

## Cardiac Spindle Cell Hemangioma: A Case Report<sup>1</sup>

Ju Young Lee, M.D., In Jae Lee, M.D., Kwang Sun Min, M.D.<sup>2</sup>, Eui Yong Jeon, M.D.,  
Yul Lee, M.D., Sang Hoon Bae, M.D.

Spindle cell hemangioma is an uncommon vascular lesion histologically resembling a cavernous hemangioma and Kaposi's sarcoma with a predilection for the extremities. There are no radiologic reports concerning cardiac spindle cell hemangioma in the current literature. We report here a case of cardiac spindle cell hemangioma.

**Index words :** Heart, neoplasms  
Hemangioendothelioma  
Heart, CT

Primary cardiac neoplasms are rare lesions that affect patients of all ages and have a reported prevalence rate of 0.001 - 0.03% in autopsy series and a rate of 0.15% in echocardiatic series (1). The majority of primary cardiac tumors are benign. The most common primary cardiac neoplasm is a myxoma, which accounts for approximately half of all cases. Other benign primary tumors include the papillary fibroelastoma, rhabdomyoma, fibroma, hemangioma, paraganglioma, and lipoma (1).

A spindle cell hemangioma is an uncommon vascular lesion that exhibits a predilection for the extremities (2 - 4). It has been previously described as a spindle cell hemangioendothelioma histologically resembling a cavernous hemangioma and Kaposi's sarcoma. It is now regarded as a benign neoplasm that is often associated with vascular anomalies. To the best of our knowledge, its occurrence in the intracardiac space has not been previously reported. Here we report a case of cardiac spindle cell hemangioma.

### Case Report

A 39-year-old female presented to the Emergency Department of our hospital complaining of general weakness, dyspnea, cold sweating, generalized edema and abdominal discomfort. The patient had suffered from occasional shortness of breath for a month. However, no treatment was given before the symptoms had become aggravated 15 days prior and the patient was unable to walk more than three steps at a time. The patient had been hospitalized at a local clinic for five days, but the symptoms persisted and progressed. During the last month, the patient gained about 6 kg of body weight due to generalized edema. At presentation in our hospital, the patient had an extremely ill looking appearance with dyspnea and complained of abdominal discomfort. She had no known underlying disease and was a healthy housewife before this episode.

An initial chest radiograph revealed cardiomegaly without any evidence of abnormal pulmonary vasculature affecting the right side of the heart. A precontrast computed tomography (CT) scan revealed heterogeneously high attenuation of a large mass filling the right cardiac atrium and ventricle (Fig. 1). No calcification was noted in the mass. On a contrast enhanced CT scan, a

<sup>1</sup>Department of Radiology, Hallym University College of Medicine

<sup>2</sup>Department of Pathology, Hallym University College of Medicine

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Address reprint requests to : In Jae Lee, M.D., Department of Radiology, Hallym University College of Medicine, 896 Pyungchon-dong, Dongan-gu, Anyang-city, Kyungki-do 431-070, Korea

Tel. 82-31-380-3885 Fax. 82-31-380-3878

E-mail: ijlee2003@yahoo.co.kr

lobulated solid mass with slightly heterogeneous peripheral enhancement was seen (Fig. 2). No significant central enhancement was noted which suggested internal hemorrhagic necrosis. Cardiac muscle invasion with pericardial extension of the tumor was suspected at the anterior wall of the right atrium. Extensive reflux of contrast material was noted from the right atrium to the suprahepatic inferior vena cava, right hepatic vein and its branches with a delayed circulation time. There was no abnormality in the aorta and its great branches. Therefore, heart failure of the right side of the heart was suggested due to a large cardiac mass in the right cardiac chamber. A moderate amount of bilateral pleural effusion, pericardial effusion and ascites were associated with the disorder. There was no evidence of a distant metastasis in scanned lung and abdomen. To exclude the

possibility of large intracardiac thrombus, extremity doppler ultrasound was performed and it revealed no evidence of deep vein thrombosis in both lower extremities. On the following day, inferior vena cavography was performed that demonstrated no evidence of thrombosis in the inferior vena cava, both iliac veins and other lower leg veins. Thus, the mass in the right cardiac chamber was considered as a cardiac tumor rather than a thrombus. Transesophageal echocardiography revealed that the large echogenic lobulated mass was attached to the wall of right atrium and right ventricle, filling the right cardiac chamber with involvement of atrioventricular groove and extension to the pericardial space (Fig. 3).

The patient underwent open heart surgery to remove the mass. On the surgical field, a large well-encapsulated botryoidal mass was found in the right cardiac cham-

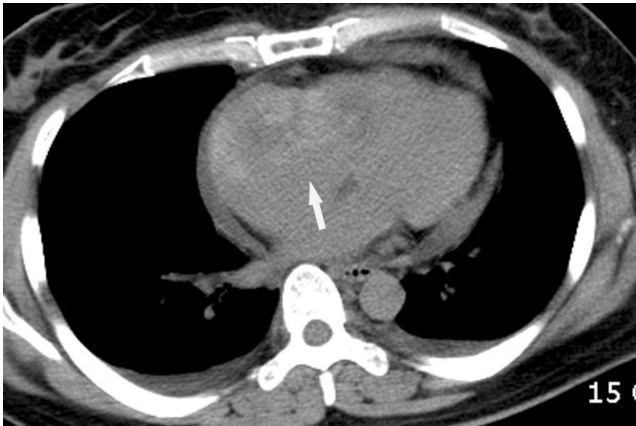


Fig. 1. A precontrast computed tomography scan reveals a large heterogeneously high attenuation mass filling the right atrium and ventricle of the heart (arrow).

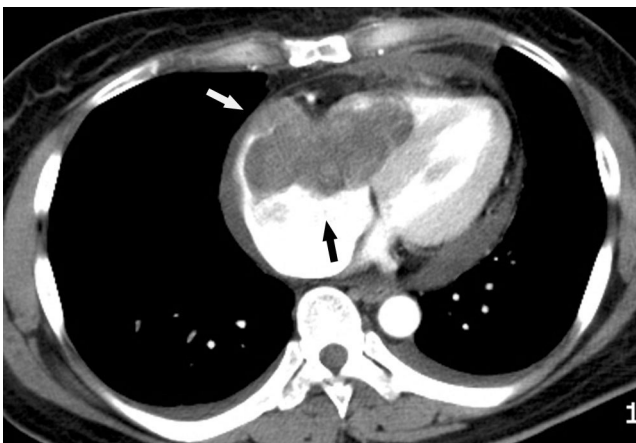


Fig. 2. A contrast enhanced CT scan shows the poorly enhanced lobulated mass (black arrow). Cardiac muscle invasion and pericardial extension of the tumor are suspected at the anterior wall of the right atrium (white arrow). Note the moderate amount of bilateral pleural effusion and pericardial effusion.

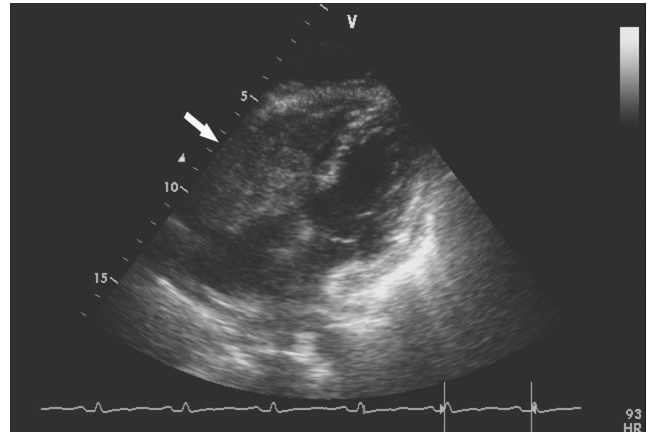
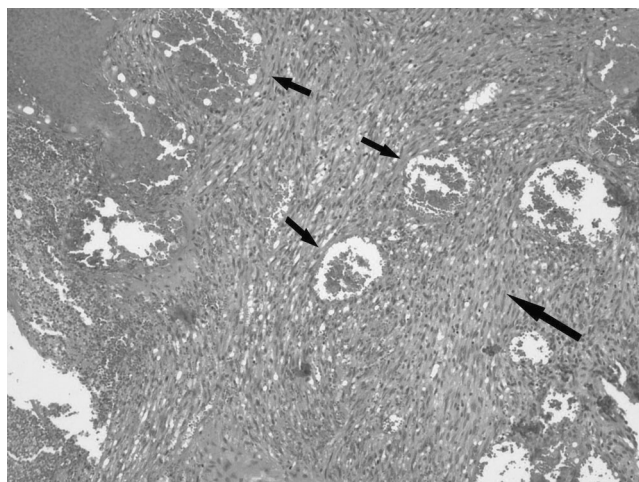


Fig. 3. A transesophageal echocardiograph shows the large echogenic lobulated mass attached to the right atrial and ventricular wall, filling the right cardiac chamber (arrow).



Fig. 4. The gross specimen shows a well-defined lobulated mass with internal hemorrhage measuring about  $7.5 \times 5.5$  cm. Note the botryoidal shape of the mass.



**Fig. 5.** The tumor consists of cavernous vascular spaces with RBCs (short arrow) and cellular areas of spindle cells (long arrow) (H & E staining,  $\times 100$ ). Note the central infarcted area of vascular spaces filled with RBCs.

ber (Fig. 4). It demonstrated broad attachment to the anterior wall of the right atrium with protrusion into the right ventricle through the tricuspid valve. A small thrombus was found at the adjacent right atrial muscle and inside of the mass. The mass extended into atrioventricular groove forming a mass at the origin site of the right coronary artery. The majority of the mass was excised but a small portion of the lesion remained in the right atrial wall. A follow-up transesophageal echocardiograph revealed about a 2.6 cm diameter sessile lesion that remained at the medial aspect of the atrioventricular groove.

The pathologic examination revealed that the mass was a vascular neoplasm with extensive central infarction, and the viable portion showed spindle cells with immunopositivity for CD 34 and factor 8. Multiple engorged round blood vessels were present in the infarction area (Fig. 5). The final pathologic diagnosis was a spindle cell hemangioma.

After the surgery, symptoms and physical activity of the patient gradually improved.

## Discussion

Spindle cell hemangioma is a distinct vascular lesion, characterized by cavernous blood vessels separated by spindle cells reminiscent of those seen in Kaposi's sarcoma. It has been described under the name of spindle cell hemangioendothelioma when first described in 1986 by Weiss and Enzinger (2). Before its original description, this lesion was probably misdiagnosed as a variety of be-

nign and malignant tumors, including angiosarcoma, Kaposi's sarcoma, cavernous hemangioma, and angiomatosis (3). Although Weiss and Enzinger indicated that spindle cell hemangioma is a vascular tumor of intermediate malignancy in their initial report, recently many investigators have argued against a malignant potential for spindle cell hemangioma (3, 5 - 9). Recently, Perkins and Weiss concluded that spindle cell hemangioma is a primary benign vascular neoplasm or malformation similar to angiomatosis. They suggested that the lesion be designated "spindle cell hemangioma" for a solitary lesion and "spindle cell hemangiomatosis" for multifocal lesions (3).

Most of the reported spindle cell hemangiomas involve the dermis and the subcutaneous tissue, and appear as a solitary or multiple nodules, usually involving the upper extremity of young adults. It follows a benign indolent course, but has a tendency for recurrence (4, 9). A radiologic report of intramuscular spindle cell hemangioma showed the presence of high-attenuation circular foci consistent with vascular spaces on an enhanced CT scan (7). MR imaging of the lesion demonstrated a heterogeneous signal intensity on both T1- and T2-weighted images.

Only one case of a spindle cell hemangioma that occurred within an internal organ (spleen) has been reported (3). An occurrence of a spindle cell hemangioma in the intracardiac space has never been previously reported. The neoplasm in this case arose in the intracardiac space, and was histologically characterized by a mixture of a large necrotic space of cavernous blood vessels and areas of spindle cells, which resembled Kaposi sarcoma. These histologic features are consistent with the vascular tumor termed "spindle cell hemangioma". Unlike the usual cardiac hemangioma, the mass was revealed as relatively well-defined lobulated lesion with poor enhancement on contrast enhanced CT. On precontrast CT, it showed heterogeneously high attenuation due to intratumoral hemorrhage and thrombus, which could initially be confused with another cardiac mass such as a myxoma or unusual large thrombus.

There were some limitations in evaluating the extent and characteristics of the mass in this case. Since CT would not reveal the exact tumor extension into the cardiac muscle and tumor composition, initially the mass was confused with another type of cardiac tumor other than a hemangioma component. MRI might be helpful in the preoperative evaluation of the composition of the mass (7).

In conclusion, cardiac spindle cell hemangioma is extremely rare and can manifest as an unusual form of a cardiac mass that can compromise cardiac function. It might be seen as a poorly enhanced intracardiac mass and may be confused with a thrombus or other tumor, including a myxoma.

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