Research Square

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Diagnostic Mystery — a Rare Right Ventricular Cardiac Hemangioma: a Case Report

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Case report

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Abstract

Background: Cardiac hemangiomas are rare in all kinds of benign cardiac tumors. Although cardiac hemangiomas affect all ages and may occur anywhere within the heart, right ventricular hemangiomas are extremely uncommon.

Case presentation: We report a 56-year-old woman presented with chest tightness and breath shortness for 3 months. Transthoracic echocardiography and coronary computed tomography angiography showed a mass located adjacent to the apex of the right ventricle but both failed to figure out where the mass originated from, remaining a diagnostic mystery preoperatively. The mass was removed successfully and the histopathological examination confirmed it was hemangioma.

Conclusions: Cardiac magnetic resonance should be the ultimate diagnostic tool of cardiac tumors. Surgical removal, associated with a low recurrence rate and long-term survival benefits, should be the first choice of therapy for cardiac hemangiomas.

Background

Cardiac hemangiomas, one kind of benign heart tumors, with an incidence of fewer than 2% of all cardiac tumors are exceptionally rare[1]. Although cardiac hemangiomas affect all ages and may occur anywhere within the heart, right ventricular hemangiomas are extremely uncommon[2, 3]. We report one of these rare cases and discuss the diagnosis of heart tumors in the literature.

Case Presentation

A 56-year-old female presented with chest tightness and breath shortness for 3 months. Given the symptoms have gradually worsened, she went to the local hospital where transthoracic echocardiography showed an elliptically hypoechoic mass, measuring about 4.0×2.8cm, located adjacent to the apex of the right ventricle (RV) in the pericardial cavity. It seemed originated from the pericardium and had an unclear border with normal right ventricular myocardium, additionally, no obvious blood flow signal had been detected in the mass. Re-transthoracic echocardiography in our institution identified the size of the mass as 4.0×3.0cm (Fig. 1A), which was similar to the result in local hospital. Coronary computed tomography angiography (CTA) was implemented for further diagnosis, but it seemed to fail to figure out where the mass originated from. It showed an infiltrating tumor appears to be connected to the RV(Fig. 1B), meanwhile the tumor showed a clear boundary with the RV on another image (Fig. 1C), suggesting its pericardial origin. It was diagnosed preoperatively as a tumor originating from the pericardium and invading the RV through discussion, but the malignancy could not be excluded completely. Consequently, the surgery was performed via a median sternotomy under cardiopulmonary bypass. After the pericardium was incised, we found that the tumor(4cm×3cm×3cm) originated from the apex of the RV and adhered to the pericardium, rather than from that as previously speculated (Fig. 2A). A part of the capsule protruded out of the heart, and the rest integrated and communicated with the RV myocardium.

The normal ventricular myocardium at the boundary between the two was thin. There was solid mass in the tumor, filling with bloody cyst fluid and there was no myocardial ischemia and reverse heartbeat. The tumor was completely removed and the defect in the right ventricle was sutured and enforced with a patch of autologous pericardium along the incision line (Fig. 2B). The post-surgery pathological diagnosis was reported to be hemangioma (Fig. 2C, D). The patient had an uneventful recovery without any complication and was discharged on the postoperative 7th day. No signs of recurrence and right heart dysfunction was observed by thoracic echocardiography during 4 months of postoperative follow-up.

Discussion

Cardiac hemangiomas represent only 1 ~ 2% of all benign heart tumors[1]. Most affected patients are asymptomatic and the symptoms depends on the tumor's location and size, which are always non-specific, such as dyspnea, arrhythmias, angina, signs of right heart failure and thromboembolic events[4, 5]. Consequently, cardiac hemangiomas are often discovered incidentally by transthoracic echocardiography and misdiagnosed as other cardiac neoplasms(e.g. cardiac myxoma)[6]. Right ventricular hemangiomas are extremely rare, especially that at the apex of the RV. According to Jiang's summary[3], the most common site of right ventricular hemangiomas is the anterior wall of the RV, but only 6.7% are located at the apex of the RV. In our case, the hemangioma is located at the apex of the RV and it grows outward reaching to 4.0cm, different from most cardiac hemangiomas which are single, relatively small subendocardial nodules(2.0 ~ 3.5cm)[7]. These characteristics make it very unusual and significantly increase the difficulty of diagnosis.

Currently, the diagnostic tools for cardiac tumors mainly include echocardiography, chest computed tomography (CT) and cardiac magnetic resonance (CMR) imaging. Transthoracic echocardiography has been the preferred diagnostic tool for cardiac tumors because of its non-invasiveness and convenience, however, it cannot distinguish the tissue level very accurately. Besides, it cannot display the blood supply of the tumor unless contrast-enhanced ultrasound is applied. Considering that there was no obvious blood flow signal in the mass on echocardiography, so we were apt to deem it was non-cardiac origin preoperatively. Contrast-enhanced CT may make up for these shortcomings, but it is unfriendly to patients who are allergy to contrast agents or with renal insufficiency. Coronary CTA and coronary angiography are also used for showing the distribution of vessels, feeding vessels to the tumor and whether the coronary arteries are oppressed[8]. In our case, the origin of the tumor remained a mystery in result of two coronary CTA images associated with key information of the origin revealing quite different findings, leaving us in a diagnostic dilemma. If we used CMR at that time, we might be able to figure out the property of the tumor and its precise relationship with the RV anterior free wall and the pericardium. The excellent contrast resolution and multiplanar capability of CMR imaging allows qualitative diagnosis and optimal anatomical evaluation of any cardiac tumor. In addition, CMR imaging enabled us to demonstrate the precise relationship among the tumor, tricuspid valve, and RV anterior free wall, which was useful for pre-surgical planning[5]. However, the implantation of pacemakers or metal objects such as biliary stent, and the high price limit the application of CMR in our country.

With regard to treatment, surgical removal is the first choice of therapy for cardiac hemangiomas[9]. After a complete resection, the prognosis is generally favorable with a low recurrence rate. Even an incomplete resection is reported to produce long-term survival benefits[4].

Conclusions

In our case, the hemangioma at the apex of the RV is extremely rare, and its diagnosis is so difficult that preoperative echocardiography and CT have not figure out the property of the tumor. CMR should be the ultimate means of diagnosis-difficult cardiac tumors. Our operation was very successful, and there was no recurrence and other complications in the subsequent follow-up.

Abbreviations

RV: right ventricle; CTA: computed tomography angiography; CT: computed tomography; CMR: cardiac magnetic resonance;

Declarations

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Authors' contributions

JF and LG performed and compiled the case and contributed to article writing. PT, XD and QZ were major contributors in article writing and image analysis. SW and YN performed the case and reviewed the report. All authors read and approved the final manuscript.

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Availability of data and materials

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Ethics approval and consent to participate

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Consent for publication

Written informed consent of clinical detail and image publication was obtained from the patient.

Competing interests

None declare.

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