

Left Ventricular Hemangioma

Thomas Strecker, MD,¹ Axel Schmid, MD,² Thorsten Zielesinski, MD,¹ Johannes Rösch,¹
Abbas Agaimy, MD³

¹Center of Cardiac Surgery, ²Department of Radiology, and ³Institute of Pathology, Friedrich-Alexander-University
Erlangen-Nuremberg, Germany

ABSTRACT

Cardiac hemangiomas are quite rare benign tumors of vascular origin often detected incidentally during routine examinations. Here we present the diagnostic evaluation and excisional biopsy of such a cardiac tumor in a 20-year-old man.

INTRODUCTION

Cardiac hemangiomas are extremely rare and mostly documented as isolated case reports. They comprise 5% to 10% of benign tumors in individuals from all age groups and may produce a variety of symptoms or be asymptomatic. Histologically, cardiac hemangiomas may display a wide range of morphology including cavernous, capillary, and mixed variants, similar to their peripheral counterparts.

CASE REPORT

A 20-year-old man with an unclear left ventricular (LV) mass was referred to our hospital for further diagnostic evaluation. The mass was incidentally detected by transthoracic echocardiography at the medical examination for the Federal Armed Forces. The patient had been in his usual state of excellent health before admission to hospital.

At admission, physical examination was unremarkable. Transesophageal echocardiography showed a large mass (50 × 50 mm) within the posterior wall of the left ventricle, but also involving parts of the inferior and lateral wall (Figure, A). The heart valves appeared normal without any signs of regurgitations or stenosis. The echo showed no impairment of either left ventricular or right ventricular systolic function. On electrocardiogram (ECG)-gated dual source computed tomography (CT), the mass had slight patchy contrast enhancement on arterial phase and distinct homogeneous enhancement on delayed imaging (Figure, B1 and B2). Cardiac magnetic resonance imaging

(MRI) was performed for further tissue characterization. The exact location of the tumor and the relationship to surrounding structures was demonstrated on steady-state free precession imaging. A diagnosis of probable cardiac hemangioma was suggested based on typical signal intensity (Figure, C1 and C2) and contrast uptake characteristics of the lesion (Figure, C3) indicating a very high degree of vascularization. Invasive coronary angiography demonstrated that the left ventricular mass was mainly supplied by end branches of the second obtuse marginal branch (Figure, D). The patient was taken to the operating theater, where a left lateral thoracotomy was performed, and an excisional biopsy of the ventricular mass at the left lateral wall was obtained. Because of the extensive growth and infiltration, it was impossible to excise the tumor mass completely, not even via a median sternotomy.

The pathological specimen was composed of small fragments of fibrofatty epicardial tissue and adjacent myocardium. The lesion was composed of a mixture of dysplastic venous vessels showing severe degrees of fibromuscular dysplasia that obliterated the vascular lumina (Figure, E1). Isolated normal-looking arteries were seen as well as transition to closely packed capillary-size channels (Figure, E2). The adjacent myocardium contained numerous dilated thin-walled blood vessels diffusely dissecting through the myocardium and seemingly communicate with the aberrant thick-walled epicardial vessels (Figure, E3). There was no evidence of malignancy. Immunohistochemical stains were consistent with a blood vessel origin, but not a lymphatic origin of the vessels. Furthermore, no reactivity for HMB45 was detected, thus excluding a lesion from the family of lymphangioleiomyomatosis. Based on these findings, the diagnosis of mixed cardiac hemangioma with arteriovenous and capillary components was made.

Because the patient had no further symptoms associated with the cardiac hemangioma and radical operation could not be performed, we decided to observe him under medical care without further surgical intervention. A repeated echocardiography and MRI 6 months after leaving the hospital showed similar results, without any evidence of progressive tumor growth.

DISCUSSION

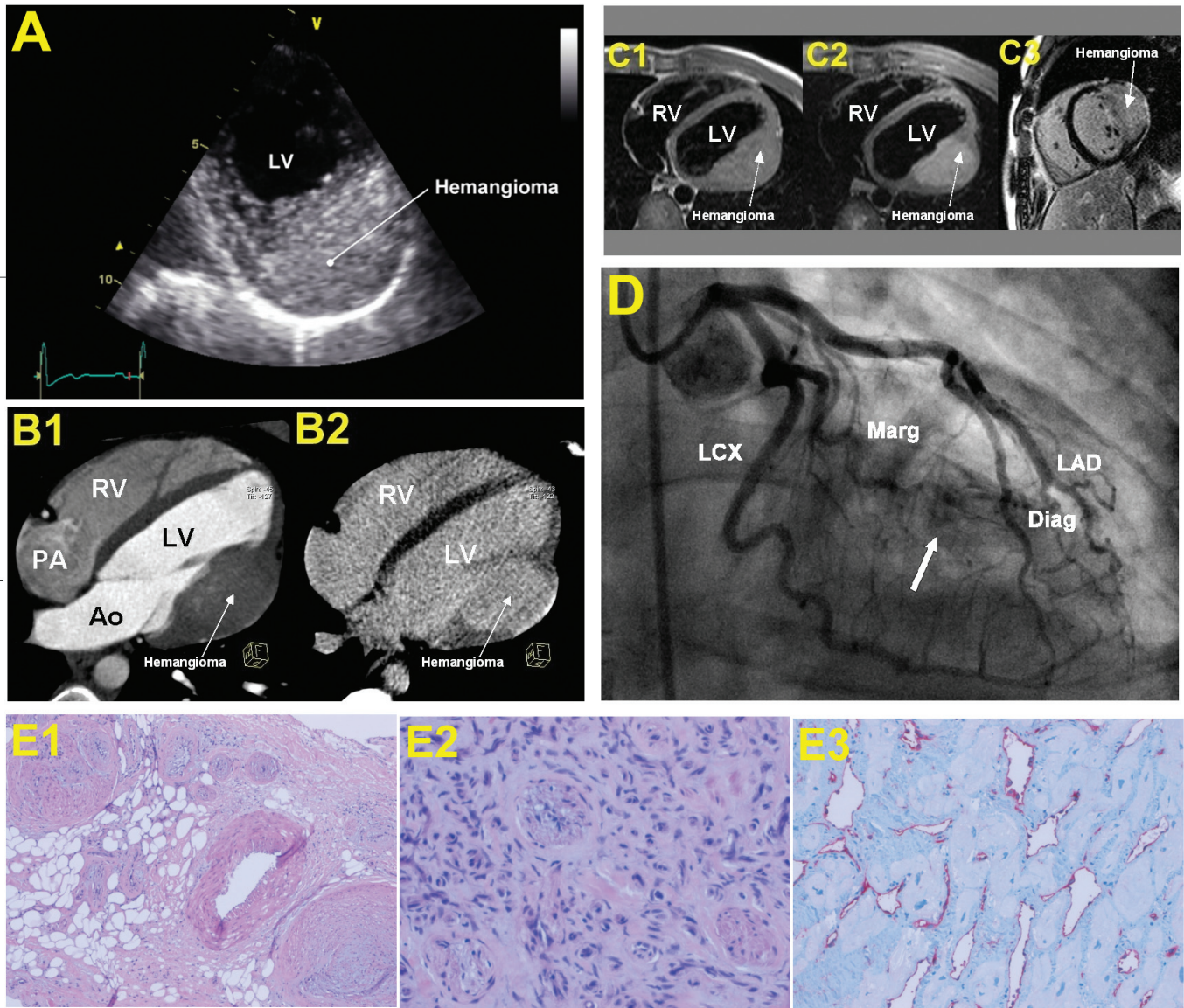
Primary cardiac tumors are very rare entities among heart diseases. Their incidence in autopsy series varied from

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Correspondence: Thomas Strecker, MD, Center of Cardiac Surgery, Friedrich-Alexander-University Erlangen-Nuremberg, Krankenhausstrasse 12, 91054 Erlangen, Germany; +49-9131-85-33985; fax: +49-9131-85-36088 (e-mail: thomas.strecker@uk-erlangen.de).

0.001% to 0.03% [Centofanti 1999]. About 75% of primary cardiac tumors are benign, with atrial myxomas being the most common. Other benign cardiac tumors are fibromas, rhabdomyomas, pericardial cysts, lipomas, hamartomas, and teratomas. The remaining 25% are malignant, and the most

common malignant tumors are various sarcomas: myxosarcoma, liposarcoma, angiosarcoma, fibrosarcoma, leiomyosarcoma, osteosarcoma, and rhabdomyosarcoma [Burke 1996]. Cardiac hemangiomas are extremely rare and mostly documented as isolated case reports. They account for 5% to 10%



A, Transthoracic echocardiography: parasternal short axis showing the large mass infiltrating the posterior wall of the left ventricle, but also involving parts of the inferior and lateral wall. B, Dual phase multi-detector computed tomography: B1, First-pass imaging depicts initial contrast uptake of the central parts of the mass; B2, Delayed enhancement acquisition shows homogeneous contrast storage of the whole tumor. C, Electrocardiogram-gated magnetic resonance imaging: C1, T1-weighted fast spin echo imaging in a long-axis view shows an isointense mass in the lateral wall of the left ventricle; C2, The mass is hyperintense on T2-weighted triple inversion recovery imaging; C3, Delayed enhancement imaging in a short-axis view, obtained 10 minutes after administration of 0.1 mmol/kg gadolinium-based contrast agent, demonstrates homogeneous intense contrast enhancement of the tumor. D, Invasive coronary angiography: the left ventricular mass (arrow) is mainly supplied by end branches of the second obtuse marginal branch. E, Histology: E1, An overview of cardiac "mixed hemangioma" showing multiple venous vessels with remarkable fibromuscular intimal dysplasia surrounded by an artery and epicardial fibrofatty tissue; E2, High-power view of the capillary components illustrating numerous small capillary-size vessels interrupted by obliterated "dysplastic" venous vessels; E3, Dilated thin-walled vessels dissecting through the myocardium and lined by swollen bland-looking endothelium highlighted by the vascular endothelial marker CD31 (no staining for the lymphatic endothelial marker podoplanin was detected). LV indicates left ventricle; RV, right ventricle; Ao, aorta; LCX, left circumflex; Marg, ramus marginalis; Diag, ramus diagonalis; LAD, left anterior artery.

of all benign cardiac tumors. Anatomically, they may occur in any part of the heart, but they are more commonly found in the lateral wall of the left ventricle (21%), the anterior wall of the right ventricle (21%), and the interventricular septum (17%) [Brizard 1993].

These benign cardiac tumors may present few non-specific or no clinical symptoms, and they often present themselves so insidiously that their diagnosis and therapy is delayed. Occasional cases can cause intracardiac blood flow obstruction, valvular dysfunction, arrhythmias, pericardial effusions with tamponade, and peripheral embolization with systemic deficits. Some cardiac tumors induce no symptoms and become evident as incidental findings, as in the present case.

Although transthoracic echocardiography is useful in the initial evaluation of suspected cardiac tumors, transesophageal echocardiography is frequently required for a more comprehensive and accurate assessment [Alam 1995]. In our case, the mass was thought to be a thrombus initially. Nevertheless, the location of the mass was not common for a thrombus. They generally involve the apex of the left ventricle, most often in the presence of akinesis or dyskinesis. The differential diagnosis included a cardiac myxoma, although myxomas usually occur in the left atrium in 75% of the cases; only 5% of them develop in the left ventricle [Tanrikulu 2009]. For further evaluation of the mass, we performed CT and MRI scans. CT adequately demonstrates the morphology, location, and extent of a cardiac mass, and its main advantage over echocardiography is in its depiction of the pericardium, great vessels, and other structures allowing the physician to look for associated extracardiac disease, including metastases [Dawson 1990]. Furthermore, the utility of MRI in the evaluation of cardiac masses is well established. The wide field of view, high contrast and spatial resolution, and multiplanar imaging capacities allow precise demonstration and location of a mass, including its anatomic relationship to the cardiac chambers and any involvement of the myocardium, pericardium, or contiguous structures [Araoz 2000].

In our case, we decided to try surgical resection because of the diagnostic uncertainty about the natural course of the tumor. Intraoperatively, the tumor showed a diffuse, poorly circumscribed pattern with involvement of both epicardial fat and adjacent myocardium. Therefore, we avoided radical

surgery and decided to perform conservative regular medical controls by echocardiography, CT, and MRI.

In the majority of cases, cardiac tumors require operative excision. Surgery should be recommended to prevent potentially life-threatening complications [Moniotte 2005; Ediae 2009]. Most benign cardiac tumors form well-circumscribed nodular masses, and they can thus be resected completely; only a few tumors cannot be completely excised because of their huge size, and only tumor debulking may be feasible for such extensive tumors [Pigato 1998]. Fortunately, our patient was asymptomatic without any evidence of progressive tumor growth at 6-month follow-up.

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