

Case Report

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Intracardiac Hemangioma - A Rare Primary Tumour with Specifics Radiological Findings

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Abstract

Hemangioma is a rare primary cardiac tumor with an incidence of 2.8% of all cardiac tumors. Found at any stage of life and cardiac chamber, some of these tumors are located in the atrium. Symptoms are not specific (arrhythmias, dyspnea, tamponade, compression of adjacent structures or embolization) and varies according to the location of the tumor and the speed of growth.

We describe the case of a 49-year-old patient who underwent surgical excision under cardiopulmonary bypass of a voluminous intramyocardial hemangioma compressing the right atrium. The mass was first seen on transthoracic echocardiography for the investigation of chest pain and palpitations and was confirmed by cardiac magnetic resonance imaging. Postoperative histopathology revealed an 8 x 7 x 3 cm cavernous hemangioma with some characteristics for an arteriovenous malformation. Due to its location in the right atrium, the patient developed during the postoperative course an atrial fibrillation requiring a long course anticoagulation. The cardiovascular rehabilitation was uneventful. The patient showed no recurrence of the initial symptoms nor the hemangioma during the following two years.

Keywords: Intracardiac Hemangioma; Intracardiac Tumor; Cardiac MRI; Cardiopulmonary Bypass; Cavernous Hemangioma.

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Introduction

Although cardiac tumors are rare entities, an accurate diagnosis is essential to initiate the proper treatment. The fourth edition of the WHO Classification of Tumors of the Lung, Pleura, Thymus and Heart classified them into benign tumors, tumors of uncertain biologic behavior, germ cell tumors and malignant tumors. [1]

Hemangioma is a rare primary benign cardiac tumor and is frequently misdiagnosed as myxomas, the latter being the only relatively common primary heart neoplasm (about 50-80% of cases) [2]

Histopathologic features of cardiac hemangiomas are characterized by benign proliferative endothelial cells lining blood vessels with increasing vascularization. Based on the predominant type of the proliferating vessels, hemangiomas are classified into cavernous, capillary and arteriovenous types. These features are identical to those of hemangiomas elsewhere in the body. They usually grow slower than the malignant tumors but despite its histopathologic benignity is considered to be dangerous due to its risk life-threatening complications like stroke, syncope or sudden death and surgical excision is suggested as first-line therapy.

This case illustrates the importance of a standardized approach to evaluate cardiac masses in order to avoid unnecessary testing and to achieve an accurate diagnosis. [1]

Case presentation

A 49-year-old male patient, active smoker, was admitted for headache associated with left hemifacial hyperesthesia and photophobia. He described several episodes of syncope, increasing dyspnea to New York Heart Association (NYHA) class III and atypical chest pain on exertion for several months. The patient was initially suspected of having migraine with aura and was admitted to our hospital for further evaluation.

Cerebral Magnetic Resonance Imaging (MRI) ruled out ischemic lesions or neoplasia. Transthoracic echocardiography (TTE) showed normal biventricular function without valvulopathy or pericardial effusion. However, it showed a 50 x 67 mm mass located in the right atrial wall without tricuspid or venous outflow obstruction. Cardiac MRI with T1, contrast-enhanced T2, and T1 Fat-Sat sequences confirmed the intramural mass in the right atrium with characteristic features of an intramyocardial hemangioma: A homogeneous lesion with well-defined contours is located posterior to the right atrium and extends 51 x 67 mm axially and 70 mm craniocaudally (Figure 1). The mass compressed the right atrium but did not appear to be endocavitary. It showed hypersignal on T2 sequences, isosignal on T1 sequences, and absence of fat on fat-sat. After gadolinium injection, contrast enhancement begins peripherally, followed by intense homogenization on late sequences. To confirm the diagnosis, the patient underwent a CT scan-guided transparietal and transpulmonary needle biopsy of the mass. The histopathologic results showed a specimen that was too hemorrhagic to be useful, and the procedure was complicated by a discrete pneumothorax and alveolar hemorrhage, which subsequently resolved spontaneously.

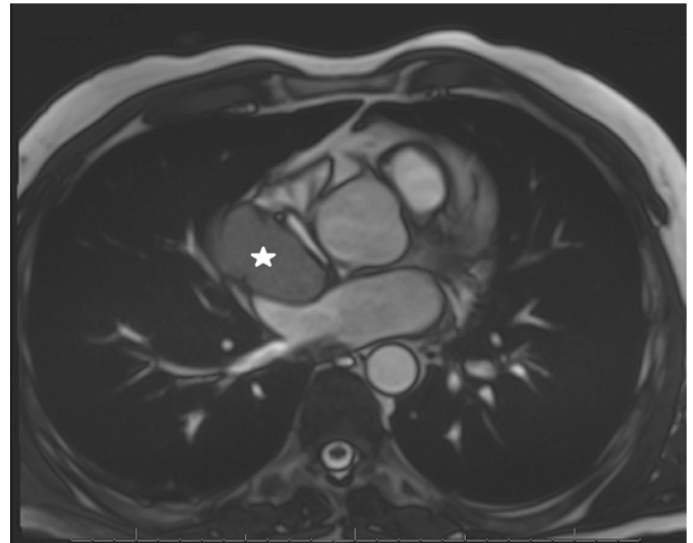


Figure 1: CMR T1-weighted images with heterogeneous, intermediate signal intensity mass (white star)

Surgical resection was planned based on the right atrial wall location, evolving symptoms and future hemodynamic implications. After a median sternotomy, CPB was established between the ascending aorta and a double venous cannulation (superior vena cava and right femoral vein). The aorta was cross-clamped and the mass was resected in toto with preservation of the sinus node area (Figure 2). A pericardial patch replaced a large defect area extending between the two vena cava and the interatrial septum.

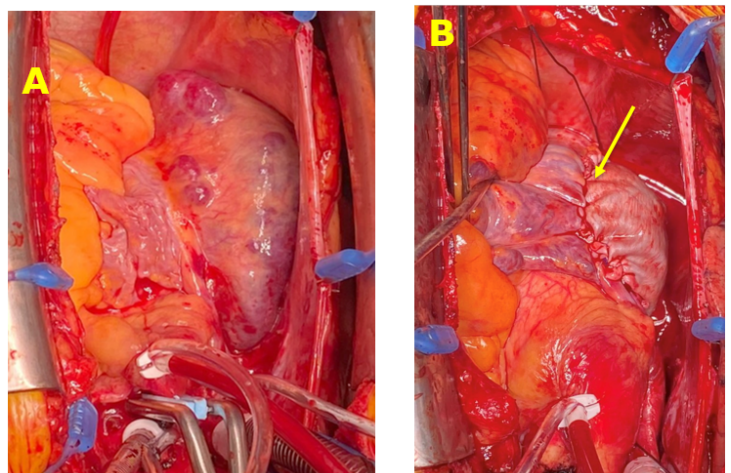


Figure 2: Operative status with the mass located in the right atrium (A). After resection with a normal right atrium and visible sutures (yellow arrow) (B).

The postoperative course was marked by an episode of atrial fibrillation, which was treated with intravenous amiodarone, and postoperative anemia, which required a transfusion of 1 red blood cell concentrate. The patient was discharged from the ICU on day 1 and the remainder of the postoperative course was uneventful. TTE showed non-dilated right cavities with normal right ventricular systolic function, left ventricular ejection fraction (LVEF) of 70% and no pericardial effusion. Histopathology revealed an 8 x 7 x 3 cm encapsulated and irregular mass with dilated vascular structures consistent with a cavernous hemangioma (Figure 3).



Figure 3: Resected hemangioma (8 x 7 x 3 cm)

The patient was discharged on postoperative day 7 with temporary oral anticoagulation. The patient showed no recurrence of the initial symptoms or the hemangioma over the next two years and remains asymptomatic.

Discussion

This case illustrates the management of an intracardiac hemangioma using a standardized diagnostic and therapeutic approach. Hemangioma is a rare primary cardiac tumor with an incidence of 2.8% of all cardiac tumors [3]. It can be found at any age, anywhere in the heart or pericardium, of different types such as cavernous hemangioma, capillary hemangioma and arteriovenous malformation and of different size [4]. Symptoms are nonspecific (arrhythmias, dyspnea, tamponade, compression or embolization) according to their location and growth. A review of 200 cases of cardiac hemangiomas reported sizes varying from 0.5 to 14 cm [5] with an average size of 4.48 cm [6]. The right atrium was the predominant location (26.2%) followed by the left ventricle (23.1%).

Hemangiomas are often misdiagnosed with more common tumors such as myxomas (60-80% of benign tumors), especially when histologically there is a myxoid background. In that same review, only 15 patients (7.5%) were correctly diagnosed preoperatively [2].

Cardiac MRI is the test of choice for diagnosis [7]. It provides information on mass size, shape, location, involvement of surrounding tissues, hemodynamic effects on ventricular and valvular function. Hemangiomas show typical radiological findings. They are heterogeneous, with intermediate signal intensity on T1-weighted images and hyperintensity on T2-weighted images. They also show rapid and strong contrast enhancement on first-pass perfusion imaging and are typically absent on LGE images, which allows them to be distinguished from more common tumors such as myxomas [8]. In the case of our patient, the diagnosis of hemangioma was confirmed on the cardiac MRI, based on those typical characteristics. Thrusting these findings, we could have avoided the needle biopsy, which turned out to be non-contributory and exposed the patient to potentially dangerous complications.

Because of the risk of obstruction, embolization, or sudden death, complete surgical resection remains the gold standard, even for benign tumors.

Conclusion

This case highlights the need for a standardized diagnostic approach to cardiac masses to avoid misdiagnosis and unnecessary, potentially dangerous procedures. The importance of accurate diagnosis and prompt intervention is underscored by the potential for life-threatening complications. Surgical excision remains the gold standard, offering the best chance for resolution of symptoms and prevention of complications.

A multidisciplinary approach involving cardiologists, radiologists, and cardiac surgeons ensures optimal patient care, with early diagnosis and intervention critical to improving outcomes. Further studies are needed to better understand the long-term prognosis of patients with cardiac hemangiomas and to refine surgical and postoperative strategies.

Authors Contributions

CB was responsible for the design, drafting, revision, and approval of the original manuscript for publication.

SC was responsible for the design, drafting, revision, and approval of the original manuscript for publication.

AG was responsible for review and approval of the manuscript for publication.

MC was responsible for review and approval of the manuscript for publication.

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DD was responsible for review and approval of the manuscript for publication.

Conflict of interest statement

No conflicts to declare for any of the authors.

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Informed Consent

Written informed consent was obtained from the patient for this publication.

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