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Enormous Right Atrial Hemangioma in an Asymptomatic Patient: A Case Report and Literature Review

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We report the case of an enormous right atrial hemangioma in an asymptomatic 42-year-old woman. The diagnosis was made by echocardiogram after the patient was found to have an abnormal EKG during a routine medical exam. The hemangioma is the largest described in English literature. The tumor was surgically resected and the patient did well postoperatively. In this case report, we discuss the discovery of the tumor and treatment of our patient. A brief discussion of cardiac hemangiomas follows. (ECHOCARDIOGRAPHY, Volume 26, September 2009)

echocardiography, *benign cardiac tumor*, *right atrium*, *hemangioma*

Case Report

A 42-year-old asymptomatic woman with a normal physical exam was referred for echocardiogram after an abnormal electrocardiogram (EKG) was obtained during a routine primary care visit. The EKG had revealed normal sinus rhythm and T-wave abnormalities. Transthoracic and transesophageal echocardiography showed a very large (8.0 × 7.0 cm) mass filling almost the entire right atrium, except for the area adjacent to the tricuspid valve orifice. The mass had a homogeneous core and relatively smooth borders (Fig. 1A and Video 1). It did not extend into the venae cavae. After intravenous injection of perflutren lipid microsphere contrast (Definity, Lantheus Medical Imaging, North Billerica, MA, USA) there was brisk opacification of the mass. This finding indicated that the mass was highly vascular in nature (Fig. 1B and Video clip 2).

Chest radiograph revealed a bulge on the right-hand side of the heart (Fig. 2). A CT image obtained immediately after the administration of intravenous contrast shows a large right atrial mass (Fig. 3A). A delayed CT image obtained approximately 5 minutes later (Fig. 3B) shows that the tumor has enhanced to the same degree as the blood pool indicating that it is highly vascular. This pattern of enhancement (no central enhancement immediately after intravenous contrast administration with delayed enhancement isodense to the blood pool) is typical of hemangiomas.[18] F-fluorodeoxyglucose (FDG) PET/CT fusion imaging (Fig. 3C) showed no hypermetabolic activity in the tumor consistent with a benign lesion. Upon entering the pericardium at the time of surgery, a large protruding mass was readily visible from the right atrium. It encompassed most of the lateral wall and appeared to involve part of the interatrial septum. After initiation of cardiopulmonary bypass, an incision was made in the right atrial wall just at the rim of the atrial tumor. There was a large intraatrial component. The atrium was removed from the tumor medially, leaving a very small portion of the lateral wall of the atrium and the right atrial appendage. The entire tumor was slowly excised en bloc and a portion of the intraatrial septum was removed. The tumor measured 11.0 × 6.5 × 4.5 cm and weighed 170 g (Fig. 4A).

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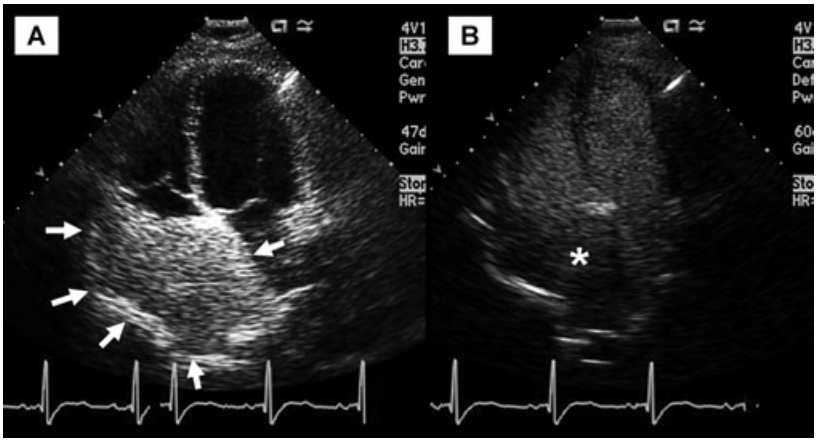

Figure 1. *Transthoracic Echocardiogram* ***A.*** *Noncontrast transthoracic echocardiogram in apical four-chamber view showing a homogenous mass filling the entire right atrium (arrows).* ***B.*** *Transthoracic echocardiogram showing brisk opacification of the right atrial mass (asterisk) after intravenous perflutren lipid microsphere contrast (Definity) injection.*

Bovine pericardial patch extending from the superior to the inferior vena cava with a large bulbous center was used to reconstruct the atrium and the lateral aspect of the superior vena cava. Surgical pathology revealed hemangioma (Fig. 4B). The patient remains stable on follow-up examinations.

Discussion

Primary tumors of the heart are rare with an incidence range at autopsy of less than three cases per 1,000 autopsies (0.0017–0.27%).

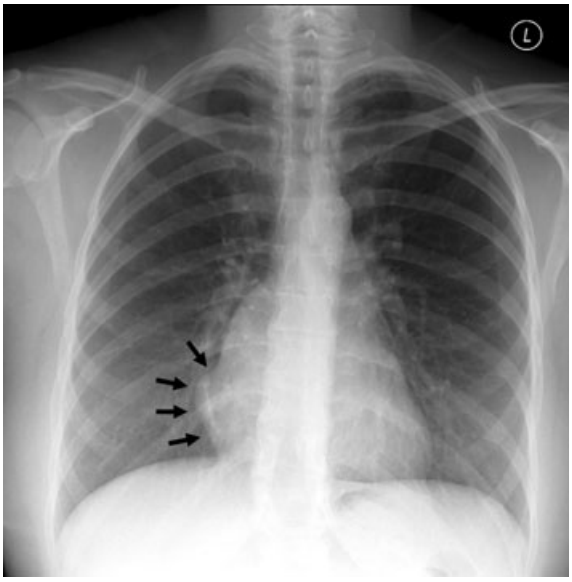

Figure 2. *Chest radiograph showing a bulge on the right-hand side of the heart (arrows) due to large right atrial hemangioma.*

Seventy-five percent of these primary cardiac tumors are benign with hemangiomas accounting for 2.8% of them.[1] Hemangiomas represent the benign proliferation of endothelial cells, which forms channels of blood in a nonencapsulated but well-demarcated structure without necrosis.[2] Cardiac hemangiomas may occur at any anatomic location within the heart and can originate from the pericardium, endocardium, or myocardium. Most hemangiomas have been reported to be small, endocardial nodules measuring between 2 mm and 3.5 cm in diameter. They may be polypoid or sessile, are usually solitary, and lack the features of infiltration.[3] Histologically, they fall into one of the three classifications: (1) cavernous (multiple, dilated, thin-walled vessels), (2) arteriovenous (dysplastic malformed arteries and veins), and (3) capillary type (smaller capillary type vessels).[4]

Hemangiomas can occur in any age group and tend to show a female predominance. They may be asymptomatic or they may present with a vast array of symptoms. Presenting the symptoms depends primarily on the tumor location, size, and extension.[5] Some of the symptoms attributed to hemangiomas include heart failure, pericardial effusion, sudden death, dysrhythmia, coronary insufficiency, and valvular stenosis.[6] Intracavitary tumors are known to potentially produce a triad of obstruction, embolization, and constitutional symptoms.[7]

There are several available modalities that allow for incidental and purposeful diagnosis of cardiac hemangiomas. On a routine chest radiograph there may be cardiac enlargement, irregular heart border, and abnormal appearance of other intrathoracic structures. Abnormal EKG may also lead to the incidental discovery of

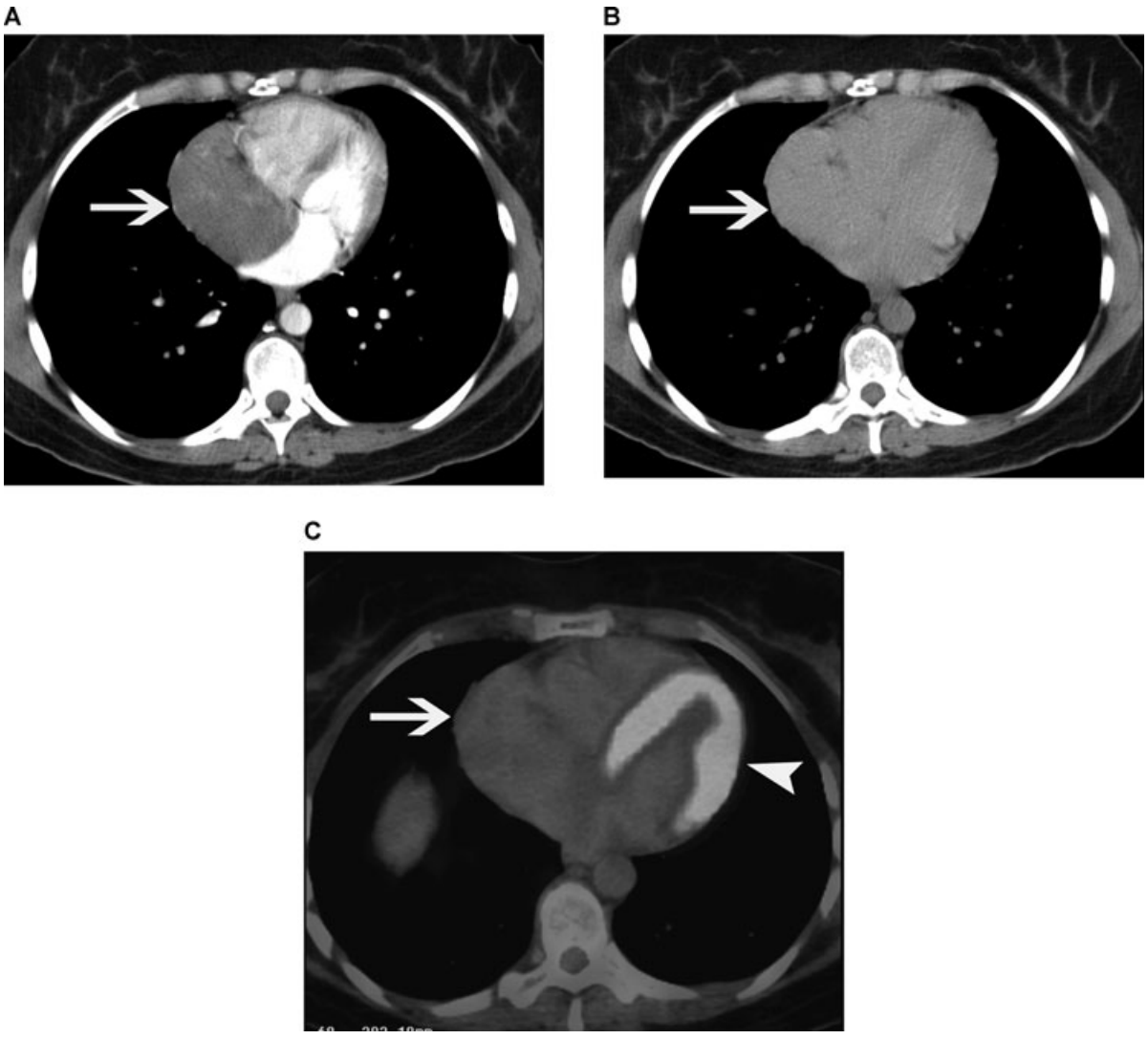

Figure 3. *Computed tomography (CT) and FDG PET/CT images.* ***A.*** *CT image obtained immediately after administration of intravenous contrast shows large right atrial mass (arrow).* ***B.*** *Delayed CT image obtained about 5 minutes later shows that the tumor (arrow) has enhanced to the same degree as the blood pool indicating that it is highly vascular. This pattern of enhancement (no central enhancement immediately after intravenous contrast administration with delayed enhancement isodense to the blood pool) is typical of hemangiomas in the liver also.* ***C.*** *^{18}F-fluorodeoxyglucose (FDG) PET/CT fusion image shows no hypermetabolic activity in the tumor (arrow) consistent with a benign lesion. Note normal activity in myocardium of left ventricle (arrowhead).*

cardiac hemangiomas (as in our patient).[8] Cardiac catheterization and angiography can localize a tumor within the chambers of the heart and outline neovascularity, respectively.[9] However, these methods are invasive and may prove to be hazardous as they lead to the risk of tumor embolization.[8] CT and magnetic resonance imaging (MRI) may also be useful in identifying the vascular origin of a cardiac tumor. CT, unlike MRI, detects calcification in tumors, is faster, easier to perform, and produces a more reliable image. MRI, on the other hand, allows for a better selection of imaging planes and better soft-tissue contrast as compared to CT. Diagnosis has been aided with the advent of two-dimensional echocardiography. In general, it is the most appropriate screening and diagnostic imaging modality for detecting cardiac tumors. It is particularly useful in screening for and diagnosing atrial tumors. Echocardiography has

Figure 4. *Surgical Pathology* ***A.*** *Gross pathological specimen of the self-contained tumor with smooth borders.* ***B.*** *Histological examination of tumor shows dilated, thin-walled vessels filled with blood. The findings are indicative of cavernous hemangioma.*

been found to be safe, reliable, cost-effective, and noninvasive.[6] It helps in assessing the accurate location, size, shape, mobility, and extracardiac extension. It is also useful for visualizing all four chambers of the heart simultaneously and assessing cardiac function.[8]

Cardiac hemangiomas have an unpredictable outcome. They may proliferate indefinitely or stop growing and involute. They may remain asymptomatic or patients may suffer sudden death as a result of any of the previously mentioned complications.[3] Hemangiomas in general usually have a good prognosis.[10] A complication of surgical resection is the risk of tumor recurrence after excision.[10] For patients undergoing surgical excision, follow-up with regular echocardiography should be maintained, as this will allow for the detection of tumor recurrence.[4] Nonresectable benign tumors tend to have a poor prognosis due to complications such as ventricular arrhythmias and sudden death.[10]

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Video clip 1. Noncontrast transthoracic echocardiogram in apical four-chamber view showing a homogenous mass filling the entire right atrium. (Codec mpeg-2)

Video clip 2. Transthoracic echocardiogram showing brisk opacification of the right atrial hemangioma after intravenous perflutren lipid microsphere contrast (Definity) injection. (Codec mpeg-2)

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