ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.surgicalneurologyint.com

Surgical Neurology International
Editor-in-Chief: Nancy E. Epstein, MD, NYU Winthrop Hospital, Mineola, NY, USA.
SNI: Neuro-Oncology
Editor
Mitsutoshi Nakada, MD
Kanazawa University, Ishikawa, Japan

SNI
Open Access

Case Report

Cardiac phoenix in the brain-occult intracranial hemorrhagic metastases from completely resected atrial myxoma

Sagar Amrutlal Ghodasara[1], Rohit Balasubramanian[1], Shriram Varadharajan[2], P. S. Shobhanaa[3]

Departments of [1]Neurosurgery, [2]Neuro-Imaging and Stroke Interventions, [3]Neuropathology, Kovai Medical Center and Hospitals, Coimbatore, Tamil Nadu, India.

E-mail: *Sagar Amrutlal Ghodasara - sagarghoda88@gmail.com; Rohit Balasubramanian - rohitstanley@yahoo.com; Shriram Varadharajan - shri86@gmail.com; P. S. Shobhanaa - shobhanaa.ps@gmail.com

***Corresponding author:**
Sagar Amrutlal Ghodasara,
Department of Neurosurgery,
Kovai Medical Center and
Hospitals, Coimbatore,
Tamil Nadu, India.

sagarghoda88@gmail.com

Received : 06 July 2020
Accepted : 22 October 2020
Published : 11 November 2020

DOI:
10.25259/SNI_410_2020

Quick Response Code:

ABSTRACT

Background: Cardiac myxomas are sporadic in nature and can often recur with a frequency of 3%, especially in middle-aged women, and 22% of the cases account to a part of Carney complex. Complete surgical removal of the myxoma is usually curative. Recurrence has been related with partial surgical excision, multicentricity, and embolism of tumor fragments.

Case Description: We report a case of a patient with single brain metastases due to tumor embolization, from a cardiac myxoma operated prior. This case is exclusive, as tumor embolization from atrial myxoma to the cerebral cortex can be possible, within a short duration. In our case, the patient was evaluated with a magnetic resonance imaging brain and a solitary hemorrhagic lesion in the eloquent cerebral cortex was observed. To determine the primary etiology, the diagnosis of probable metastases was thought of, and a thorough workup was planned. Surprisingly, no primary lesion was detected, and as a histological diagnosis was required, he underwent a navigation-guided excisional biopsy of lesion. The biopsy was indicative of a metastatic deposit from an atrial myxoma.

Conclusion: In eloquent cortex lesions, gross total resection is challenging for a neurosurgeon especially when the patient has no significant neurological deficits. Timely gross total resection of a solitary metastatic lesion can improve the patient's outcome and can enhance early recovery with less or no morbidity.

Keywords: Cerebral metastasis, Eloquent visual cortex lesion, Excisional biopsy, Left atrial myxoma, Magnetic resonance imaging, Positron emission tomography

INTRODUCTION

Cardiac myxoma is the most common benign heart tumor, frequently found in the left atrium in about 86% of the cases.[12,15] Recent evidence has revealed that cardiac myxomas are slowly proliferating benign neoplasms. Complete resection of cardiac myxoma and its cardiac appendages can cure a cardiac myxoma, but its incomplete resection, can result in multifocal tumors and embolisms due to its recurrence. Independent cerebral metastatic growth, intraparenchymal hemorrhages, and oncotic aneurysms are uncommon neurological sequelae of atrial myxomas.[6] Ischemic strokes are usually the most common neurologic

presentation, followed by syncope, psychiatric symptoms, and headache.[4] The existence of its malignant counterpart is controversial; however, it's recurrence after excision has been reported.[10] Recurrence has been attributed to incomplete excision, multifocality, and embolism of tumor fragments.[7] Systemic embolism from the left atrial myxoma is particularly frequent, and the brain is the most common metastatic site for cardiac myxomas (approx.45%).[13] Due to the persistent risk of brain metastases and aneurysm formation, early diagnosis and intervention is desirable. Long-term follow-up of the patients with atrial myxoma even after complete surgical excision is mandatory and a thorough explanation of the potential risks of recurrence of the cardiac tumor must be explained to the patient, along with the possibility of late development of cerebral lesions. Literature has reported very few such cases. We report this rare case, to highlight weird pathological course of this disease and importance of neurosurgical decision-making in tackling cerebral metastatic part.

CASE HISTORY

A 63-year-old male was admitted with chief complaints of moderate left-sided hemicranial headache for 1 month which was on and off in nature and not associated with any aura. The headache progressively increased over 7 days and was associated with vomiting. He presented with an unsteady gait along with giddiness. He underwent an excision of a left atrial myxoma, a year ago and was on regular follow-up with an uneventful cardiac evaluation. A cardiologist's evaluation with a 2D ECHO showed no residual lesion in heart. A magnetic resonance imaging brain [Figure 1] was done which was suggestive of a left parieto-occipital multicystic hemorrhagic SOL with the potential of being a hemorrhagic metastasis or a cavernoma. The positron emission tomography revealed a mildly hypermetabolic enhancing lesion in the left parieto-occipital area with no active metabolic lesion elsewhere in the body.

He underwent left parieto-occipital craniotomy with navigation-guided excision of the lesion. Intraoperatively, there was a yellowish-gray soft lesion (6.0 × 4.4 × 5.9 cm) extending up to the left occipital horn of lateral ventricle with moderate vascularity. There were 2–3 satellite lesions observed in the hemorrhagic and necrosed tumor bed. Postoperatively, he improved well neurologically, with an improvement of gait imbalance.

HPE report [Figure 2] was suggestive of a myxomatous lesion with extensive hemorrhage and secondary degenerative changes consistent with myxoma metastasis. At present, he has been discharged and is on regular follow-up and has been advised radiological evaluation at regular intervals.

Neuroimaging

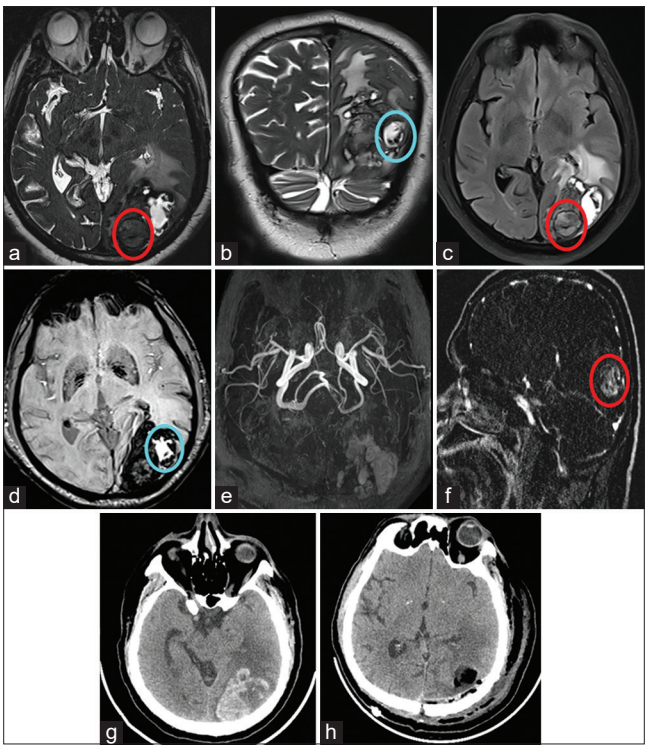

Figure 1: Top row – Axial and coronal T2-weighted (a and b) images show large heterogeneous hemorrhagic lesion in the left occipital region with perilesional edema and mass effect. Differential soft tissue is noted along posterior aspect (red circle) on T2 and FLAIR (c) with hypointensity and blooming on SWI (d) as compared to adjacent hematoma (blue circle) showing hyperintensity with peripheral blooming. Bottom row – Axial TOF MRA images (e) show no obvious vascular abnormality and postcontrast sagittal subtracted T1 MPRAGE images (f) show intense enhancement within differential soft tissue (red circle on f). Preoperative plain CT (g) shows hemorrhagic lesion and postoperative CT shows resection cavity with air pockets and near complete resection of lesion (h).

Histopathology

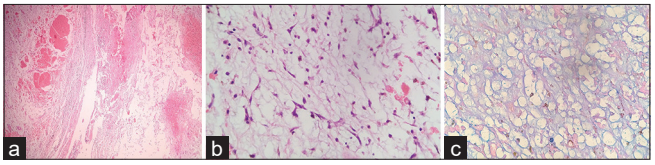

Figure 2: (a) Normal brain parenchyma on right with circumscribed hemorrhagic lesion on left – ×5 zoom. (b) Hypocellular tumor with scattered bland spindle cells in basophilic matrix – ×40 zoom. (c) Alcian blue highlights basophilic matrix.

DISCUSSION

Cardiac myxoma is the most common benign heart tumor, that is, sporadic (93% of cases) with a female preponderance.

Cardiac myxoma can also be a component of an autosomal dominant syndrome called Carney complex. Carney complex is characterized by spotty pigmentation (blue nevi and lentigines), myxomas (cardiac, cutaneous, and mammary), endocrine overactivity (Cushing's syndrome and acromegaly), testicular tumors, and Schwannomas.[14] The gross appearance of cardiac myxomas is variable – internally, they are heterogeneous and frequently contain cysts, necrosis, and hemorrhage.[3] The histogenesis of cardiac myxoma remains a theme of most debates. The two main hypotheses are that the tumor cells are derived either from multipotential mesenchymal cells or from endocardial neural tissue. It is speculated that patients may have an immune response to the neoplasm or to the heart muscle mediated by the presence of neoplasm, and the reaction leads to constitutional symptoms.[7]

Metastatic lesions have been reported to be diagnosed up to 8 years later than the primary lesion. In the majority of cases, the cerebral arteries, including the retinal arteries, are affected. However, it has been postulated that tumor tissues may grow into the walls of the vessels causing focal disruption of the internal elastic lamina, providing a nidus for cerebral hemorrhage and subsequent growth of metastatic tumor tissue.[5] It is most likely that the intracerebral hemorrhagic metastases resulted from the silent emboli of the cardiac myxoma occurring in the preoperative period or due to operative manipulation. These lesions are usually multiple and most commonly located in frontoparietal regions of brain. Our patient had a solitary lesion with the epicenter in the left occipital cortex. Transient or permanent visual loss may result from involvement of the retinal arteries. One study found significant association between occurrence of embolism and presence of a villous tumor surface.[1] Tumor fragments that may have metastasized into cerebral vessel walls may enlarge, causing vessel occlusion and delayed infarction[9] or they may penetrate through the vessel wall (vascular transgression), forming intra-axial metastases.[2]

The fate of the tumor fragment, which embolizes to the cerebral vessel within the central nervous system, still remains controversial. Two late complications have been reported: either a tumor fragment may grow and present as an expanding mass lesion, or it may develop a vascular aneurysm at the site of the embolus.[8] A study recently showed overproduction of CXC chemokines (a subgroup of small chemokine proteins), interleukin-8, and growth-related oncogene by myxoma cells.[11] Overproduction of CXC chemokines may explain malignant potential of the lesion. Metastatic brain lesions sometimes present earlier than the diagnosis of primary lesion.

As the natural history of this disease is unknown, the standard of care for these intraparenchymal lesions remains controversial. Thus, a high index of suspicion is required so that diagnosis can be effectively made and patients can be put under surveillance.

Our patient underwent a surgery for myxoma 1 year back. As our patient had a cortical bleed in an accessible location, excision biopsy was possible, which helped us arrive at a diagnosis. In patients who present with multifocal intracerebral hemorrhages, biopsy from a noneloquent region is mandatory to establish the cause for bleed.

CONCLUSION

A standard of care in the management of cardiac myxoma patients with cerebral metastases is yet to be established. While surgery may be appropriate in cases with one or two isolated brain metastases, palliative radiotherapy could be administered to patients with multiple brain metastases. Vigorous workup must be pursued if the patient becomes symptomatic yet again or develops fresh neurological manifestations.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Acebo E, Val-Bernal JF, Gómez-Román JJ, Revuelta JM. Clinicopathologic study and DNA analysis of 37 cardiac myxomas: A 28-year experience. Chest 2003;123:1379-85.
2. Bazin A, Peruzzi P, Baudrillard JC, Pluot M, Rousseaux P. Cardiac myxoma with cerebral metastases. Neurochirurgie 1987;33:487-9.
3. Burke AP, Virmani R. Cardiac myxoma. A clinicopathologic study. Am J Clin Pathol 1993;100:671-80.
4. Ekinci EI, Donnan GA. Neurological manifestations of cardiac myxoma: A review of the literature and report of cases. Intern Med J 2004;34:243-9.
5. Hwang BJ, Connelly MM, Lev MH. Distinctive MR imaging appearance of hemorrhagic cerebral aneurysms associated with atrial myxoma. AJR Am J Roentgenol 2001;177:925-7.
6. Jean WC, Walski-Easton SM, Nussbaum ES. Multiple intracranial aneurysms as delayed complications of an atrial myxoma: Case report. Neurosurgery 2001;49:200-2.
7. Keeling IM, Oberwalder P, Anelli-Monti M, Schuchlenz H, Demel U, Tilz GP, *et al*. Cardiac myxomas: 24 years of experience in 49 patients. Eur J Cardiothorac Surg 2002;22:971-7.
8. Kumar A, Sachdev A, Singh R, Lehl SS, D'Cruz S, Mahapatra M,

et al. Left atrial myxoma presenting as pseudobulbar palsy. Neurol India 2002;50:219-21.
9. Price DL, Harris JL, New PF, Cantu RC. Cardiac myxoma. A clinicopathologic and angiographic study. Arch Neurol 1970;23:558-67.
10. Rodrigues D, Matthews N, Scoones D, Aziz F, Nath F. Recurrent cerebral metastasis from a cardiac myxoma: Case report and review of literature. Br J Neurosurg 2006;20:318-20.
11. Sakamoto H, Sakamaki T, Sumino H, Sawada Y, Sato H, Sato M, *et al*. Production of endothelin-1 and big endothelin-1 by human cardiac myxoma cells--implications of the origin of myxomas. Circ J 2004;68:1230-2.
12. Shapiro LM. Cardiac tumours: Diagnosis and management. Heart 2001;85:218-22.
13. Shimono T, Makino S, Kanamori Y, Kinoshita T, Yada I. Left atrial myxomas. Using gross anatomic tumor types to determine clinical features and coronary angiographic findings. Chest 1995;107:674-9.
14. Stratakis CA, Kirschner LS, Carney JA. Carney complex: Diagnosis and management of the complex of spotty skin pigmentation, myxomas, endocrine overactivity, and schwannomas. Am J Med Genet 1998;80:183-5.
15. Yoon DH, Roberts WC.Sex distribution in cardiac myxomas. Am J Cardiol 2002;90:563-5.

How to cite this article: Ghodasara SA, Balasubramanian R, Varadharajan S, Shobhanaa PS. Cardiac phoenix in the brain-occult intracranial hemorrhagic metastases from completely resected atrial myxoma. Surg Neurol Int 2020;11:383.