



Cardiac Hemangioma in the Left Ventricular Septum

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Abstract

Keywords

- cardiac
- tumor
- cardiac hemangioma

Background Primary cardiac tumors are an exceedingly rare benign group of tumors that may remain asymptomatic for a prolonged duration or could lead to significant clinical events.

Case Presentation A 64-year-old female patient underwent echocardiography prior to elective knee surgery due to the presence of palpitations and dyspnea. This revealed the existence of a mass located on the left side of the interventricular septum, which was resected successfully.

Conclusion Surgical resection represents the primary therapeutic approach for the management of cardiac hemangiomas. Failure to perform timely resection may elevate the risk of developing total atrioventricular block and experiencing sudden death.

Introduction

Primary cardiac tumors remain a rare group of diseases with an incidence ranging from 0.02 to 0.056%.^{1,2} The majority of these tumors, approximately 75%, are benign.³ Among the benign cardiac tumors, atrial myxoma is the most common, while sarcoma is the most common malignant tumor.⁴ The group of the benign cardiac tumors also includes nonmyxomatous tumors: lipomas, fibromas, fibroelastoma, rhabdomyoma, and hemangiomas. This case report focuses specifically on cardiac hemangiomas, which are extremely rare primary cardiac tumors characterized by the presence of abnormal blood vessels. They can occur in both infants (often associated with Kasabach–Merritt syndrome) and adults, with an incidence ranging from 2.8 to 5% of all primary cardiac tumors.^{5,6} Cardiac hemangiomas can be differentiated based on their localization (intra-, peri-, and paracardial) and origin (endocardium, myocardium, and epicardium).⁵ Morphologically, they can be categorized into cavernous, capillary, and arteriovenous types.⁷ Cardiac hemangiomas

typically present as well-circumscribed, nonencapsulated masses, exhibiting a reddish-purple hue attributable to their vascular nature. They can vary considerably in size and may be associated with surface thrombi due to their propensity to cause turbulent blood flow.^{8,9} Cardiac hemangiomas can occur in various parts of the heart, but the right atrium is the most common, followed by the left ventricle, right ventricle, and left atrium.⁷ Therefore, limited information is available regarding cardiac hemangiomas localized in the septum.

This case report describes a symptomatic cardiac hemangioma in the left ventricular septum.

Case Report

A 64-year-old female patient presented at the hospital with a diagnosed vascularized tumor extending from the inferoseptal to the anteroseptal region of the left ventricular septum. The patient was scheduled for elective knee surgery.

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Prior to the surgical procedure, a cardiac evaluation, including echocardiography, was conducted due to instances of rapid palpitations and shortness of breath. These symptoms had been occurring intermittently over the past few weeks, with the initial occurrence reported approximately 2 years before admission.

Upon emergency medical examination in the emergency room (ER), no signs of myocardial ischemia or arrhythmia were observed. Echocardiography revealed a $2.6 \times 1.8 \times 1.3$ cm mass situated on the left side of the interventricular septum (►Fig. 1). The left ventricular ejection fraction was within normal range, and no evidence of left ventricular obstruction was noted. A positron emission tomography and computed tomography (PET-CT) scan was conducted as an additional examination to confirm the finding. It indicated mild metabolic activity within the left ventricular mass (►Fig. 2). On the day of admission, the patient was in a stable hemodynamic condition, with laboratory results indicating preexisting chronic kidney disease of grade 3a. Other laboratory values, including troponin levels, fell

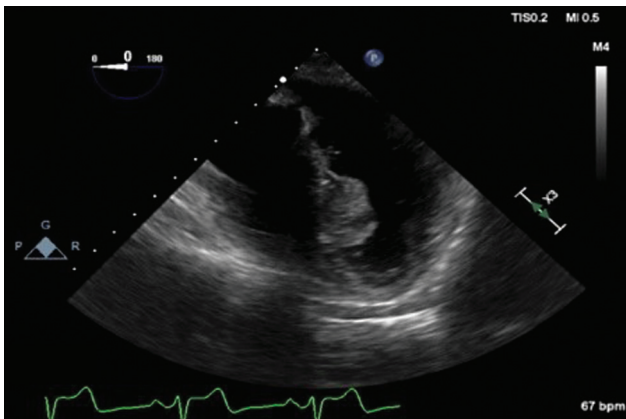


Fig. 1 Preoperative transesophageal echocardiography confirms the tumor at the left ventricular septum, measuring 3×2 cm (projection at the mid-esophageal level).



Fig. 2 Preoperative positron emission tomography computed tomography (PET-CT) shows the cardiac mass, which appears to be connected to the left ventricular septum with mild metabolic activity.



Fig. 3 Macroscopic view. The largest dimension of the specimen was 4.2 cm. The surface appeared to be covered by the endothelium and the myocardial muscle. The cut surface showed a well-demarcated spongiotic, reddish colored lesion.

within the normal range. Surgery was performed on the patient on the subsequent day via a median sternotomy. After aortic and right atrial cannulation, a cardiopulmonary bypass was established. Myocardial protection was achieved using antegrade and retrograde cardioplegia techniques. A transverse aortotomy was performed to visualize the tumor, which extended around 1.2 cm from the aortic annulus on the muscular part of the ventricular septum to approximately 3.4 cm toward the apex. Extraction of the tumor mass was done with forceps from the septum. After being grasped with forceps, the mass experienced a slight collapse and minor bleeding. Complete in toto resection of the tumor was easily achieved, and the specimen was sent for further pathological analysis (►Figs. 3 and 4). An intraoperative frozen section confirmed the diagnosis of a benign cardiac hemangioma. Closure with a pericardial patch was not deemed necessary, and no residual ventricular septal defect (VSD) was observed. Following successful weaning from the cardiopulmonary bypass without any arrhythmias, sternal closure was performed. Postoperatively, the patient was transferred to the intensive care unit (ICU) for 1 day, followed by 1 day in the intermediate care unit (IMCU) and 11 days in the regular unit. The postoperative electrocardiography (ECG) displayed a left bundle branch block, which exhibited slight regression on the day of discharge. Subsequent echocardiography demonstrated the absence of a residual mass. After a total of 14 days of hospitalization and an uneventful recovery, the patient was discharged to a rehabilitation clinic. Subsequently, after 1 year of follow-up, there were no indications of cardiac events or tumor recurrence.

Pathological Findings

Macroscopic and histopathological analyses of the extracted mass were performed (►Figs. 3 and 4). Upon macroscopic examination, the tumor presented as well-defined, encapsulated, reddish-blue masses (►Fig. 3). Immunohistochemical markers such as Ki-67 and ERG were employed to assess the tumor's vascularity and growth potential^{8,9} (►Fig. 4). ERG, a

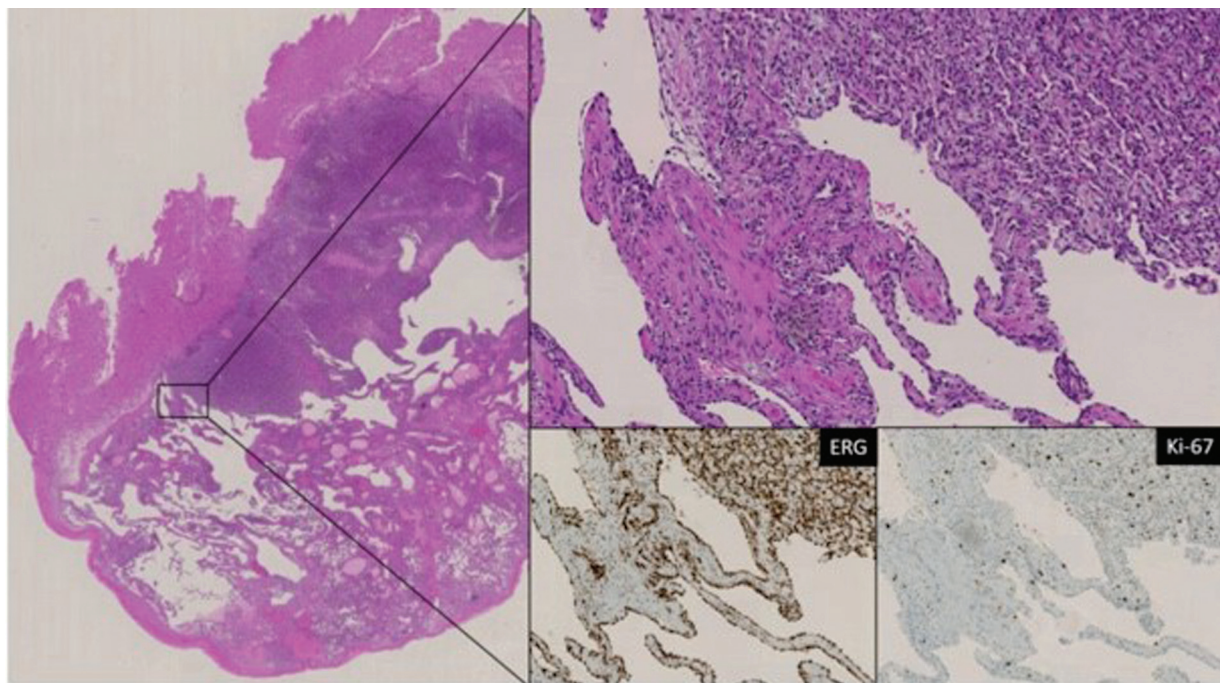


Fig. 4 Histopathological view. On overview, histology shows a well-demarcated lesion with capillary and cavernous areas. The lining is characterized by flat cells without atypia and immunohistochemical confirmation of endothelial differentiation. The Ki-67 immunohistochemistry underlines the low proliferative nature of the lesion.

member of the ETS transcription factor family, is frequently utilized as an endothelial marker in the context of tumor angiogenesis and vascular neoplasms.⁹ In cardiac hemangiomas, a lower Ki-67 labeling index is typically observed, reflecting the benign and generally slow-growing nature of these tumors.⁹ By employing both Ki-67 and ERG in a complementary manner, pathologists can determine the proliferative rate and evaluate the vascular component of the cardiac tumor.

Discussion

A cardiac hemangioma is a rare benign tumor that can remain asymptomatic for an extended period or manifest with mild symptoms. Its growth exhibits a slow progression, and there is no documented potential for metastasis. Nevertheless, depending on their location and size, cardiac hemangiomas have been linked to various complications, including stroke, thromboembolic events, angina, syncope, dyspnea, arrhythmias, pericardial effusion, right ventricular outflow tract obstruction, heart failure, atrioventricular block, and even sudden death.⁸ According to Li et al,⁷ who analyzed 200 cases of cardiac hemangioma, tumors situated in the intra-atrial or interventricular regions are associated with a higher risk of total atrioventricular block and sudden death. Therefore, despite the rarity of cardiac hemangiomas, imaging diagnostics such as echocardiography, CT, magnetic resonance imaging, and PET-CT should be conducted if suspicion arises. Upon confirmation of the diagnosis, surgical resection is recommended.¹⁰ Depending on the location of the tumor, the surgical approach should vary. In this particular case, a left transatrial approach could also be considered, especially

since the tumor was situated close to the septal leaflet of the mitral valve. This procedure can be performed through a median sternotomy or minimally invasively via a lateral thoracotomy. The approach provides excellent exposure, allowing for safe tumor extraction with minimal manipulation.¹¹ Moreover, given the unstable nature of cardiac hemangiomas, the establishment of a safe extraction technique is imperative. Panos and Myers described a small series using a video-assisted approach and a “laparoscopic basket.”¹² Furthermore, with the development of minimally invasive video-assisted and endoscopic cardiac surgery, these approaches may also need to be considered, depending on the location of the cardiac tumor. Small study series by Panos and Myers,¹² Ko et al,¹³ and Tang et al¹⁴ have described cases of left atrial and biatrial cardiac myxoma extraction using minimally invasive methods, with no limitations and excellent results.

In summary, surgical removal is a safe and feasible approach for treatment of cardiac hemangiomas. Depending on the location and surgeon's preference, different surgical methods could be employed. Minimally invasive techniques are increasingly utilized due to their safety, reduced trauma, shorter recovery times, and improved cosmetic outcomes. However, data on surgical treatment for cases of cardiac hemangioma are very limited, and further data are required to determine the best therapeutic options.

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None.

Conflict of Interest

None declared.

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