

Malignant primary tumor in left atrium with mitral valve invasion, pulmonary veins and cerebral metastasis: A case report

Tumor primario maligno en aurícula izquierda con invasión a válvula mitral, venas pulmonares y metástasis cerebral. Informe de un caso

Miguel Reynoso-Hermosillo¹, Jaime Sandoval-García¹, Baltazar López-Rosales¹, Antonio Esparza-Villarruel¹, José M^a Díaz-Sifuentes², David García-Martínez², and Wenceslao G. Ángeles-Bueno^{3*}

¹Department of Heart Surgery, High Specialty Medical Unit; ²Biomedical Research Unit 02; ³Department of Surgical Oncology, High Specialty Medical Unit. Specialty Hospital, Centro Médico Nacional de Occidente, Instituto Mexicano del Seguro Social, Guadalajara, Jalisco, México

Resumen

Introducción: La incidencia de tumores primarios cardíacos se calcula en 0.0017 a 0.28%, de los cuales el 25% es maligno y ocupa principalmente cavidades derechas. **Caso clínico:** Paciente que inicia con síntomas neurológicos por metástasis cerebrales de tumor primario cardíaco en cavidades izquierdas, que infiltra válvula mitral y venas pulmonares. Se realiza resección tumoral con reemplazo valvular mitral y remodelamiento auricular con uso de orejuela izquierda. **Conclusiones:** Las técnicas de imagen son fundamentales para el diagnóstico de esta afección, ya que la mayor parte de los tumores cardíacos se muestra asintomática hasta etapas tardías.

Palabras clave: Tumor primario cardíaco. Sarcoma intimal auricular. Metástasis cerebrales. Tratamiento quirúrgico.

Abstract

Introduction: The incidence of cardiac primary tumors is estimated to be 0.0017-0.28%, of which 25% are malignant, occupying mainly right cavities. **Clinical case:** Patient present with neurological symptoms due to brain metastasis of primary cardiac tumor in the left atrium, infiltrating the mitral valve, and pulmonary veins. Tumor resection was performed with mitral valve replacement and atrial re-modeling with the use of the left appendage. **Conclusions:** Imaging techniques are fundamental for the diagnosis of this pathology since the majority of cardiac tumors are asymptomatic until late stages.

Key words: Cardiac primary tumor. Primary intimal sarcoma. Brain metastasis. Surgical management.

Introduction

Malignant primary cardiac tumors are extremely rare neoplasms of varied histopathology. They account for 0.008% of cancer cases; the most common is

sarcoma, with the angiosarcoma subtype being the most prevalent¹.

Symptom presentation occurs in a quite varied way, which depends on the size and localization. Therefore, imaging studies are necessary to obtain morphological information about a tumor and its functional

Correspondence:

*Wenceslao G. Ángeles-Bueno

Belisario Domínguez, 1000

Col. Independencia

C.P. 44340, Guadalajara, Jalisco, México

E-mail: dr.wenceslao1@hotmail.com

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consequences². Left atrial sarcomas tend to be more solid and less infiltrative³.

The treatment of choice is complete resection, with valve reconstruction or replacement if necessary to maintain free margins. In selected cases, heart transplantation is considered the final treatment once the presence of metastasis has been ruled out⁴. In case of metastasis or if marginal resection is not possible, treatment should be complemented with radiation therapy. Although chemotherapy has not been shown to increase survival, it does reduce the risk of adverse events and, therefore, using it is recommended in the presence of metastasis, incomplete resection or in patients not eligible for surgical intervention⁵.

Clinical case

Thirty-four-year-old male patient, allergic to sulfonamides, without further medical history or comorbidities. In September 2017, he started suffering left hemi-cranial headache, which subsided with analgesics; 2 months later, the headaches were intensified and were accompanied by vertigo, with the condition even becoming incapacitating, which is why he attended the Regional General Hospital.

Diagnostic approach was started with computed tomography (CT) and nuclear magnetic resonance imaging (MRI) (Fig. 1), in which a left parieto-occipital tumor, cystic lesion with hyperintense halo enhancement was observed. He was referred to the Neurosurgery Department, where he underwent craniotomy, which identified a parasagittal parietal tumor with a thickened capsule containing an oily yellow fluid.

On histological study, a poorly-differentiated Sertoli cell tumor was observed, and a CT-scan was thus performed, which revealed a mediastinal tumor. Subsequently, a transthoracic echo-sonogram (TT-ECHO) was carried out, which documented a 17 mm × 19 mm tumor on the roof of the left atrium, with a broad base; an MRI was also performed, with findings of a 34 mm × 30 mm × 37 mm tumor in the left atrium that occupied 50% of the atrium, with infiltration to the right inferior pulmonary vein.

Cardiac tumor resection with mitral valve replacement was decided; the reported findings were: left atrial roof tumor that compromised the mitral annulus and mitral valve anterior leaflet, as well as sessile, indurated right inferior pulmonary vein with malignant characteristics (Fig. 2).

The procedure was carried out under balanced general anesthesia, Class II invasive surveillance, with a mid-sternal incision, cardiac exposure, and double cava

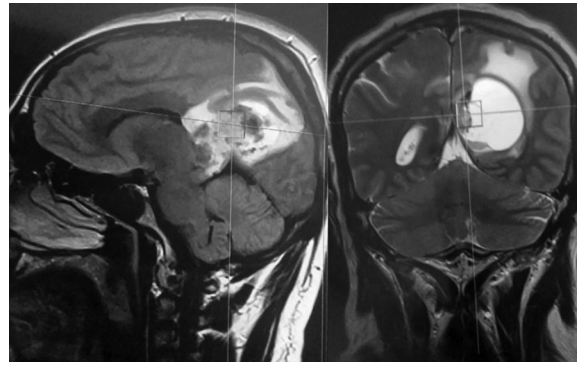


Figure 1. Magnetic resonance where the brain tumor is shown.

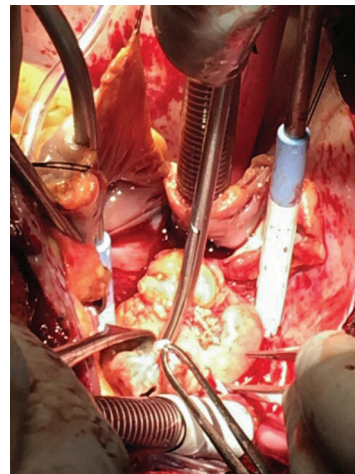


Figure 2. Left atrial cardiac tumor.

cardiopulmonary bypass; high trans-septal approach was carried out, which was expanded with superior vena cava section (Fig. 3). Extensive tumor resection was then performed, which sacrificed the left atrial roof, the mitral valve and annulus, and replacement of the invaded native mitral valve with a mechanical valve (31 mm) (Fig. 4), and left atrial reconstruction were carried out. Plasty was performed using the ipsilateral appendage and superior vena cava anastomosis, as well as atrial septal defect closure, and right atrial closure. Pump withdrawal at the first attempt in nodal rhythm supported by a temporary pacemaker to then go into sinus rhythm; trans-surgical TT-ECHO was requested, where the absence of tumor was observed, as well as adequate prosthetic valve mobility; mediastinal drains were placed and the procedure was completed.

The patient was taken to the post-surgical intensive care unit, where he had pneumonia, acute liver failure, and acute kidney injury, with adequate resolution. He was admitted to the heart surgery area to continue

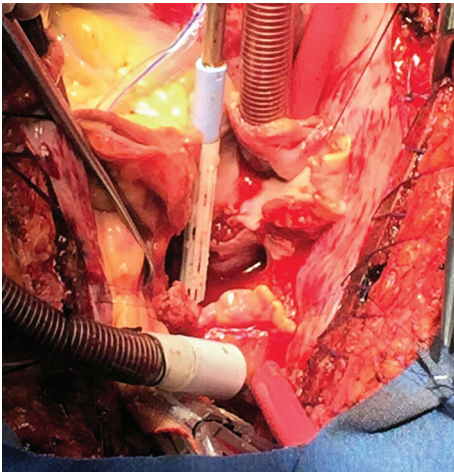


Figure 3. Surgical approach.

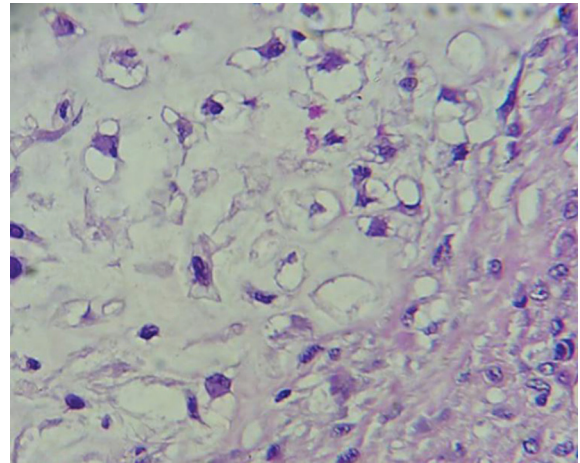


Figure 5. Chondroid metaplasia areas; cells with irregular nuclei immersed in chondroid matrix are observed (40x, hematoxylin-eosin staining).

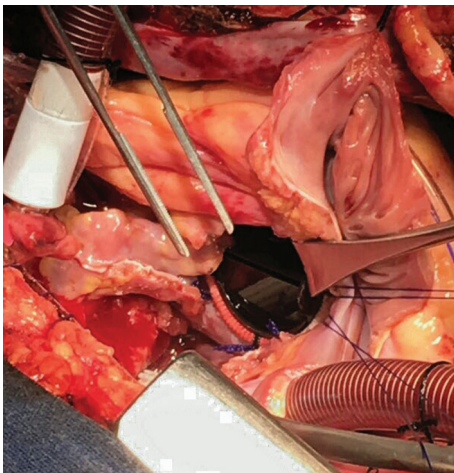


Figure 4. Placement of a 31-mm mechanical mitral valve prosthesis.

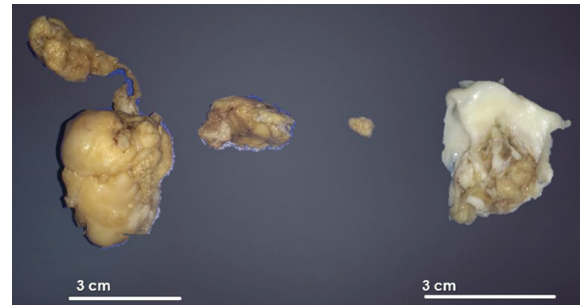


Figure 6. Macroscopic plane: light brown, irregular tumor with polypoid appearance. At right, a segment of the valve affected by tumor proliferation is observed.

treatment and follow-up: the control CT revealed post-operative changes in the left atrium anterior and superior wall without residual tumor tissue being observed, as well as normal-sized cardiac cavities and all four pulmonary veins permeable without filling defects; TT-ECHO without evidence of tumor lesion with normally-functioning prosthetic valve and preserved left ventricular ejection fraction. Home discharge was indicated.

Discussion

Malignant primary cardiac tumors are extremely rare malignancies of varied histopathology. The most common type is sarcoma with intimal subtype⁶, which is the one presented by the patient (high-grade primary intimal atrial sarcoma) (Figs. 5 and 6), the pathological characteristic of which was *MDM2* gene amplification⁷.

In this country, there are two published surgical case series of primary cardiac tumors, with a prevalence of 0.73% and 0.16%, and malignant strain has been found in 12.5% and 16%, respectively; sarcoma is the predominant type^{8,9}. Isolated case reports have been described where clinical presentation includes cardiac symptoms, in addition to rare cases of malignant fibrous lymphomas and histiocytomas¹⁰⁻¹³.

Symptoms of this anomaly are highly varied, with obstruction-associated congestive heart failure, embolization, constitutional symptoms, and arrhythmias specially being observed, although, in this case, the patient started with neurological signs due to brain metastasis, in spite of the fact that left atrial sarcomas generate metastases lately, since they tend to be more solid and less infiltrative than those on the right side³.

Due to its difficult clinical diagnosis, imaging studies can be used to provide morphological information on a cardiac tumor and its functional consequences. The

main goal is to characterize tumor localization, size and vascularity, and hemodynamic consequences and to assess for infiltration. A TT-ECHO should be performed in all patients and can be used to define the relationship with other structures and the exact fixation site. While MRI helps define the tumor grade, vascularity, and infiltration, it also allows, together with echocardiogram, a better perspective for diagnosis and helps to differentiate between malignant and non-malignant tumors². In the reported case, CT helped to identify the tumor site in the mediastinal region to then identify the site of said tumor using TT-ECHO.

In the treatment of malignant tumors confined to the heart that has no infiltration, a complex tumor resection can be performed as, for example, total resection of right cavities, with pulmonary blood flow being assured by means of Fontan circulation. An *ex situ* resection can be carried out in case the tumor involves the left atrial posterior wall or the large vessels. When the tumor completely involves the left side without evidence of metastases, it is plausible using a total artificial heart⁴. In selected cases, heart transplantation is considered the final treatment option once the presence of metastases has been ruled out.

With this patient, the operation was successfully practiced, and complete resection was achieved with adequate margins, confirmed by histopathology examination.

The reported survival for patients with cardiac tumors remains poor. The reported mean is 6-18 months, which is significantly higher when complete resection is achieved in comparison with those who do not have metastases at presentation⁶. The authors carried out a 9-month follow-up at the outpatient clinic with no evidence of recurrence.

Conflicts of interest

The authors declare that they have no conflict of interests.

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Ethical disclaimer

Protection of people and animals. The authors declare that no experiments were performed on humans or animals for this research.

Confidentiality of data. The authors declare that no patient data appear in this article.

Right to privacy and informed consent. The authors have obtained informed consent from the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

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