Cardiac hemangioma presenting as a primary cardiac tumor

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Case Report

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Abstract

Cardiac hemangiomas are very rare benign cardiac tumors. They can present at any age and clinical presentation varies according to location and size. In our patient the tumor was located in the left atrium. Left atrial hemangiomas, especially those attached to the left atrium wall, may be mistakenly diagnosed as myxomas. Therefore, we report a rare case of a cardiac hemangioma diagnosed clinically as cardiac myxoma. The tumor was removed surgically for further microscopic examination.

Introduction

Cardiac hemangiomas (CH) are rare primary tumors of the heart and constitute only 2.8% of primary cardiac tumors [1]. Symptoms depend on the tumor's location. The diagnosis can be made clinically through echocardiography, computed tomography (CT), cardiac magnetic resonance (CMR), or combinations of them to rule out other conditions and differential diagnosis. Histopathologic examination is the gold standard for definite diagnosis, while surgical resection is considered the treatment of choice. Microscopically hemangiomas are characterized by benign proliferative endothelial cells lining blood vessels with increased vascularization. The vascular channels are lined by endothelial cells with moderately pleomorphic, sometimes atypical nuclei and focal tuft formation. Mitoses are extremely rare. Electron microscopy shows interdigitating endothelial cell borders without desmosomes or tight junctions. Histopathologic features of CH are identical to those of hemangiomas elsewhere in the body. Based on the predominant type of the proliferating vessels, hemangiomas are classified into cavernous, capillary and arteriovenous types. Cardiac hemangiomas are benign tumors and lack the ability to metastasize, so they usually grow at a slower rate than malignant tumors. Moreover, they are well circumscribed by an outer surface. However, case reports of hemangiomas invading the conductive tissue of the heart suggest that they are unlike the usual benign tumors. Therefore, despite its histopathologic benignity, CH is regarded as clinically dangerous, owing to the risk of life-threatening complications like syncope, stroke, and even sudden death. Owing to its rarity, to our knowledge this disorder is derived from case reports, and no comprehensive review or clinical guideline is available. Therefore, the aim of this study is to provide a comprehensive case analysis of CH based on literature review.

Case Presentation

A 87- years old female was referred to the hospital with complaints of dyspnea, dizziness and weakness for the past few months. The symptoms had deteriorated 3 weeks before her visit to the hospital. Her past history was unremarkable. Transthoracic echo-doppler cardiogram revealed an isoechogenic mass in the left atrium. There was no pericardial effusion.

Physical examination revealed arterial blood pressure 100/80 mmHg, heart rate 120 bpm/min, mild jugular regurgitation, and regular cardiac rhythm without murmurs.
After completing the examinations since there was no significant compression of other cardiac and vascular structures, a preliminary diagnosis was made for a cardiac myxoma with a possible thrombus and was referred for surgical excision. The patient underwent a more detailed ultrasound-cardiogram and she subsequently underwent coronary angiography as a work-up before surgery. The next step in management was surgery and excision of the mass for further microscopic examination.

The surgical intervention was successful, where the fixed mass in the inter-atrial septum was removed without affecting the mitral valve and septum defect.

The mass was sent to the histopathology laboratory for further processing and examination. Macroscopically the lesion resulted in a well-encapsulated mass filled with blood with thin and by moderate extent of somewhat thicker areas of the wall.

Hematoxylin and Eosin (H&E) stained sections showed dilated, congested vascular channels filled with red blood cells and lined with benign endothelial cells, without atypia.

Microscopically, the lesion showed dilation of the engorged vessels comprising multiple thick- and thin-walled blood-filled vascular channels. There was no evidence of atypia or necrosis.

Immunohistochemical CD31 staining of endothelial cells flooring dilated vessels.

Immunohistochemical staining indicated positivity for CD31 and CD34. The endothelial cells lining the cavernous spaces reacted strongly with CD34 and SMA (Smooth muscle actin).

The histological findings along with immunohistochemical results were consistent with benign cavernous hemangioma of the left atrium.

**Discussion**

Hemangiomas are nonmalignant vascular tumors consisting of benign proliferation of network blood vessels arising predominately in the skin, and less commonly involving intra-cardiac location. Histological features of cardiac hemangiomas are similar to that of extracardiac hemangiomas, composed of vascular engorgement channels bordered by non-atypical benign endothelial cells. The lesion may infiltrate the wall compartment including endocardium, myocardium, epicardium, pericardium or it can expand in the whole cardiac tissue and cavity. These rare tumors with ambiguous etiology, can be precisely classified after radiologic examination and meticulous surgical resection. [6, 7] Most cardiac hemangiomas are asymptomatic and discovered incidentally by echocardiography, CT, MRI or at autopsy. Asymptomatic cardiac hemangiomas are also discovered during evaluation of other heart conditions [8]. Cardiac hemangiomas often have combined features of capillary, cavernous, and arteriovenous hemangiomas. Among them, the cavernous and capillary types are reportedly encountered more frequently. The pattern of the tumor can be either displayed as a solitary one with multiple, dilated thin-walled vessels such as in our case report of cavernous hemangiomas or as a combination feature of different histopathology patterns. Papillary fibroelastoma is a tumor arisen from endocardium cells
extended adjacent to the atrioventricular valves. It is believed that PF has predilection for elderly compared to children, due to mechanical stress of calcified valves. Fibromas are usually solitary masses, which originate from connecting tissues and usually are located into interventricular septum. The biopsy shows unencapsulated fibrous tissue proliferation, with sparse capillaries in the center easily invading structure nearby. Also, other rare tumors are reported included arteriovenous malformation tumors of the heart, cricoid aneurism with dysplastic malformation of arteries and veins, lipoma, myxoma, leiomyoma and mesothelioma which are often an incidental finding. [9] According to our literature review, the majority of the cases reported were diagnosed during symptomatic clinical investigations, nevertheless some of them were accidental findings in non-complaint patients. The range of symptoms associated with hemangiomas, depend on the tumor’s location and evolution, including compression and infiltration of the adjacent compartment, rupture, bleeding, embolization and infection.[6,7,8] Atrial hemangiomas, especially those attached to the septum, may be mistakenly clinically and radiological diagnosed as myxomas. However, there are no myxoma cells or ring structures in cardiac hemangiomas, and cellular areas with numerous capillaries are usually present.[8] Some low-grade angiosarcomas may be difficult to be distinguished from hemangiomas. However, lack of mitotic activity, cellular pleomorphism, necrosis, and cellularity can discern a hemangioma from an angiosarcoma.[10] Hemangiomas can also be differentiated from left ventricular thrombi on the basis of their shape, boundary, echogenicity, vascularity, and medical history. Left ventricular thrombi can develop in several situations. Contrast echocardiography was shown to aid and to be a hallmarked device for diagnosis and differentiation of cardiac masses.[7] Unlike other benign tumors, the life cycle of a hemangioma is different in that there is a phase of rapid proliferation that is followed by spontaneous involution. This is especially true for the capillary and cutaneous cavernous varieties while the deep cavernous type almost always never regress and malignant transformation is extremely rare. Surgical resection of the tumor is the treatment of choice and the outcome is usually favorable. Follow-up is recommended to identify any recurrence, even though the rate of recurrence is unknown.

Conclusions

Our patient had a cardiac cavernous hemangioma that presented with nonspecific cardiac symptoms. Identification of the tumor was made with transthoracic echocardiography, but the final diagnosis was made only after surgical resection and further histopathological examination of the tumor. The patient had a favorable outcome after the resection and is under follow-up for the possibility of recurrence.

Declarations

Ethical Approval

Not applicable.

Competing interests
The authors declare that they have no competing interests.

**Authors' contributions**

**Contributions**

Leart Berdica wrote and prepared this manuscript and also made the immunohistopathological diagnosis. Erisa Kola and Daniela Nakuci and Edlira Horjeti jointly provided the analysis and interpretation of pathological findings related to this case. Mehdi Alimehmeti observed and made the necessary regulations and was the supervisor. All authors read and approved the final version of the manuscript.

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**Consent for publication**

The patient provided consent and gave permission to have his case and diagnostic images presented in the medical literature.

**Availability of data and materials**

Free access.

**References**


**Figures**

![Image A](image1.png) ![Image B](image2.png)
Figure 1

Fig. A Transesophageal echocardiography showing a ball-like tumor attached to the anteroseptal wall of the left atrium

Fig. B Coronary angiography - demonstrated normal coronary arteries and a vascular blush through the tumor.

Figure 2

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Figure 3

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