symptoms caused by her intracranial metastases. She was not recommended to receive chemotherapy, and SRS was not successful in reducing the size of her multifocal lesions. Instead, she responded favorably to en bloc removal of each myxomatous lesion, with the improvement of cerebral edema and neurological symptoms postoperatively. This patient has already experienced recurrence of her primary cardiac lesion and it is possible that she may experience recurrence of intracranial lesions, as well. Given this risk, she will require longitudinal follow-up for the neurosurgical and cardiac dimensions of this complex diagnosis. Ultimately, her goal is to undergo surgical resection of the primary cardiac tumor, for which she has been cleared given follow-up imaging without significant intracranial disease.

In summary, this unique case report demonstrates repeat tumor resection as an effective method for managing symptomatic intracranial metastatic myxoma in a patient with poor response to radiation therapy and multiple recurrences, with follow-up showing improvement in neurological symptoms and mass effect secondary to her intracranial lesions, as well as the absence of recurrence on imaging.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The author(s) confirms that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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