Right atrial angiosarcoma mimicking a myxoma: from a wrong diagnosis to a difficult decision

Miksomayı andıran sağ atriyal anjiyosarkom: Yanlış bir tanıdan zor bir karara

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The case described deals with a 27-year old man who underwent surgical approach to remove a large right atrial mass diagnosed as cardiac myxoma. During the operation several biopsies were performed because of its gross aspect. An angiosarcoma was diagnosed, thus, a surgical excision was the only possible procedure. Six months later, a computed tomography scan revealed the presence of substitutive and hemorrhagic cerebral lesions. The patient expired 10 months after surgery. The best therapy for primary angiosarcoma would have been an early and complete surgical excision. Unfortunately, often an important involvement of vital cardiac structures is present; in this case an orthotopic cardiac transplantation could be considered.

Key words: Angiosarcoma; myxoma; cardiac tumor; heart transplantation.

Bu yazıda, kardiyak miksoma tanısı konan büyük bir sağ atriyal kitlenin çıkarılması için cerrahi tedavi uygulanan 27 yaşında erkek hasta sunuldu. Kitlenin oldukça büyük olması nedeniyle ameliyat sırasında birkaç biyopsi uygulandı. Anjiyosarkom tanısı kondu, dolayısıyla cerrahi eksizyon tek çareydi. Altı ay sonra bilgisayarlı tomografi sonucunda hemorajik serebral lezyonların olduğu görüldü. Hasta ameliyattan 10 ay sonra kaybedildi. Primer anjiyosarkom için en iyi tedavi yöntemi erken ve tam bir cerrahi eksizyon olacaktır. Ne yazık ki sıklıkla hayati kardiyak yapıların önemli bir bölümünün de tutulumu görülür, bu durumda ortotopik kardiyak transplantasyon düşünülebilir.

Anahtar sözcükler: Anjiyosarkom; miksoma; kardiyak tümör; kalp nakli.

Primary cardiac neoplasms are uncommon both in adult and pediatric age groups with a varying incidence, from 0.0017% to 0.28% according to different autoptic series.[1] Among these, sarcomas without any differentiation, angiosarcomas, leiomyosarcomas, and rhabdomyosarcomas are the most common histologic types.[2] They occur most frequently in the third to fifth decade and are more common in males (M:F ratio 2:1).[3] Despite the new therapeutic measures available, the survival of patients with cardiac sarcomas is still limited and it ranges from six months to two years.[2] Herein, we present a case of a young man with an extensive neoformation involving the right atrium.

CASE REPORT

In August 2004, a 27-year-old man was referred to our department for the surgical management of a right atrial mass. The patient had no medical history except for pericarditis symptoms (dyspnea, fever, chest pain). On admission, physical examination was normal and heart sounds were not muffled. Transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) confirmed the presence of a large endocavitarian mass (56.3x53.2 mm) attached on the posterior and inferior right atrial wall. A computed tomography (CT) demonstrated a dishomogeneous mass with irregular lobulated border which filled half of the atrial cavity (Fig 1). Exams seemed to present a good cleavage plane so the patient underwent sternotomy to remove the atrial mass. In spite of preoperative examinations after pericardiotomy the right atrium appeared mammillated with a large mass apparently infiltrating the atrial wall. So, multiple transatrial biopsies were performed. The chest was closed and the patient re-sent to cardiologic ward. Histologic diagnosis of angiosarcoma was made with immunochemistry studies and showed a moderate cellular differentiation and no foci of hemorrhage or necrosis. In some sites the stroma was scanty and

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dominated by spindle-shaped cells surrounding vascular structures. In other sites papillary structures or nests of spindle-shaped were observed, with a mitosis index of 21 mitosis/HPF (high-power fields). The tumoral cells were positive for vimentin, CD 34 and CD 31, negative for keratin, desmin, actinin and protein S100.

After the diagnosis of primitive malignant cardiac tumor, a cardiac-gated magnetic resonance imaging (MRI) was carried out. It confirmed the right atrial enlargement containing a large circular neoformation, with polilobated borders, attached to the lateral and superior wall. The inferior vena cava outlet was respected, whereas the superior vena cava appeared crushed by the mass. Then, MRI revealed a tumor with transversal diameters of 5x8 cm.

There were not any pathologic findings across other cardiac structures. Considering the volume of the tumor and the difficulty of its complete surgical resection, the option of a heart transplant was evaluated. However, the heart transplantation center refused to perform the operation because of the poor prognosis and high risk of recurrence. So much so that, in absence of evidence of distance metastases, an attempt for surgical resection was made. Cardiopulmonary bypass (CBP) was instituted by means of aortic and right femoral vein cannulation. Moreover, the operation was performed in circulatory arrest in order to obtain a better surgical exposure with a bloodless operation field and allow the largest possible excision. The tumoral mass was easily located, widely filling the right atrial cavity it appeared enlarged. A partial excision of the mass (7x5 cm) was allowed only, due to a wide implant base on the right atrial free wall. A right atrial reconstruction with a bovine pericardium patch was carried out too.

Postoperative outcome was uneventful, and the patient underwent three cycles of post-operative chemoterapeutic treatment (Epirubicina, Mesna, Ifosfamide). A month late, an MRI showed hepatic involvement. Then, the patient showed signs of neurological disfunction (cephalea, cognitive disorders) six months later. Actually, a CT scan revealed the presence of substitutive and hemorrhagic lesions (Fig. 2); the patient expired 10 months after surgery.

DISCUSSION

The evolution of cardiac surgery and, in particular, the development of CPB has allowed the attempt of resection of virtually all primary cardiac tumors.[4] Distant metastases occur in 66% to 89% of the patients, and the reported mean survival is from three to 6.6 months. This metastatic spread is independent from the surgical approach, whether wide resection or complete heart transplantation.[5] The diagnosis of these soft tissue tumors is often delayed because the symptoms are nonspecific and do not occur until the primary tumor is large enough to involve the vital cardiac structure.[1,6] Actually, most cardiac sarcomas interfere with cardiac function, they usually cause symptoms of congestive

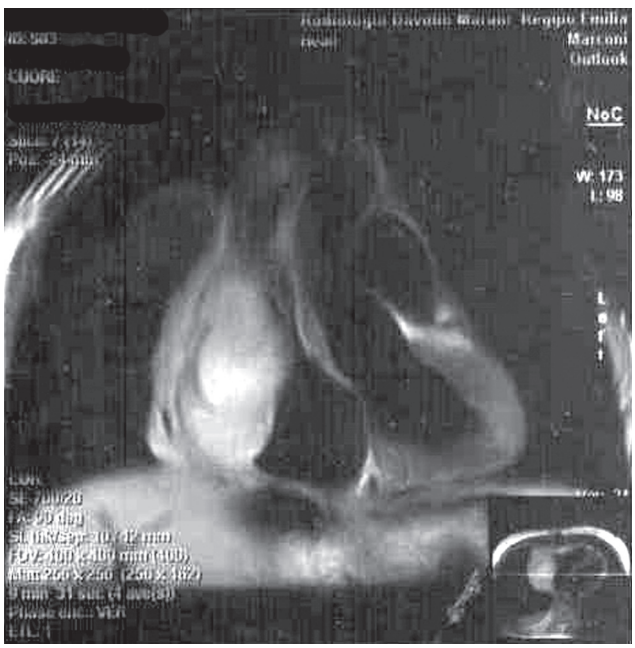

Fig. 1. Pre-operative computed tomography scan showing a large right atrial mass.

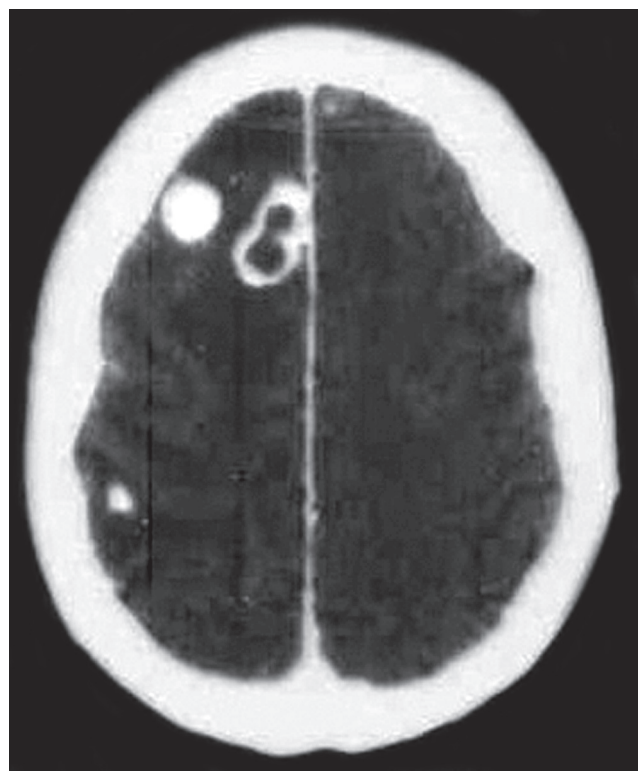

Fig. 2. Computed tomography scan revealing late cerebral hemorragic and substitutive lesions.

heart failure, simulating mitral stenosis, dyspnea, chest pain, palpitations, fever, arrhythmias, pericardial effusion, syncope, hemoptysis, tumor embolism, constitutional symptoms (e.g., weight loss, anemia), cerebral ischemia or myalgia as a result of the wide anatomic local extension with severe cardiac damage and obstruction of the blood flow.[1,7] For these reasons, excision is an attractive option to relieve symptoms and prevent early death but it is merely a palliative solution. Cardiac transplantation might be the only hope for palliation in patients with involvement of vital cardiac structures such as valves, arteries, and conduction tissue. Therapeutic management must mainly consider histologic subtypes and involves chemotherapy, radiotherapy and surgery.[8] The role of adjuvant chemotherapy before or after surgical resection remains controversial. As a matter of fact, even if polychemotherapies may optimize the rate of therapeutic response, there is no evidence of increasing survival rate except for a few cases reported by Sorlie et al.,[9] Percy et al.[10] and Nakamichi et al.[4,8] Moreover, there has been only an isolated report of a cardiac angiosarcoma that responded to a treatment with liposomal doxorubicin with a disease free-survival of 11 months in spite of incomplete resection.[3] As underlined by Uberfuhr et al.,[5] at present the best option might be an orthotopic cardiac transplant for patients with unresectable malignant primitive cardiac tumor with no evidence of distant dissemination. However, reports about transplantation show disappointing results and a standard approach has not been codified yet to such a disease. Aravot et al.[11] and Grandmougin et al.[2] reported two exceptional cases in which cardiac transplantation, for a neurofibrosarcoma and a rhabdomiosarcoma respectively, obtained a survival of more than five years.[6] On the contrary, there are many reports with survival times from six to 16 months,[5] probably related to immunosuppressive treatment that might have triggered an explosive tumor growth, promoting the dissemination, before heart transplantation, of undetectable micrometastases. An obvious restriction for transplantation, that made this option more arguable, is the scarcity of donor organs. In fact, an ethical question is whether the use of donor hearts for patients with malignant cardiac tumors is acceptable in view of the continuing organ shortage. For this reason and for dubious results previously described, our patient was not enlisted for heart transplantation. Therefore, given the small number of cases of angiosarcoma and the equivocal results of the treatments described in literature, a difficult decision had to be faced. We chose to perform the widest possible surgical excision. Although arguable, this option was taken in order to relieve the patient symptoms and improve his quality of life despite the short expected survival. Furthermore, besides medical considerations, the patient's young age, his and his parents' expectation, due to the wrong initial diagnosis of benign tumor, led us to operate him. What made it even worse is that cardiac surgeon is not used to such aggressive and malignant disease at such young age. Unfortunately, this young have no other treatment option[5] and still remain a surgeon's weakness in front of a poor prognosis.

Hopefully, in future, this kind of patients, not eligible for cardiac transplantation might benefit from total artificial heart or from xenotransplantation, although there is still more work to be done to make these techniques more available and safer.

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