

Brain Metastasis of Atrial Myxoma: Case report

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SUMMARY

Metastasis of an atrial myxoma to the brain is extremely rare¹. Thus far there are only 17 cases reported, including our present case. Most of the brain metastases manifest only in 3 to 6 decades, after an average time frame of one to two years after surgical removal of parental tumour. We present a case of brain metastases of atrial myxoma in a teenager of the youngest age among all reported cases, unusually as early as 15 years old³. The progress of the metastatic process had been insidious for three years after heart surgery. The imaging demonstrated a rather sizeable tumour by the time when the patient is symptomatic. The location of the metastatic tumour is anyhow superficial to the cortical surface, enabling complete surgical excision of the tumour easily achievable with favourable outcome.

KEY WORDS:

Atrial Myxoma, Brain Metastasis, Cardiac Benign Tumor, Cardiac Myxoma

INTRODUCTION

Primary cardiac tumors are rare (up to 0.5% in autopsy series). Amongst them, atrial myxomas are the most common and account for about 40-50% of the cases. The majority of these benign primary cardiac tumors are sporadic lesions (93% of cases) and usually occur in middle-aged women and the remainings are familial episodes. Complete surgical removal of the myxoma and its cardiac attachment is usually curative. The frequency of recurrences in cardiac myxomas varies between 3% for sporadic cases and 22% for cases of Carney complex. Recurrence has been related to incomplete excision, multifocality, and embolism of tumor fragments. Metastasis of an atrial myxoma to the brain is extremely rare¹. Thus far there are only 17 cases reported, including our present case. Most of the brain metastases manifest only in 3 to 6 decades. We report the youngest patient who has brain metastases of atrial myxoma unusually as early as 15 years old³.

CASE REPORT

A 15 years old Chinese girl with a background history of left atrial myxoma with total tumor excision done at Institut Jantung Negara on 20th April 2006, presented to us on 13 November 2009 with progressive right sided headache, nausea and vomiting which was markedly worsened for the past 1 week. There was no history of fever, trauma or seizure. Clinically she was alert, intact higher mental functions and no neurological deficit elicited. Full blood count (FBC), blood urea and serum electrolyte (BUSE), liver function test (LFT) and prothrombin time (PT)/ partial thromboplastin time (PTT) were unremarkable.

Computer tomography (CT) brain showed multiple enhancing solid-cystic lesions at grey white junctions of the right parietal and occipital lobes with significant perilesional edema and midline shift. One enhanced nodule with wall and calcification was noted at the right Parieto-Occipital area sized 2.4 x 1.7 cm. It has a cystic component measuring 7.4 x 4 cm. Another enhancing nodule measuring 1.9 x 1.4 cm is found at the grey white matter junction anterolateral to the aforementioned cystic component.

Right craniotomy and excision of the tumor was performed on 14th November 2009.

Intraoperatively, the tumor was oval in shape, well encapsulated, dark red in colour and poorly vascularised. There was dense adhesions between the tumor and the brain parenchyma. About 40 mls of straw-coloured cystic fluid was aspirated. The solid component as well as the cyst wall was otherwise removed completely from the surrounding brain parenchyma.

Post operatively she was well and her symptoms resolved completely. She attended our out patient clinic for follow up a year later. There is no new sign or symptom. Magnetic Resonance Imaging revealed no recurrence of the previously excised tumour. There is no expansion of the other lesion. No new treatment was initiated. She is currently under close follow up at the local hospital.

DISCUSSION

Atrial Myxoma is a rare primary cardiac tumour of mesenchymal origin affecting less than 0.05% of the population. It may present with many clinical syndromes. 60% of the patients are seen primarily by cardiologists with palpitations, exertional dyspnea, or syncope. In 30% of cases, the patients present with constitutional symptoms and non-specific manifestations, e.g. fever, loss of weight, malaise, leucocytosis, elevated erythrocyte sedimentation rate, hypergammaglobunaemia. This is explained by the dislodgement of the friable and spongy component of the atrial myxoma into the systemic circulation. The other 30% present with embolization. The central nervous system is one of the most susceptible areas of embolization most often resulting in ischemic strokes. The emboli, in the form of tumour or blood clot usually enter the large-diameter Middle Cerebral Artery and get occluded at its bifurcation areas. Independent cerebral metastatic growth, intraparenchymal hemorrhage and oncotic aneurysm are uncommon neurological sequelae of Atrial Myxoma⁴.

The central nervous system (CNS) manifestations include progressive headache, nausea or vomiting due to the increase

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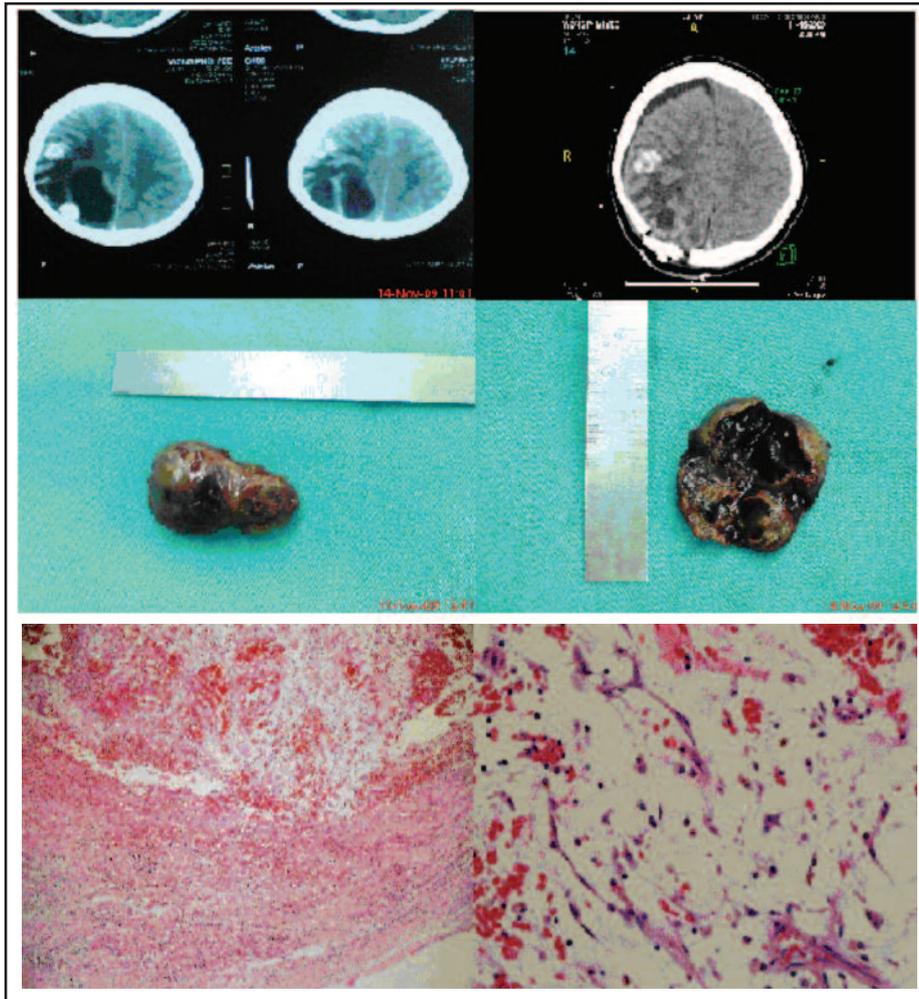


Fig. 1: Upper left : Contrast-enhanced CT brain of the patient showed multiple small rounded well enhanced lesions seen over the right parietal and occipital areas. The occipital lesion showed huge cystic component with mass effect. Upper right : Post operation CT brain image shows complete excision of the superficially located lesion. Middle left : Gross pathology: the excised tumour measures 5 x 3 cm. It has a well circumscribed margin. It is brownish in colour with shiny gelatinous surface Middle right : Intra-tumoral hematoma was seen after the tumor was incised by the surgeon. Bottom: Sections under microscopic examination showed circumscribed fragments of fibrocollagenous tissue with extensive area of hemorrhage. Under light microscopy, myxomatous component is seen. It is characteristically composed of stellate or fusiform shaped cells surrounded by loose stroma with abundant basophil cells infiltration. Tying together the patient's background history of atrial mycoma, the lesion represents metastasis from its cardiac origin.

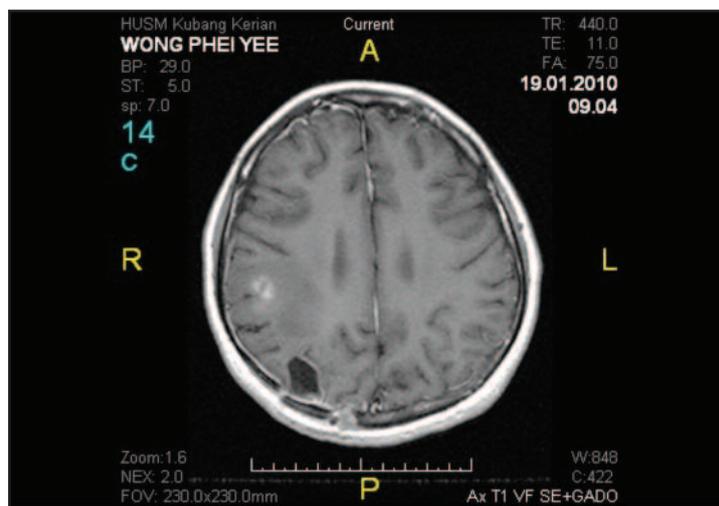


Fig. 2: Follow-up Magnetic Resonance Imaging revealed gliosis over the previously operated site. The right parietal lesion does not show expansion in size. cardiac origin.

of intracranial pressure, limbs or body weakness or numbness and new onset of seizure. These signs and symptoms should warrant the clinician to arrange for CNS imaging and referral to neurosurgical colleague.

The CNS manifestations of atrial myxoma may be due to the tumor emboli causing the cerebral ischemia/infarct, cerebral aneurysmal ruptures or the enlargement of tumor emboli exerting compression to the surrounding neural structures².

The diagnosis was concluded eventually on Histopathological examination. Stout standard is applied in the pathologic diagnosis making. At gross pathological examination, the tumour has a gelatinous appearance. Microscopic examination reveal hypocellular, scattered stellate or fusiform cells in a relatively avascular and highly myxoid stroma. The histopathological findings of the index case represent the characteristic features of myxoma. Correlating with the background history of atrial myxoma, the lesions are most likely metastases from their cardiac origin.

CONCLUSION

Long term follow up of the patients with atrial myxoma even after complete surgical excision is mandatory and thorough explanation of the potential risks of recurrence of the cardiac tumor, the late development of cerebral lesions and the enlargement of the cerebral lesions is necessary. Vigorous work-up must be pursued if the patient again becomes symptomatic or develops new central nervous system manifestations.

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