

Case Reports

Sequelae and Surgical Management of Giant Cardiac Myxoma

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Abstract

Cardiac myxomas are a rare phenomenon within the general population, and although there are reports of them, giant myxomas are not common in the medical literature. This report presents a case of a giant left atrial cardiac myxoma in a 57-year-old female patient who was largely asymptomatic until she presented with a diffuse thromboembolic stroke. This case report highlights the importance of surgical management of cardiac myxomas and discusses the difference in open vs minimally invasive surgical resection of giant cardiac myxomas.

Keywords: Myxoma; stroke; minimally invasive surgical procedures

Case Report

Presentation and Physical Examination

A 57-year-old female patient was transferred from a smaller peripheral hospital for the workup of a suspected stroke. Her symptoms began the afternoon before she was transferred, when her friend noticed that she suddenly began to lean toward the left and had left-sided weakness.

The patient was then taken to an outside hospital's emergency department, where an initial computed tomographic scan of her head revealed no acute abnormalities. Her National Institutes of Health Stroke Scale (NIHSS) score was 12 at the time, though, and positive for partial lateral gaze paresis, flaccid left upper and lower extremities, limb ataxia in both left extremities, right partial facial paralysis, and dysarthria. Given her NIHSS score at this time, she was subsequently given intravenous tissue plasminogen activator for strong suspicion of acute stroke. Computed tomographic angiographic imaging of the head and neck was obtained but did not show evidence of large vessel occlusion. The patient was then transferred from this outside hospital to the reporting institution's neurological intensive care unit for further stroke workup and management.

In the neurological intensive care unit, a physical exam revealed that the patient was in no acute distress. She was alert and oriented to person, place, time, and event and able to follow instructions, but she did have mild dysarthria. Cranial nerve assessment found that she had a mild right gaze preference, but she overcame midline and right facial droop. Motor exam revealed 4/5 strength in her left upper extremity and left lower extremity, with 5/5 strength in her right upper extremity and right lower extremity. Sensation was intact bilaterally at all levels. Coordination was found to be intact through a finger-to-nose test and a heel-to-shin test. Her repeat NIHSS score was 4. The rest of her physical examination was within normal limits. An initial electrocardiogram was obtained, showing normal sinus rhythm with right atrial enlargement, right ventricular hypertrophy, and right axis deviation. Initial magnetic resonance imaging of the head was performed that demonstrated diffusion-weighted imaging hyperintensities involving the right thalamus, bilateral cerebellar hemispheres, left parietal lobe, right frontal and right parietal lobes,

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and bilateral temporal lobes that were concerning for acute thromboembolic disease (Fig. 1A, Fig. 1B). Per the patient's stroke workup, a 2-dimensional echocardiogram with Doppler study was obtained that showed evidence of a severely enlarged left atrium, with a large, heterogeneous, dense, mobile 4.0×9.0 -cm mass with a wide base attachment to the interatrial septum that occupied most of the left atrium and prolapsed into the left ventricle (Fig. 2, Fig. 3).

Medical History

The patient's medical history was notable for hypertension without current use of antihypertensives and a transient ischemic attack 5 years previously.

Differential Diagnosis

The differential diagnosis for a pedunculated cardiac mass includes cardiac myxoma, thrombus (both infectious and sterile), metastasis, and primary malignant cardiac tumor.

Technique

While the patient was in the neurological intensive care unit, the neurology team performed neurologic checks every hour. Both the cardiology and cardiothoracic surgery teams were consulted for management of myxoma. Per cardiothoracic surgery recommendations, open atrial myxoma surgical resection was performed on day 6 of hospitalization. During surgery, a 9.0×5.0

Key Points

- Cardiac myxomas are extremely rare, especially in asymptomatic patients.
- Myxomas may be a medical emergency if they cause recurrent strokes or mitral valve inlet obstruction.
- Surgical management is imperative for treatment, but no guideline recommendations regarding traditional open surgery vs minimally invasive techniques for giant myxoma resections exist.

Abbreviation

NIHSS, National Institutes of Health Stroke Scale.

$\times 2.4$ -cm tan-red, papilliferous, lobulated mass with an attached $2.5 \times 1.5 \times 0.3$ -cm portion of endocardium and myocardium and a stalk arising from the level of the foramen ovale was resected en bloc and submitted in formalin for pathologic review. Laboratory studies and cultures revealed no white blood cells, organisms (on Gram stain), anaerobes, fungi, or acid-fast bacilli involvement. On pathology review, the mass was shown to comprise several "myxoma cells," with bland, elongated nuclei and eosinophilic cytoplasm within a myxoid and hemorrhagic background. Hemosiderin-laden macrophages were also seen, but there was no evidence of nuclear atypia, mitotic figures, or infiltrative features. Thick-walled muscular arteries with smooth muscle hypertrophy were noted at the stalk of the mass. Cardiomyocytes were noted to be located within

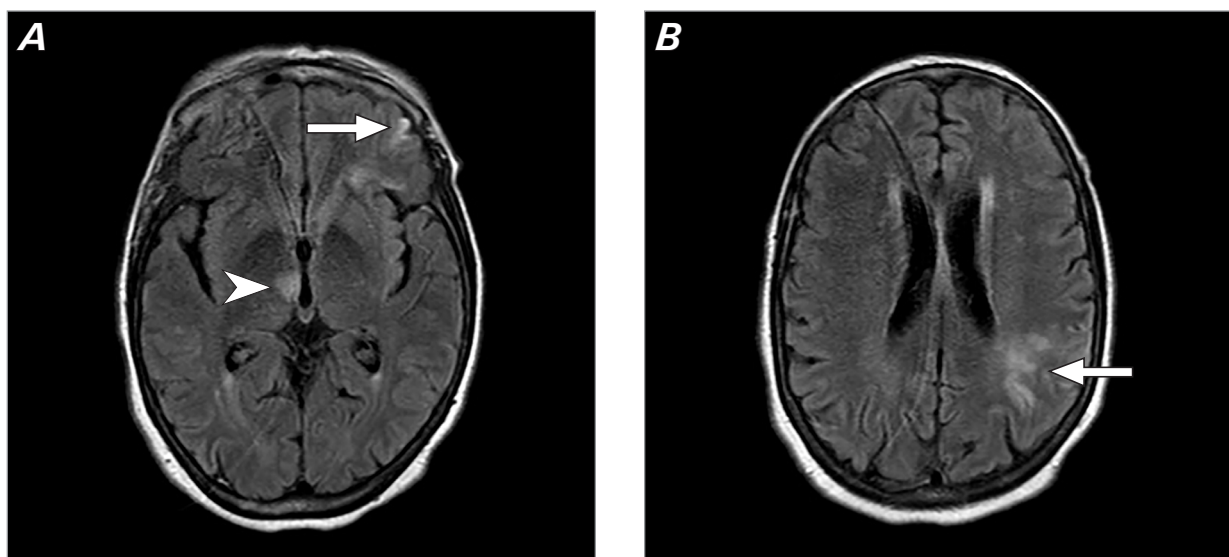


Fig. 1 Diffusion-weighted magnetic resonance imaging in the axial plane without contrast reveals hyperintensities in the (A) right thalamus (arrowhead) and left frontal lobe (arrow) and (B) left parietal lobe (arrow).

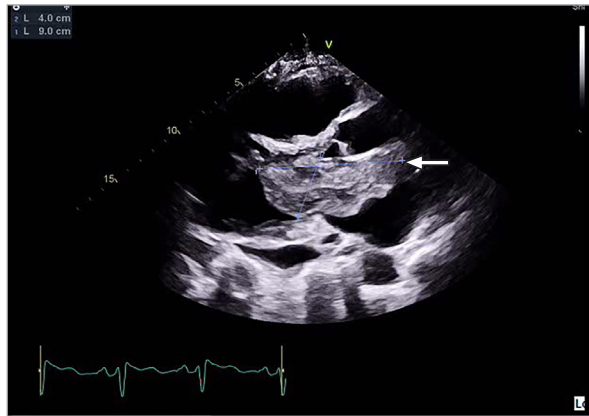


Fig. 2 A 2-dimensional transthoracic echocardiogram provides a parasternal long-axis view of the patient's myxoma. The arrow shows the myxoma protruding from the mitral valve. The myxoma measured 4.0 × 9.0 cm.

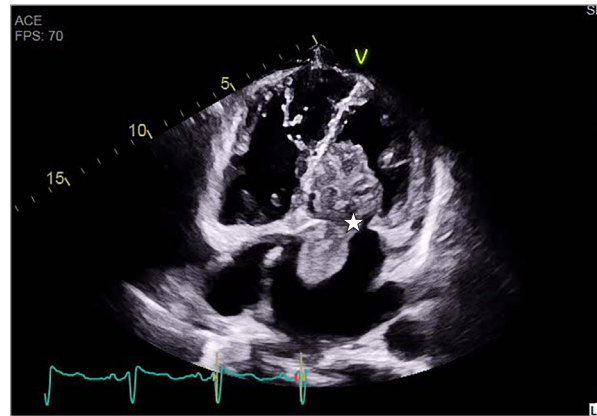


Fig. 3 A 2-dimensional transthoracic echocardiogram provides a subxiphoid view of the patient's myxoma. The star indicates the myxoma's site of attachment to the interatrial septum. The myxoma measured 4.0 × 9.0 cm.

0.1 cm of the resection margin, which is consistent with a negative margin for tumor cells.

Outcome

The conclusion of the patient's clinical syndrome was explained by the left atrial myxoma serving as a source of her cardioembolic stroke at presentation to the outside hospital. Cardiology and cardiothoracic surgery were immediately consulted for management and removal of left atrial myxoma to decrease stroke burden and improve symptoms. The patient tolerated the procedure well and recovered completely from surgery with improved symptoms, except for experiencing intermittent atrial fibrillation with rapid ventricular response after surgery, likely due to cardiac irritation during the procedure.

Latest Follow-Up

This patient was eventually discharged home on oral anticoagulation (apixaban 5 mg twice daily) for at least 3 months, aspirin 81 mg daily, metoprolol 50 mg twice daily, atorvastatin 20 mg daily, and colchicine 0.6 mg twice daily, with outpatient follow-up appointments scheduled with cardiology, cardiothoracic surgery, and neurology.

Discussion

Primary malignancies of the heart are rare in the general population, with metastasis of distant tumors being much more common.¹ Benign tumors are by far the

most common type and include cardiac myxomas, fibromas, lipomatous tumors, fibroelastic papillomas, and hamartomas. Primary malignant tumors are much rarer, but among these malignant tumors, sarcomas such as angiosarcomas or leiomyosarcomas are the most common.² Of all the primary tumors listed in this section, cardiac myxomas are by far the most prevalent.^{1,2} They consist of many cells encased in a mucopolysaccharide stroma. The cells originate from a multipotent mesenchyme capable of differentiating into neuronal or endothelial tissue.³ The incidence of these myxomas is extremely low, with an incidence rate of 0.0017% to 0.19% found on autopsies. In the general population, the reported prevalence is only 0.03%.⁴ Certain genetic syndromes, such as Carney complex, exist that are associated with an increased incidence of myxomas as well as multichambered myxomas.⁴ When cardiac myxomas do occur, they most commonly do so alone, without other tumors of the heart, and only extremely rarely do they undergo malignant transformation and metastasis.⁴

Diagnosis typically depends on cardiac imaging, such as echocardiography, followed by pathologic confirmation after mass removal.¹ Echocardiographic features such as size, movement characteristics, and attachment point can help differentiate cardiac tumors from other causes of cardiac masses, such as thrombosis and vegetation.¹ Most myxomas are in the left atrium and present with 1 or more symptoms from the myxoma triad: mitral stenosis or obstruction, peripheral embolism, and systemic symptoms. One study from France found that approximately two-thirds of patients experienced mitral valve obstruction, one-third experienced embolization,

and one-third experienced systemic symptoms.⁵ The patient whose case is reported here falls only into the second category, presenting with embolization but without mitral valve obstruction or other systemic symptoms. Of note, the French study also found that two-thirds of patients presented with auscultatory changes on exam, but the current patient presented with no such findings. Myxomas on gross examination fall into 2 broad categories: (1) friable and villous or (2) smooth. Myxomas that cause embolization, such as in the current patient, are more likely to be friable or villous.

Prior case reports have identified other patients with asymptomatic cardiac myxomas, but this phenomenon is the minority among patients with myxomas. One report found 20 prior cases of asymptomatic presentation in patients with cardiac myxoma.⁶ Among patients who were asymptomatic, smaller tumors were more likely, but even these small tumors may show symptoms of the myxoma triad.⁶ One prior case highlights a completely asymptomatic myxoma that measured approximately 4 × 4 cm, but myxomas have been reported to range from 1 cm to 15 cm in size.⁵ Given this range, an asymptomatic presentation in the current patient, who had a tumor measuring 9 cm, is unexpected. The clear consensus for treatment is surgical removal, and most patients have excellent outcomes, with rates of recurrence as low as 3%.⁶ There is debate as to the feasibility of minimally invasive surgery vs the traditional open approach. The decision-making process must consider the size and burden of the tumor on the patient, but reports of the minimally invasive approach are promising and highlight a lower complication rate and shorter length of stay compared with the traditional open approach.⁶ More research must be done to uncover the true benefits of this approach on a larger scale.

In conclusion, the case of an incidentally discovered cardiac myxoma following an acute cerebrovascular attack is presented. The patient whose case was reported was largely asymptomatic before this inciting stroke, having only hypertension and prior transient ischemic attacks, which in retrospect may have been attributable to the myxoma but were not attributed to it at the time of diagnosis. Because the patient had had no prior cardiac imaging, the myxoma was not discovered until it was of a massive size. This case highlights the necessity of surgical management of these myxomas and the possible complications that may arise even in patients who lack clear prior symptoms.

Article Information

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