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Primarycardiac Osteosarcoma: A Multidisciplinary Case Report

Case Reports

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Abstract

Objective: Primary cardiac osteosarcoma is a rare entity. The most common location of this tumor is the posterolateral wall of the left atrium. Clinical presentation of the tumor depends on the tumor size and the proximity to cardiac valves, so most osteosarcomas are often asymptomatic and heart failure symptoms arise when the growing mass impairs cardiac hemodynamics or invades surrounding structures. In these cases a multidisciplinary approach is essential for a complete classification and treatment of the disease.

Materials and Methods: We here describe a case of a 62-year-old woman who presented with high-frequency atrial fibrillation and severe dyspnea. Imaging studies showed a huge, firm mass occupying more than half of the left atrium with a large osteoid component, infiltrating the mitral valve and the atrio-ventricular sulcus. The complete therapeutic plan was composed with the contribution of cardiologists, cardiac surgeons, pathologists, radiologists and oncologists. The palliative surgical treatment was required for the severe hemodynamic impair unresponsive to medical treatment. Histologic examination confirmed the macroscopic diagnosis.

Results: The Patient recovered from the operation and was admitted for chemotherapy treatment, but a severe pneumonic complication between the first two cycles of therapy led her to death about two months after the diagnosis.

Conclusion: From a surgical point of view, we suggest the described bi-atrial access to have the widest exposition of the mass and nearest structures. When a complete excision can be planned, cardiac autotransplantation should be considered. A sort of cardio-oncological collegial group is essential for the treatment of this and all cases in which a neoplasm has involvement on the cardiovascular system. This also applies to those cases in which cancer treatment may be limited by concomitant cardiac pathologies.

Keywords: cardiac tumor, heart team, multidisciplinary, sarcoma

Introduction

Cardiac tumors are abnormal tissue growths in the heart or heart valves. The tumors can be malignant or benign. If they begin growing in the heart and stay there are called primary tumors, while tumors that start in another part of the body and move to the heart are called secondary tumors. Primary cardiac tumors are very rare and most likely benign. But, even benign tumors can cause problems because of their size and location: they can embolize to vital

organs or obstruct cardiac structures and valves. Primary cardiac osteosarcoma is a rare and aggressive neoplasm, usually localized in the posterior wall of the left atrium. This position allows the tumor to grow significantly before giving signs and symptoms.

Case Report

We here report the case of a 62-year-old previously healthy woman who presented on October, 10, 2016 with progressive dyspnea, asthenia, and palpitation for about two weeks before admission in our Emergency Room. The Patient was anxious, dyspnoic, with low blood pressure values. Cardiac auscultation revealed a systolic murmur grade 4/6 over the left sternal border. The electrocardiogram showed atrial fibrillation with rapid ventricular response. Standards antero-posterior radiologic exam of the thorax showed bilateral pleural effusion, mainly at the left side.

Laboratory investigations, including complete blood cell count, electrolytes and renal function were within normal range. Liver enzymes were: ALT=162 U/L and AS =126 U/L. C-reactive protein was 19.8 mg/dl, N-terminal-pro-B-type natriuretic peptide 1484 pg/ml.

Transthoracic Doppler-echocardiography revealed a big, heterogeneous mass in the left atrium, adhering to the posterior atrial wall and posterior leaflet of the mitral valve causing moderate mitral stenosis, moderate mitral regurgitation and moderate pulmonary hypertension (45 mmHg). The mass protruded in the enlarged left atrial cavity, was firm with a small portion protruding into the mitral valve and seemed to infiltrate the atrio-ventricular sulcus (Figure 1). Left ventricle was not dilated (EDV=57 ml; ESV= 31 ml), with a moderately reduced systolic function (EF=45%).

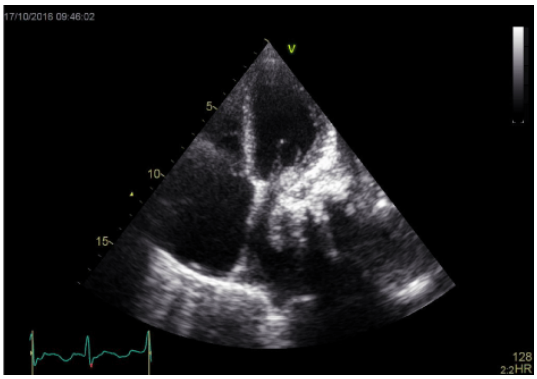

Figure 1: Echocardiogram showing the fixed left atrial mass; mitral leaflets are opened, mitral orifice is completely occupied by the tumor.

After few hours the rhythm turned sinusal and dyspnea relapsed completely. The characteristics of the mass did not depose for a benign lesion (mixoma), so further examinations were performed in the days that followed.

Cardiac computed tomography (CT) confirmed the presence of a solid, dyshomogeneous mass, with irregular margins, occupying almost totally the left atrial cavity (Figure 2). The dimensions of the mass were 7x4 cm. The mass was implanted in the infero-postero-lateral wall of the left atrium and showed two gross components: one hypodense that develops anteriorly and the other highly calcified that develops posteriorly. There was not obstruction of the omolateral pulmonary veins. CT examination did not show metastatic disease but only two mediastinal lymphatic nodes with increased volume.

Cardiac magnetic resonance confirmed the infiltration of the fat tissue of the atrio-ventricular sulcus (Video 1, a, b).

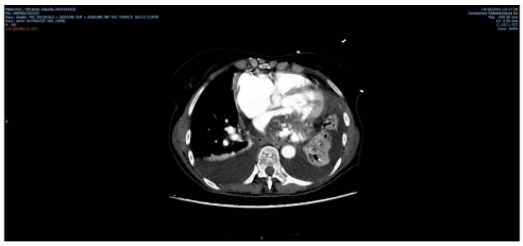

Figure 2: CT scan of the heart; to note the wide calcified region of the tumor.
Video1: Preoperative (a) and postoperative (b) magnetic resonance long axis video showing the hemodynamic obstruction and the result after surgical excision.

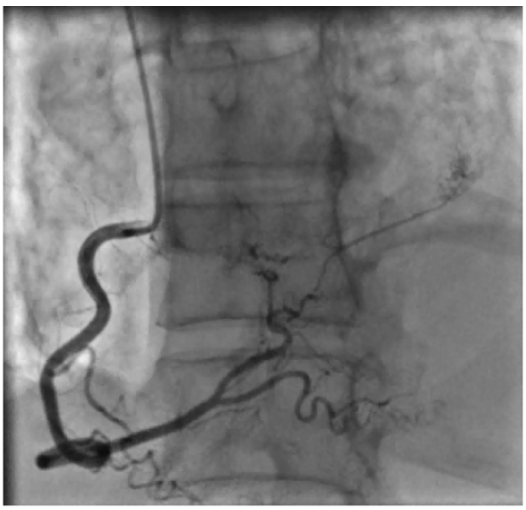

Figure 3: Coronary angiography of the right coronary artery: note the two-vessels supply coming from postero-lateral branch.

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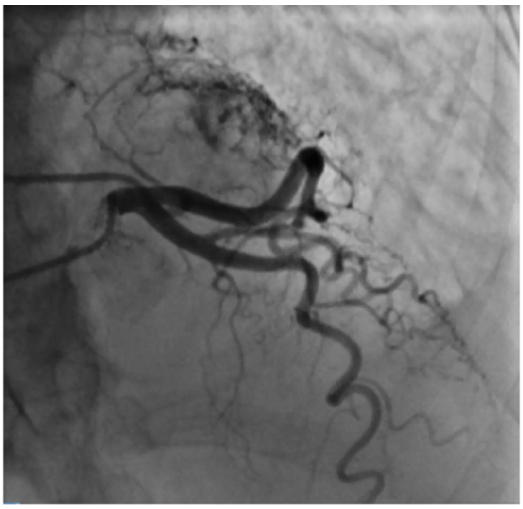

Figure 4: Coronary angiography of the left coronary system: note the vascularization of the tumor coming from the circumflex branch.

Surgical treatment

The patient was mobilized and showed a dyspnea at the minimal effort, with recidives of atrial fibrillation. Considering the extreme functional limitation and the theoretical possibility to treat the neoplasia after an immune-hystological diagnosis, a surgical operation to remove as much as possible of the malignant lesion was indicated and performed on October, 19, 2016.

Coronary angiography performed preoperatively showed a double vascularization of the tumor both from left and right coronary arteries (Figure 3 and 4).

The mass was reached through a biatrial-transeptal access (Giraudon incision), in order to have the widest control of the lesion (right atrial-interatrial septum-left atrial, via atrial roof access, (Figure 5). An extemporary histological examination was indicative for sarcoma. Intraoperative examination revealed the tumor to be deeply attached to the posterior wall of the left atrium and the posterior mitral leaflet. The atrial mass was partially resected, reaching the calcified, posterior portion. The tumor had a very slippery surface, so that was difficult even to grab it with surgical instruments. It was removed by small pieces with the aid of the electrocautery. We tried to free the posterior leaflet of the mitral valve from the mass as much as possible. Anuloplasty was excluded because the mitral annulus was invaded by the tumor. The patient tolerated the surgical procedure well and was discharged at rehabilitation facility on November 3, 2016.

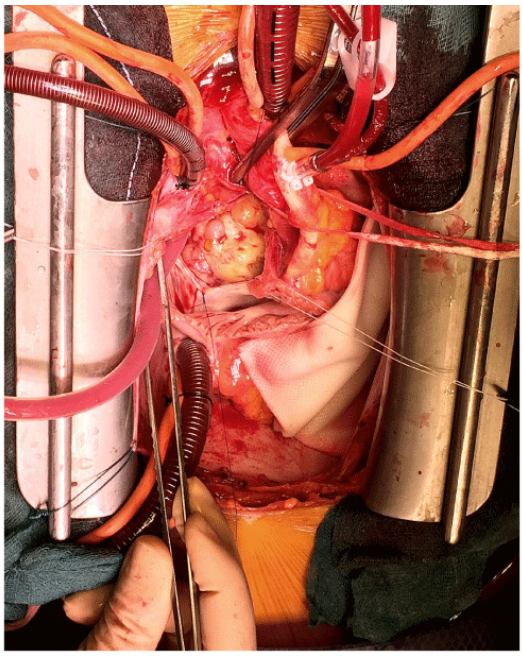

Figure 5: Intraoperative surgical view from the head of the Patient: the smooth surface is evident, as well as the complete invasion of left atrial cavity. Mitral valve was hidden by the tumor.

Medical treatment

Histopathological analyses revealed a heterogeneous malignant neoplasia. Areas of fibrous stroma including sparse tumour cells with atypia and areas with perivascular hypercellular cuffs alternate with tracts of cartilaginous tissue with high atypia and osteoid matrix (Figure 6-8). In this context, foci of necrosis are also present. The mitotic count is around 8 x 10 high power field (HPF). Atypical mitoses are seen. The morphology deposes for malignant mesenchimal proliferation, with areas of undifferentiated pleomorphic sarcoma and areas of osteoid matrix and condrosarcomatous tissues, thus leading to the diagnosis of primary cardiac osteosarcoma, according to the WHO classification of 2015 [1]. Immunohystochemical study reported positivity of Murine Double Minute 2 (MDM2), that distinguish low grade osteosarcoma (positive) from benign fibrous and fibro-osseous lesions (negative) [1].

Based on this hystochemical picture, chemotherapy with adriblastin and ifosfamid was indicated and performed during a second admission of the Patient in our Oncology Department, from December 5 to 7, 2016. The therapy was well tolerated and the Patient was discharged with a new cycle planned for January 2017.

Unfortunately, the patient died on December 16, 2016 after a severe, bilateral pneumonic event.

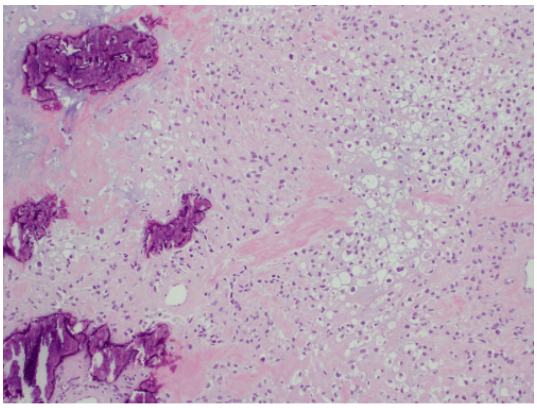

Figure 6: Areas with cartilaginous tissue and osteoid matrix.

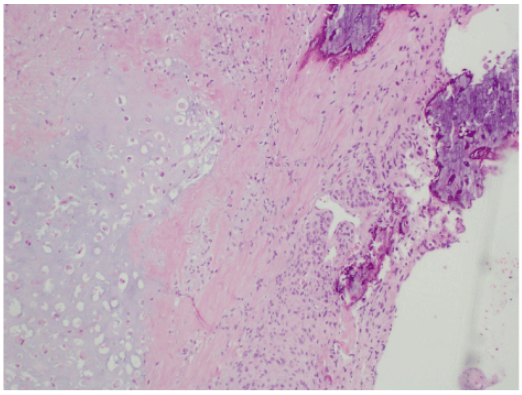

Figure 7: Areas with cartilaginous tissue and osteoid matrix.

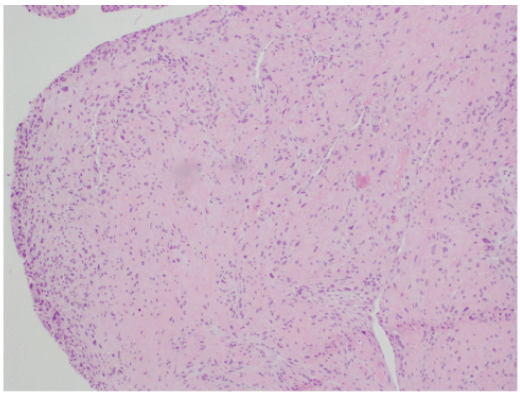

Figure 8: Areas with fibrous stroma with atypia.

Discussion

Malignant tumors constitute less than 25% of primary cardiac neoplasms. However, both primary sarcomas and benign tumors are often found in the left atrium. As a consequence of their location and similar clinical presentation, primary cardiac sarcomas can be easily confused with a benign myxoma, therefore we must carefully consider imaging features (immobility of the mass, neovascularity, multicentricity, calcification and invasion into the heart structures) to raise the suspicion for a cardiac sarcoma before proceeding to surgical / medical therapy. The localization into the left atrium of the tumor is as well a good indicator for its nature, given that sarcomas often develop from the postero-lateral wall of the left atrium and do not involve the interatrial septum [2].

At our best knowledge, we can argue from the Literature (we here resume the most recent reviews that widely contain all other references [3] that the present case is the 54th described case of primary cardiac osteosarcoma. The most important limitation in the treatment of this tumor is the impossibility to obtain a complete surgical excision, exception made for few cases discovered very early in their development. Nevertheless, surgical palliative / diagnostic procedure is often needed to avoid that the patient dies by heart failure complications and to obtain a precise immune-hysto-chemical diagnosis on which to base adjuvant therapy. From a surgical point of view, we suggest the described bi-atrial access to have the widest exposition. When a complete excision can be planned, cardiac autotransplantation should be considered.

Conclusions

We set up a cardio-oncological collegial group for the treatment of this and all cases in which a neoplasm has involvement on the cardiovascular system. This also applies to those cases in which cancer treatment may be limited by concomitant cardiac pathologies.In the future, thanks to the progress of anticancer therapies, the treatment of concomitant heart diseases will probably be increasingly used in these patients.

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