A Very Rare Cardiac Hibernoma in the Right Atrium: A Case Report

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ABSTRACT

Cardiac lipomas are rare tumors often detected incidentally during routine examinations. They usually remain asymptomatic for a long time and cause arrhythmia, heart valve dysfunction, or embolization in the later stages. In this article, we report a case of a 64-year-old patient with a very rare cardiac hibernoma located in the right atrium. Transesophageal echocardiography and computed tomography have been shown to be useful for differentiating between benign and malignant tumors in order to plan surgery. The treatment of choice for these tumors is resection. The tumor was excised with the use of cardiopulmonary bypass surgery. Histology confirmed diagnosis of a benign cardiac hibernoma.

CASE REPORT

A 64-year-old Caucasian woman with symptoms of heart failure was referred for evaluation of a right atrium (RA) mass. She noted progressive dyspnea, fatigue, arterial hypertension, dizziness, and one instance of vision disturbance. At admission into hospital, physical examination was otherwise unremarkable. The patient presented a blood pressure of 140/90 mmHg and a heart rate of 80 beats per minute (bpm). On auscultation she had a faint 2/6 systolic murmur heard best at the right medial sternal border. The electrocardiogram (ECG) showed sinus rhythm. The chest radiograph was normal without pulmonary congestion, infiltration, effusion, or pneumothorax. The laboratory tests were within the normal range. The cardiac catheterization study showed normal coronary arteries without significant stenoses and normal left

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ventricular ejection function (75%) with the following pressures: left ventricle end-diastolic 10 mmHg and aorta 165/85 mmHg. Transesophageal echocardiography (TEE) revealed a large tumor mass projected down from the superior portion of the RA with attachment to the atrial septum and the superior vena cava (SVC) measuring 5.6×2.1 cm (Figure 1). The heart valves appeared not pathological without any regurgitation or stenosis. The TEE showed no impairment of either left ventricular or right ventricular systolic function. Computed tomography (CT) revealed an inhomogeneous RA mass extending 4.5 cm in diameter. It was located cranial in the RA and produced a narrow passage of the SVC in the cavity (Figure 2).

The patient was taken to the operating theater, where a median sternotomy was performed and cardiopulmonary bypass was installed via aorto-bicaval cannulation with moderate hypothermia (33°C) and antegrade application of cold crystalloid cardioplegia (4°C, Custodiol; Köhler Chemie, Alsbach, Germany) after clamping of the aorta. After vertical opening of the RA, the implantation of the tumor could be seen subendocardiac close to the atrial septum, extending cranial from the SVC, underneath the lateral wall of the RA to the nodus atrioventricularis. The tumor did not reach the tricuspid valve. The tumor mass appeared refractile, sessile, and was hardly excised. A few biopsies were taken and revealed fatty connective tissue in the immediate sectioning analysis. The defect in the RA wall was closed with a patch of bovine glutaraldehyde-fixed pericardium. A patent foramen ovale was closed with a direct suture line. The patient was successfully weaned from cardiopulmonary bypass without any signs of cardiac failure. First the ECG showed ventricular escape rhythm and later AV-node rhythm. Because of ineffective atrium and ventricular stimulation, an epimyocardiac 2-chamber pacemaker insertion was considered. At the end of surgery the patient was transferred to the intensive care unit in a stable hemodynamic condition (blood pressure 100/60 mmHg, heart rate 80 bpm in pacemaker rhythm) with 0.055 μg/kg per minute norepinephrine and 6.9 µg/kg per minute dobutamine. It was possible to taper and withdraw inotropic support with stable hemodynamic parameters over the following hours. The patient was extubated 9 hours after surgery and transferred back to regular care on postoperative day 5.

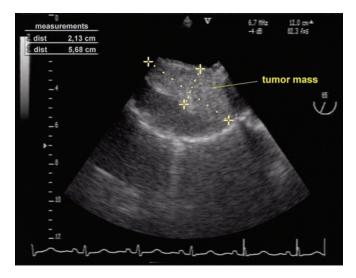


Figure 1. Transesophageal echocardiography showing the right atrium mass on the superior aspect of the atrial septum.

Histological examination of the tumor mass resulted in a diagnosis of hibernoma without evidence of malignancy. The resected tissue consisted of multiglobular, vacuolated fat cells with remnants of cardiomyocytes and granular, eosinophilic cytoplasm intermingled with mature fat cells (Figure 3).

Before the patient left the hospital, pacemaker control revealed a good ventricular sensing and pacing, but an exit bloc in the atrium. Because of a high-frequency AV-node rhythm and a difficult new implantation in the atrium, the pacemaker was converted to only ventricular stimulation. Echocardiography showed a normal right and left ventricular function with an EF of 60%, a good contractility of all areas, normal heart valves, and no relevant pericardial effusion. A further TEE control 6 months after surgery confirmed the previous findings. There were no intracardiac thrombi and the atrium septum had lightly thickened.

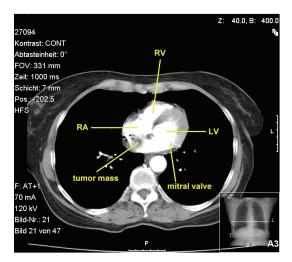


Figure 2. Transversal computed tomography of the cardiac hibernoma. RA indicates right atrium; RV, right ventricle; LV, left ventricle.

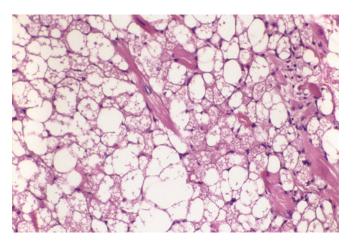


Figure 3. Histology of the resected tissue reveals multiglobular, vacuolated fat cells with remnants of cardiomyocytes and granular, eosinophilic cytoplasm intermingled with mature fat cells.

DISCUSSION

Primary cardiac tumors are very rare entities among heart diseases. They occur in 0.001% to 0.03% according to reports of autopsy series (Centofanti 1999). About 75% of primary cardiac tumors are benign, atrial myxomas being the most common. Other benign cardiac tumors are rhabdomyomas, fibromas, hemangiomas, pericardial cysts, lipomas, hamartomas, and teratomas. The remaining 25% are malignant and the most common malignant tumors are various sarcomas: myxosarcoma, liposarcoma, angiosarcomas, fibrosarcoma, leiomosarcoma, osteosarcoma, and rhabdomyosarcoma (Vander Salm 2000).

Clinical symptoms of these cardiac tumors are absent or not specific and often present themselves so insidiously that their diagnosis and therapy is delayed. They 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 (Schrepfer 2003).

Radiologic evaluation of cardiac tumors usually begins with chest radiography, which occasionally reveals abnormal findings including cardiomegaly, signs of heart failure, abnormalities of cardiac contour, and pleural effusions. The further diagnosis and management of these neoplasms has been greatly facilitated by the development of noninvasive cardiac imaging (Grebenc 2000). Although transthoracic echocardiography is useful in the initial evaluation of suspected cardiac tumors, TEE is frequently required for a more comprehensive and accurate assessment (Alam 1995). CT adequately demonstrates the morphology, location, and extent of a cardiac neoplasm, 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 cardiac-gated magnetic resonance imaging in the preoperative evaluation of cardiac masses is well established. The wide field of view, high contrast, and spatial resolution and multiplanar imaging capabilities allow precise demonstration and localization of a mass, including its anatomic relationship to the cardiac chambers and any involvement of the myocardium, pericardium, or contiguous structures (Freedberg 1988).

In the majority of cases, these tumors require operative excision. Surgery should be recommended to prevent potentially life-threatening complications. Most benign cardiac tumors can be resected completely; only a few, because of their huge size, cannot be, and only tumor debulking may be possible. Although the long-term prognosis of asymptomatic cardiac tumors is often good, fatal histories have been reported for untreated symptomatic tumors. Therefore, if a cardiac tumor is found to cause symptoms, indication for operation should be liberal (Courtis 2004).

In conclusion, the presented case shows a patient with a very rare cardiac hibernoma in the RA. Cardiac tumors produce a large variety of symptoms. The most useful diagnostic tools are echocardiography and CT, which in almost all cases precisely locates the tumor and defines its extent. The treatment of choice for these cardiac tumors is surgical resection.

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REFERENCES

Alam M, Rosman HS, Grullon C. 1995. Transesophageal echocardiography in evaluation of atrial masses. Angiology 46:123-8.

Centofanti P, Di Rosa E, Deorsola L, et al. 1999. Primary cardiac tumors: early and late results of surgical treatment in 91 patients. Ann Thorac Surg 68:1236-41.

Courtis J, Marani L, Amuchastegui LM, Rodeiro J. 2004. Cardiac lipoma: a rare cause of right-to-left interatrial shunt with normal pulmonary artery pressure. J Am Soc Echocardiogr 17:1311-14.

Dawson WB, Mayo JR, Muller NL. 1990. Computed tomography of cardiac and pericardial tumors. Can Assoc Radiol J 41:270-5.

Freedberg RS, Kronzon I, Rumancik WM, Liebeskind D. 1988. The contribution of magnetic resonance imaging to the evaluation of intracardiac tumors diagnosed by echocardiography. Circulation 77:96-103.

Grebenc ML, Rosado de Christenson ML, Burke AP, Green CE, Galvin JR. 2000. Primary cardiac and pericardial neoplasms: radiologic-pathologic correlation. Radiographics 20:1073-103.

Schrepfer S, Deuse T, Detter C, et al. 2003. Successful resection of a symptomatic right ventricular lipoma. Ann Thorac Surg 76:1305-7.

Vander Salm TJ. 2000. Unusual primary tumors of the heart. Semin Thorac Cardiovasc Surg 12:89-100.