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Case report

Brain metastasis of cardiac myxoma: case report

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Summary

Cardiac myxoma is the most common benign heart tumor. Cardiac myxoma can be a sporadic lesion (93% of cases) and usually occurs in women over 30 years. Complete surgical removal of the myxoma and its cardiac attachement is usually curative. The frequency of recurrences in cardiac myxomas was 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. We report a case with multiple brain metastases revealed by mycotic aneurysm and seizure intractable.

Keywords: Brain metastasis;Cardiac myxoma; Surgery

Résumé

Métastases cérébrales d'un myxome cardiaques: à propos d'un cas

Les myxomes sont des tumeurs cardiaques bénignes. Ces sont des lésions sporadiques (93% des cas) et habituellement observées chez la femme de la trentaine. L'exérèse chirurgicale complète emportant la base d'insertion de la tumeur est le seul traitement curatif. La fréquence des récidives est de 3% pour les cas sporadiques et de 22% pour le Syndrome de Carney. Les récidives sont liées à une exérèse incomplète, à une multiple localisation de la tumeur ainsi qu'à une migration d'un fragment tumoral. Nous en rapportons un cas avec de multiples métastases cérébrales révélées par un anévrysme mycotique et des crises d'épilepsie.

Mots-clés: Chirurgie; Métastase cérébrale; Myxome cardiaque

Introduction

Primary cardiac neoplasms are rare in humans. The incidence of primary cardiac tumors in autopsy series range from 0.02 to 2.8% - less than metastatic cardiac tumors (4%) [1]. They are predominantly benign heart tumor, which is found in the left atrium (about 86% of lesions) [1]. Complete surgical removal of the myxoma and its cardiac attachment is usually curative. Recurrence has been related to incomplete excision, multifocality, and embolism of tumor fragments. Since most myxomas are located in left atrium, systemic embolism is the most frequent metastatic complication for cardiac myxoma [2].

Case report

A 53 year-old female patient was admitted in the hospital for seizure intractable by drug therapy. Her medical story disclosed same disease: mycotic aneurysm, high blood pressure. Echocardiography revealed a large tumor mass of the left atrium. Tumor excision was performed under cardiopulmonary bypass in 2001. Pathologic diagnosis was reported as "benign myxoma". She was readmitted to the hospital with the symptoms of headache, vertigo and general seizure in april 2005. Brain magnetic resonance imaging (MRI) showed multiple hemorrhagic lesions (largest one with 3cm in diameter). No surgery was performed and she has been discharged. She was readmitted to the hospital with the symptoms of seizure intractable, weakness and transient visual loss in February 2006. Computer Tomography (CT) showed multiple hemorrhagic lesions located in right frontal lobe and bilateral occipitoparietal lesion with oedema (Figure 1 and 2). Left parietooccipital lesion was excised. The diagnosis of myxomas brain metastasis was confirmed. Pathologic

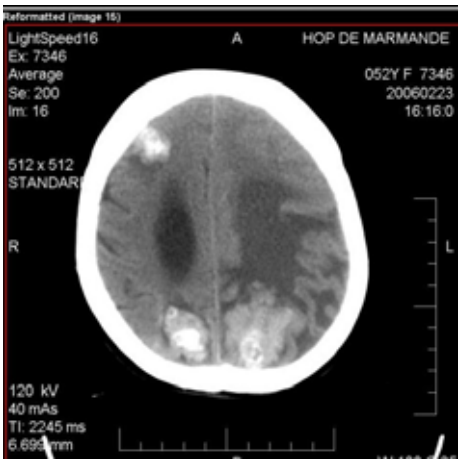

Fig. 1: Brain CT showing multiple cerebral lesions and oedema

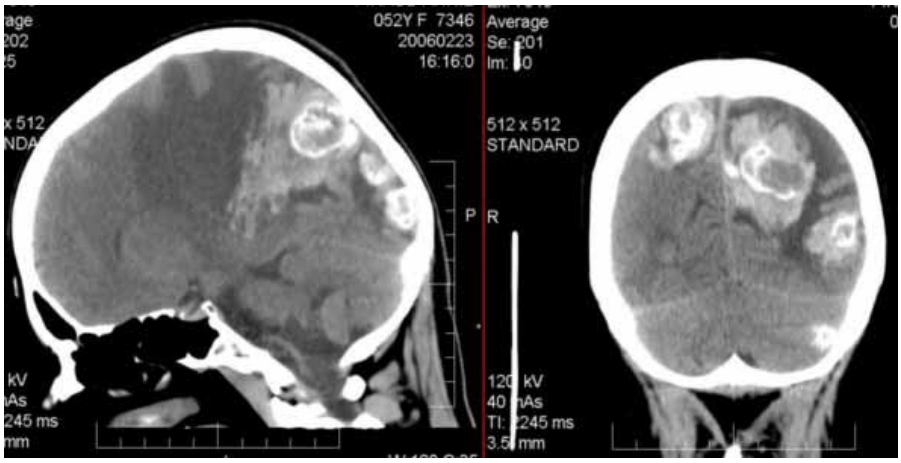

Fig. 2: Brain CT showing multiple cerebral and left cerebellum lesions and oedema

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diagnosis was again reported as benign myxoma consistent with emboli. Exploration to reveal primary source of emboli did not show any evidence of recurrence in heart. Patient did not receive whole brain radiation nor chemotherapy. She stayed at hospital during two weeks in postoperative.

Discussion

Cardiac myxoma can be a sporadic lesion (93% of cases) and usually occurs in women over 30 years [1]. Cardiac myxoma can also be a component of autosomal dominant syndrome called Carney Complex. Carney complex is characterized by spotty pigmentation (blue nevi and lentigines), myxomas (cardiac, cutaneous, and mammary), endocrine over-activity (Cushing Syndrome and acromegaly), testicular tumors, and schwannomas [3]. Cardiac myxomas of Carney complex are histologically indistinguishable from sporadic cardiac myxomas and most often arise in left atrium; however, they exhibit no age or sex preference and can present a multiple concurrent lesion in any cardiac chamber. The gross appearance of cardiac myxomas is variable. Internally, they are heterogeneous and frequently contain cysts, necrosis, and haemorrhage. It is characterized histologically by amorphous myxoid matrix polygonal to stellate cells that are arranged singly, in small clusters, or rudimentary vascular formations. Histogenesis of cardiac myxoma remains a theme of debates, the 2 main hypotheses are that the tumor cells are derived either from multipotential mesenchymal cells or from endocardial neural tissue [4]. The common clinical signs are rhythm disturbances, myocardial ischemia, pulmonary oedema, syncope and cardiac arrest. They do normally lead to the diagnostic hypothesis of an intracardiac mass. Active illness is often accompanied by elevation of erythrocyte sedimentation rate (ESV) and C-reactive protein (CRP), hyperglobulinemia and anemia. Constitutional symptoms may be mediated by interleukin-6, produced by myxoma itself. Significant immunological alterations in myxoma patients seem to occur pre- and post-operatively for ESR, serum protein electrophoresis, CRP, fluorescence-activator cell sorter, interleukin -2R, and intracellular adhesion molecule. After myxoma surgical removal, all those biochemical analysis return to normal. Therefore, it is speculated that patients may have an immune response reaction to neoplasm for heart muscle mediated by the presence of neoplasm, and the reaction leads to constitutional symptoms. Currently, the presence of cardiac masses is easily detectable by routine imaging techniques: echocardiography, MRI, and computerized tomography (CT). The primary modality for imaging is echocardiography, which usually con firms the suspected diagnosis. But in rare cases there are masses, which cannot be exactly identified by this technique. Complete surgical removal of the myxoma and its cardiac attachment is usually curative. The frequency of recurrence in cardiac myxomas varies between 3% for sporadic cases and 22% for Carney complex cases [5]. Recurrence has been related to incomplete excision, multifocality, and embolism of tumors fragments. Cardiac myxomas seem to recur more often in young males and in those who have a family history of the tumors [3]. Published figures for embolism range from 6 to 43% [5]. Right atrial and right ventricular myxomas cause recurrent pulmonary that can produce severe pulmonary hypertension and death. In addition, atrial myxomas can occasionally become infected with bacteria or fungus with consequently endocarditis and septic emboli. Since most myxomas are located in the left atrium, systemic embolism is particularly frequent. In majority of case, the cerebral arteries, included retinal arteries, are affected. Therefore, cerebral metastases are most frequent sites for cardiac myxomas. Transient or permanent visual loss may result from involvement of the retinal arteries. Occlusions of peripheral arteries and embolization into visceral, renal, and coronary arteries can also occur, and complete obstruction of abdominal aorta and arteries by a large tumor embolus originating from the left ventricle and aortic saddle embolism have even been reported [2]. One study found significant association between occurrences of embolisms and a villous tumor surface [3]. Tumors fragments that have metastasized to cerebral vessel walls may enlarge, causing vessel obstruction and delayed infarction or they may penetrate through the vessel wall, forming intra-axial metastases [6]. The standard management of patients with cerebral metastases has not been established because of the rarity of the cases [7-9] (Table 1). Metastatic brain lesions sometimes are present earlier than the diagnosis of primary lesion. However, metastatic lesions diagnosed up to 8 years later than the primary lesion was also reported. These lesions are usually multiple and most commonly located in the parietooccipital regions. Surgery was performed in some cases having one or two metastatic lesions (Table 1). Only one patient other our cases received whole brain radiotherapy with 50Gy doses for multiple cerebral metastases in the literature. One patient received postoperative chemotherapy with ifosfamide and doxorubicin and lived longer without recurrence [26].

Conclusion

In conclusion, standard of care in the management of cardiac myxoma patients with cerebral metastases has not been established. Surgery may be appropriate with in case with one or two isolated brain metastasis. Palliative radiotherapy could be administrated to patients with multiple brain metastases. However, chemotherapy should not be considered, at least within the context of current literature.

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