

Right ventricular haemangioma as a rare cause of chest pain: a case report

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Background

Cardiac haemangioma is a rare primary cardiac tumour. Most patients with cardiac haemangioma have no typical symptoms, and some may present with non-specific manifestations, such as shortness of breath, heart palpitations, or cardiac insufficiency, making it difficult to distinguish cardiac haemangioma from other diseases. We report a case of cardiac haemangioma that present with chest pain. This haemangioma was finally completely excised to relieve the patient's symptoms and avoid poor prognosis.

Case summary

A 14-year-old boy presented with an intermittent and progressive non-exertional chest pain for 2 weeks. Echocardiography showed a space-occupying mass at the right ventricular apex, which was later confirmed by computed tomography angiography and magnetic resonance imaging (MRI). The mass was successfully resected, and postoperative pathology confirmed a cardiac cavernous haemangioma. The patient had an uneventful postoperative recovery at the 8-month follow-up.

Discussion

Cardiac haemangioma is a benign tumour with no typical clinical manifestations, and very few patients may present with chest pain. Preoperative echocardiography, computed tomography, and MRI are helpful for diagnosis, and surgery can relieve symptoms and may improve the prognosis of patients with cardiac haemangioma.

Keywords

Cardiac haemangioma • Echocardiography • Magnetic resonance imaging • Chest pain • Surgery • Case report

Introduction

Primary cardiac tumours are very rare, and the literature is dominated by case reports. They are found incidentally in 0.002–0.03% at autopsy, and ~75% of cardiac tumours are benign, among which haemangiomas accounted for only 2.8%.¹ Most patients with cardiac haemangiomas have no symptoms, and some may present with non-specific manifestations, such as decreased exercise endurance, shortness of breath,

heart palpitations, and cardiac insufficiency, making it difficult to distinguish cardiac haemangioma from other diseases. At present, multimodality imaging is the main method to diagnose cardiac haemangioma, and there are no clear treatment guidelines for haemangioma. According to previous case reports, most asymptomatic cardiac haemangiomas are treated conservatively, while symptomatic cardiac haemangiomas are surgically removed.

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Learning points

- Cardiac haemangioma is a rare benign cardiac tumour that can present with chest pain as the only manifestation. Therefore, in cases of patient presenting with chest pain without a family history or risk factors of heart disease, a diagnosis of cardiac mass may be considered.
- Even though cardiac haemangiomas demonstrate high signal intensity on T2-weighted images in most cases, it can also show mixed iso- and low signal intensity on T2-weighted imaging in tumours that have less sinusoids.
- Surgical resection is the treatment of choice for symptomatic cardiac haemangioma and is considered to be curative in most cases.

Timeline

Time	Events
Day 1	Admission due to patient's progressive chest pain. Transthoracic echocardiography suggested a space-occupying lesion in the right ventricle with a normal cardiac function (left ventricular ejection fraction = 67%).
Day 2	Rechecked echocardiography identified a large, hyperechoic, well-circumscribed mass located in the right ventricular apex.
Day 3	Computed tomography pulmonary angiography found a round-shaped tumour in the right ventricular apex, which was iso-density, attached to the interventricular septum.
Day 4	Cardiac magnetic resonance imaging showed a right ventricular apex mass with well-defined borders, with mixed iso- and low signal intensity on T2-weighted imaging.
Day 6	Surgical excision was performed.
Day 7–13	The postoperative rechecked electrocardiogram, echocardiogram, and laboratory results were unremarkable and the patient discharged on Day 13.
8 months	The postoperative recovery was uneventful, and coronary computed tomography angiography showed no abnormality.

Case presentation

A 14-year-old boy presented with an intermittent and progressive non-exertional chest pain for 2 weeks. There were no associated shortness of breath, dizziness, palpitations, or a history of syncope or presyncope. Transthoracic echocardiography in the primary care clinic showed a mass in the right ventricle, and the patient was then referred to our cardiology department. His breathing rate and blood pressure were normal. On physical examination, the apical impulse was not palpable without obvious heaves or thrills, and the first and second heart sounds were normal without additional heart sounds, murmurs, or frictional sounds. Lungs were clear on auscultation with no adventitious sounds. The laboratory results were unremarkable. He had no cardiovascular risk factors, such as drug or alcohol use or a family history of heart disease. He also denied recent infectious symptoms or a history of chest trauma. The electrocardiogram on

admission indicated sinus rhythm with an average heart rate of 84 b.p.m., unremarkable PR and QRS intervals, normal axis, and no evidence of chamber enlargement or myocardial ischaemia. Repeated transthoracic echocardiography showed a solitary, well-demarcated, hyperechoic mass attached to the right ventricular apex (*Figure 1*, white arrow; *Videos 1* and *2*). Computed tomography pulmonary angiography performed to rule out pulmonary thromboembolism revealed a round-shaped tumour (10 mm × 8 mm) in the right ventricular apex that was isodense and attached to the interventricular septum (*Figure 2*, black arrow). Cardiac magnetic resonance imaging (MRI) showed a mural ventricular mass with well-defined borders and mixed iso- and low signal intensity on T2-weighted imaging (*Figure 3*, white arrow).

Surgical excision was performed under cardiopulmonary bypass conditions (*Figure 4A*; *Video 3*). Cardioplegia was performed with del Nido solution after cross-clamping the ascending aorta. The surgeon made a 5 cm incision on the right atrium, and then the right ventricle was completely observed via the tricuspid valve. A round 10 mm × 12 mm tumour mass was found in the right ventricular apex adjacent to the interventricular septum. Finally, the entire tumour and its base tissue were successfully resected (*Figure 4B*), and cardiac haemangioma was confirmed via histopathology, in which it was characterized by a disorganized collection of blood vessels (*Figure 4C*). The surgery was successful, and postoperative electrocardiogram and laboratory results were unremarkable. The postoperative echocardiogram showed normal right ventricular size and function. The patient was discharged on the 7th postoperative day and had an uneventful postoperative recovery at the 8-month follow-up.

Discussion

Cardiac haemangiomas are vascular tumours composed of capillaries or cavernous vascular channels that develop from endothelial cells, and most are benign. Histologically, cardiac haemangiomas are heterogeneous, resembling cavernous or capillary haemangiomas most frequently and intramuscular haemangiomas or arteriovenous malformations less frequently.² In this case, the mass was histopathologically confirmed as a cavernous haemangioma.

Cardiac haemangiomas can affect patients of all ages and can occur in any part of the heart but most commonly involve the right atrium and the left ventricle.³ Right ventricular cardiac haemangiomas are relatively uncommon, and most are located in the anterior wall.⁴ In a study of 30 cases of right ventricular haemangioma, the masses were located in the right ventricular apex in only 2 cases, in which patients had no symptoms.⁴

Cardiac haemangiomas are often clinically insignificant and are mostly diagnosed incidentally. In general, the symptoms produced by

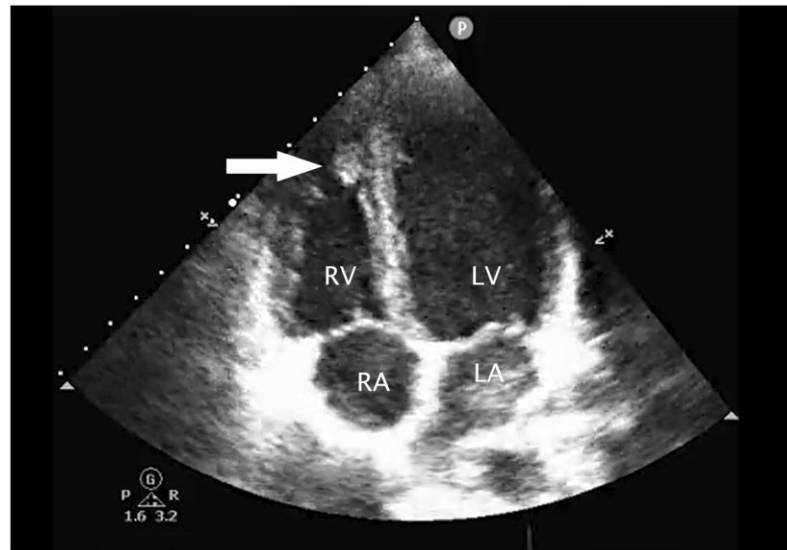


Figure 1 Admission echocardiogram showing a solitary, well-demarcated, hyperechoic mass (white arrow) attached to the right ventricular apex.



Video 1 Two-dimensional transthoracic echocardiography from the apical four chamber view.



Video 2 Magnified image of the right ventricle.



Figure 2 Computed tomography pulmonary angiography showing a round-shaped tumour (10 mm × 8 mm) in the right ventricular (black arrow) adherent to the interventricular septum and with regular borders, suspected to be benign.

tumours do not depend on their histology but on their location, scope, and growth speed. Clinical symptoms include decreased activity endurance, shortness of breath, heart palpitations, coronary insufficiency, atypical angina, outflow tract obstruction, or even sudden death.³ When they grow in the interatrial septum or atrioventricular node, they can cause conduction disturbances, atrioventricular block and a variety of rhythm disturbances. When the tumour grows in the left ventricle, the patient may present with symptoms of angina and systemic embolism events, and a small number of patients may develop ventricular arrhythmias. Epicardial or intrapericardial tumour growth may cause pericardial effusion and tamponade or angina pectoris by compression of the coronary arteries.^{5,6} In this case, the

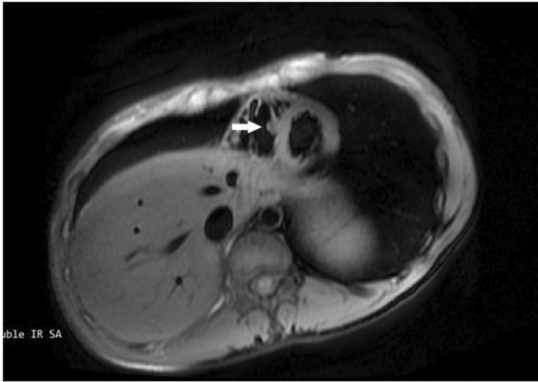
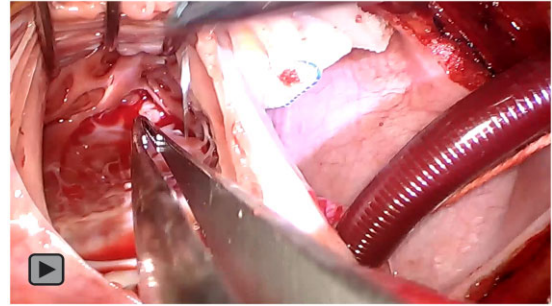


Figure 3 Cardiac magnetic resonance imaging showing the mass (white arrow) with a mixed iso- and low signal intensity on T2-weighted imaging.



Video 3 Surgical excision under cardiopulmonary bypass conditions.

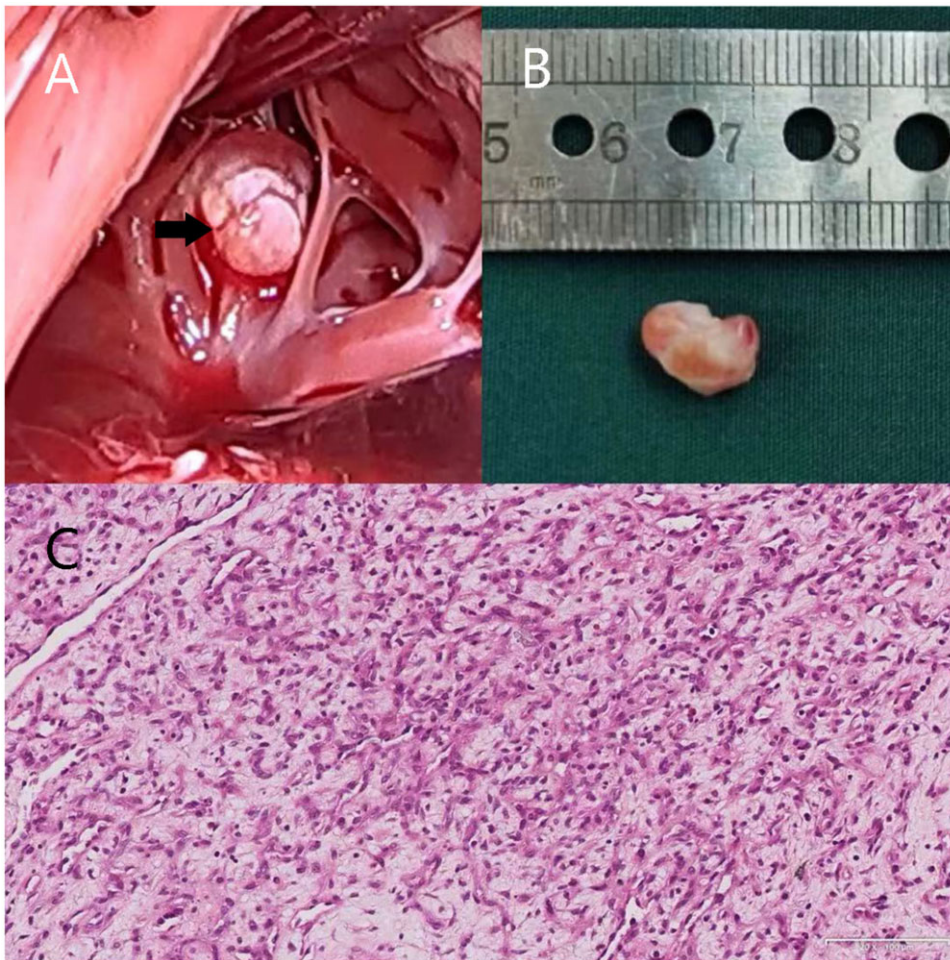


Figure 4 Right ventricular haemangioma (black arrow) seen during the operation through an incision on the right atrium (A). The excised tumour has a smooth and yellowish surface with little bleeding (B). Cardiac haemangioma was confirmed by histopathology following surgical removal, in which it was characterized by a disorganized collection of blood vessels (haematoxylin-eosin staining, $\times 40$) (C).

haemangioma was located in the right ventricular apex near the interventricular septum, and the patient's main complaint was chest pain.

Transthoracic echocardiography are the best means for primarily evaluating cardiac haemangiomas. The typical echocardiographic findings in cardiac haemangioma are hyperechoic lesions. On MRI, the extent of intramural development can be detected more accurately, and the blood supply and tissue characteristics of haemangiomas can also be determined. On cardiac MRI, the tumours demonstrate intermediate and high signal intensity on T1- and T2-weighted images, respectively. Cardiac haemangioma with high signal intensity on T1- and T2-weighted images has also been reported.⁷ Via gadolinium infusion, rapid homogeneous enhancement is characteristic of capillary haemangioma due to the high vascularity of the tumour, while cavernous haemangioma shows a progressive enhancement. In this case, the cardiac haemangioma demonstrates a mixed iso- and low signal intensity on T2-weighted imaging, which has never been reported before. This may be related to less sinusoids and more solid components in the tumour, which was confirmed in pathology.

In patients with haemangioma, coronary angiography is used to evaluate the feeding arteries to the tumour and tumour blushes, which occur as pooling of contrast medium in the sinusoids or vascular lakes within the tumour; however, cavernous haemangiomas have large vascular spaces with slow flow and therefore typically do not enhance at angiography. Considering the young age of the patient in our case, there were no risk factors for coronary heart disease, the diagnosis of haemangioma by transthoracic echocardiography and cardiac MRI was relatively clear, and the patient had indications for surgery. No coronary angiography was performed preoperatively. However, coronary computed tomography angiography (CTA) during follow-up was normal, which indicated that the cardiac haemangioma was a local lesion and had not affected the coronary arteries. In the described case report, although the accurate mechanism of the patient's chest pain was not clear, coronary steal represented the likely cause of the patient's symptoms because no coronary impingement was found on coronary CTA.

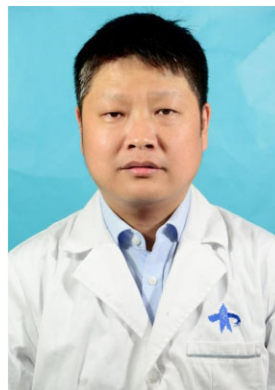
Clinically, it is important to distinguish benign and malignant tumours, and haemangiomas have to be differentiated particularly from angiosarcomas, which are also highly vascularized. The latter often presents as invasive hyperplasia to adjacent tissues, such as the myocardium and pericardium, which can be well observed on MRI.⁸ As shown in our case, benign tumours are often round in shape without lobules or burrs. For angiosarcoma, the typical presentation on MRI is a mosaic pattern on T1 images, which is due to the mixture of connective tissue structures, vascularized areas and bleeding into the tumour tissue.⁸

Clinically, propranolol is the drug of choice for the treatment of liver or skin haemangiomas, and there have been successful cases of oral propranolol in the treatment of infant cardiac haemangiomas.^{9,10} However, there is no alternative treatment for adult cardiac haemangioma except surgery. Many scholars believe that cardiac haemangioma has the possibility of embolism, rupture and sudden death, therefore, in symptomatic children and adults, surgical resection is recommended. Benign cardiac tumours such as a haemangioma in childhood have an excellent prognosis when completely excised and appear to have a good short-term prognosis even following subtotal excision.^{9,11} In this case, the chest pain was completely alleviated after surgery, and the patient did well during the scheduled 8-month follow-up.

Conclusion

Cardiac haemangioma is often misdiagnosed or ignored due to asymptomatic or atypical clinical presentation; however, echocardiography, computed tomography, and cardiac MRI can offer valuable insights into this disorder, and pathologic examination is the gold standard. Although the surgical indication for cardiac haemangioma remains controversial, the outcomes in our case and most reported cases involving surgical excision were favourable over the years.

Lead author biography



Fengwei Guo, MD, is attending doctor of cardiovascular surgery. He is engaged in cardiovascular surgery medical and research work. He has carried out a number of normal and minimally invasive cardiac surgery and extracorporeal life support (ECMO) for critically ill patients.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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We would like to thank our patient's family for allowing us to present this case and to use the patient's photographs.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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