

Primary Cardiac Angiofibroma

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Cardiac Angiofibroma is an uncommon intracardiac tumor. Thus far, only 4 cases of the rare intracardiac tumor have been reported. The present case-report describes an intracardiac angiofibroma in a 57-year-old healthy female. The patient was incidentally diagnosed with a left ventricle mass during echocardiography. We performed cardiac imaging, surgical excision and histological evaluation of the mass. The angiofibroma demonstrated features different from the relatively common cardiac tumors such as fibroma, myxoma and angiosarcoma. The cardiac MRI showed slightly high signal intensity on both T1 and T2, with the central core of lower signal intensity. The resected tumor was a whitish and rubbery mass. Histologically, the tumor showed the benign vascular proliferations associated with the surrounding collagen deposition. (Korean Circ J 2013;43:636-639)

KEY WORDS: Cardiac tumor; Magnetic resonance imaging.

Introduction

Primary cardiac tumors are rare across all age groups, with a reported prevalence of 0.001% to 0.03% in the autopsy series.¹⁾

Cardiac angiofibroma is a rare cardiac tumor, with only 4 cases having been reported worldwide. Among them, there has been no report that has discussed the tumor's characteristics on the cardiac MRI.

Thus, we report a case regarding an angiofibroma which has primarily originated from the left ventricle of the heart.

Case

A 57 year-old female without any medical history was admitted

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for a left ventricular (LV) tumor that was discovered incidentally. The tumor was found by echocardiography which was performed for the baseline evaluation of hypertension.

The echocardiography showed a round-shaped, immobile and echogenic mass attached at the LV apex. The valve and LV functions were normal (Fig. 1D).

Cardiac MRI was performed to determine the tumor's tissue type and its relations to other cardiac structures. It showed a slightly high signal intensity on both T1 and T2 weighted images. The gadolinium-enhanced cardiac MRI showed a hypoperfused tumor core and peripheral enhancement (Fig. 1).

In addition, the lesion showed the centripetal enhancement pattern during the first-pass infusion of the gadolinium-containing contrast (Fig. 2). The features of such are associated with the vascular tumors such as liver hemangioma.

The mass showed peripheral enhancement with the central sparing on the delayed enhanced imaging (Fig. 1C). It suggested that the tumor had an abundant fibrous content as well as a vascular content.²⁾³⁾

In this patient, the initial preoperative diagnosis of the mass was fibrous tumor of the LV apex. The patient was referred to a cardiac surgeon for the removal of the cardiac mass. Intraoperatively, a whitish, solid mass was detached from the base of the anterolateral papillary muscle (Fig. 3).

On the histologic evaluation, it showed a somewhat well-demarcated collagenous mass arising from the myocardium (Fig. 4A, arrows). The high-power view showed multiple irregular vascular ch-

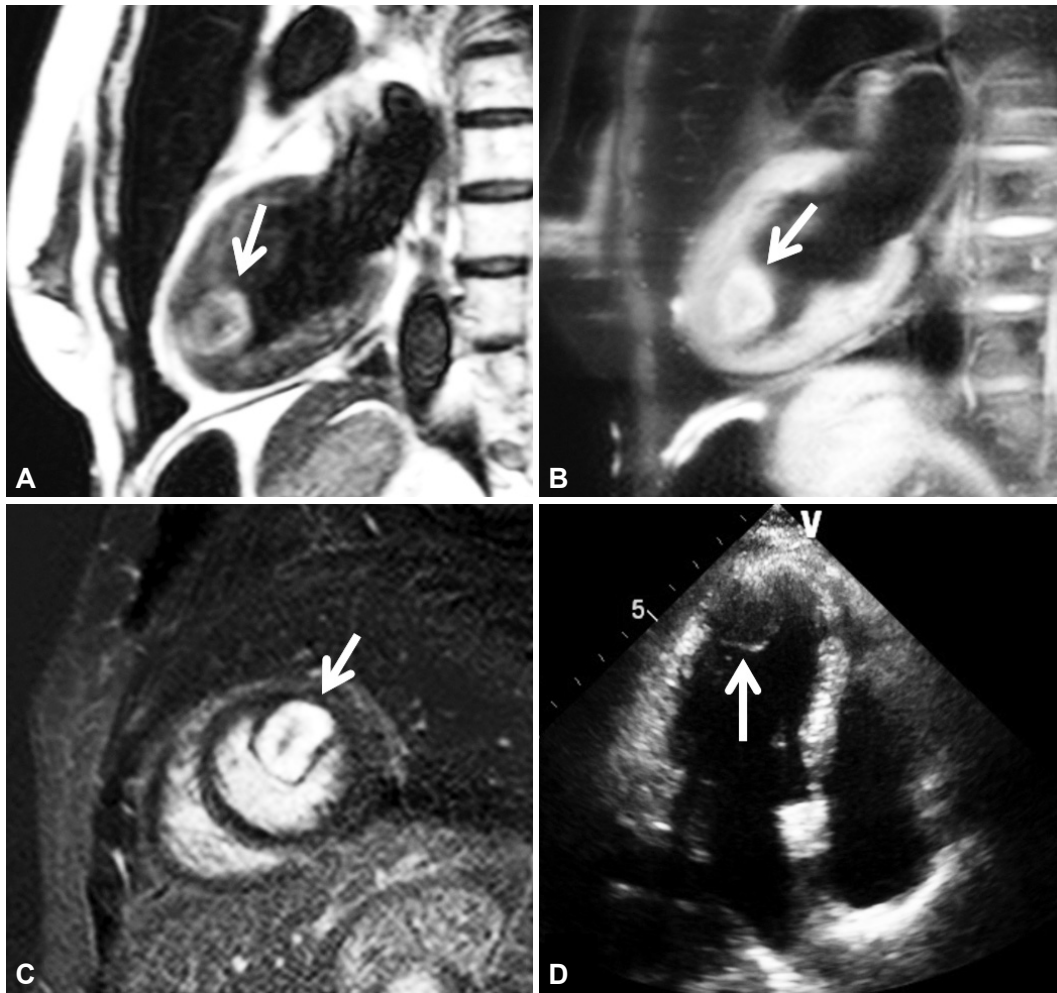


Fig. 1. A: T1 weighted image shows high signal intensity (arrow). B: T2 weighted image shows high signal intensity (arrow). C: delayed enhancement (phase-sensitive inversion recovery MR image) shows strong enhancement with central sparing (arrow). D: transthoracic echocardiographic images: four chamber view shown. It shows round shaped and echogenic mass attached at the left ventricular apex (arrow).

annels (Fig. 4B) with intervening dense collagen deposition in the mass. The desmin immunohistochemistry and trichrome staining confirmed that the stroma consisted of dense collagenous tissue, not smooth muscle (Fig. 4C and D). From these histologic findings, we diagnosed the mass as angiofibroma. The patient is currently doing well without any evidence of recurrence at the 2-year follow-up.

Discussion

Cardiac angiofibroma is a rare cardiac tumor, with only 4 cases having been reported worldwide. Two of the cases were diagnosed at childhood and related to the systemic congenital defects. In the first case, tuberous sclerosis was associated with the tumor which was found in the child.⁴⁾ In the second case, Beckwith-Wiedemann syndrome was associated with the tumor.⁵⁾

Cao et al.⁶⁾ reported a case regarding angiofibroma which was located in the right atrium and inferior vena cava. It was unclear wh-

ether it was a primary cardiac tumor or vascular tumor extending to the intracardiac cavity. The last case was reported from Russia in 1986.⁷⁾ None of these cases had the cardiac MRI evaluation; and all 4 cases could not be confirmed as a primary cardiac tumor.

Some tumors, like fibroma, hemangioma and rhabdomyoma, usually arise from the ventricle. Angiofibroma is distinguished from these ventricle-origin tumors by their histologic and image findings such as the cardiac MRI.

In this case, the cardiac angiofibroma showed the centripetal enhancement pattern during the first-pass infusion of the gadolinium-containing contrast. The features of such are associated with vascular tumors such as liver hemangioma. It suggested that the tumor had an abundant fibrous content as well as a vascular content.

However, fibromas have higher fibrous content and small vascular content. Therefore, the tumors often demonstrate little or no contrast-material enhancement. Similarly, rhabdomyoma is hypointense to myocardium after the contrast-material administration.

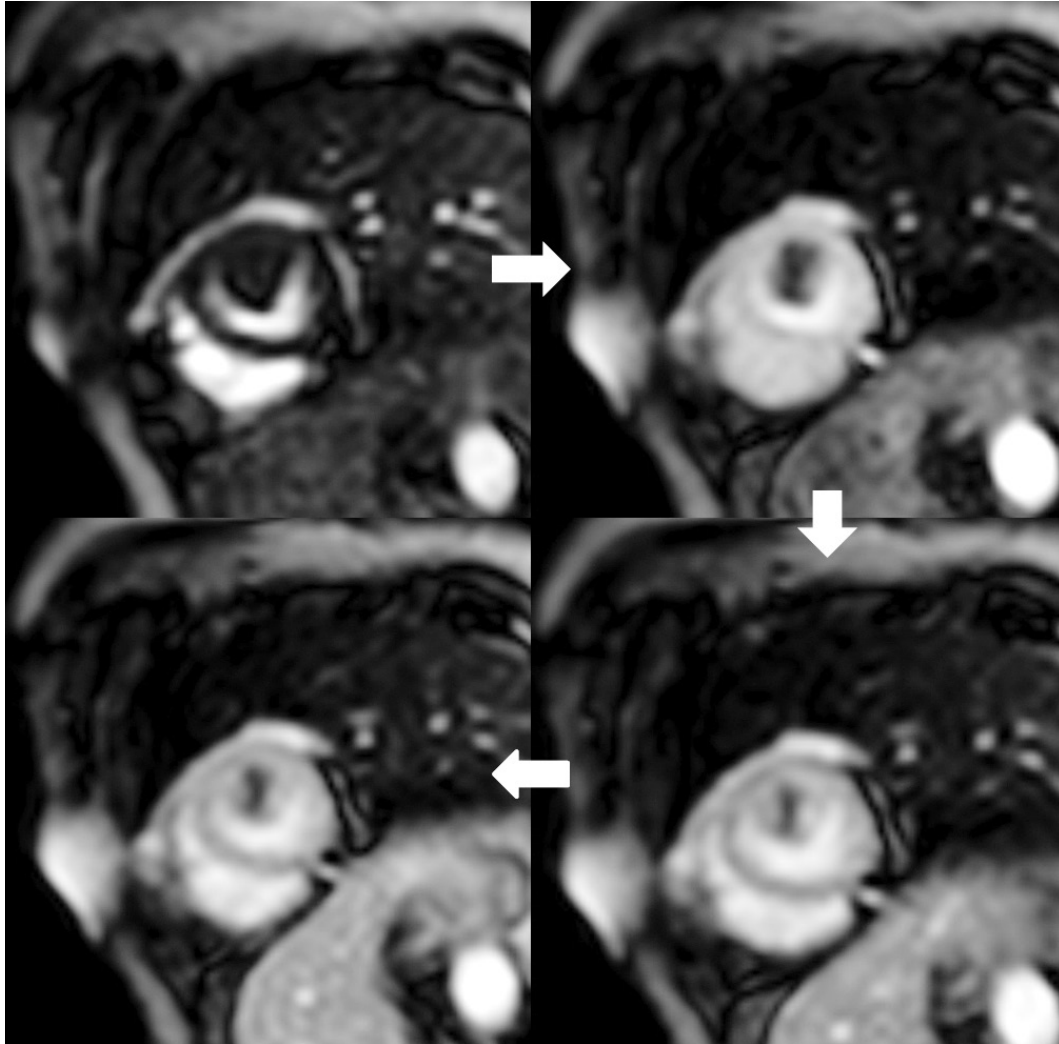


Fig. 2. Gd-DTPA first pass perfusion MRI in short axis view. The lesion shows centripetal enhancement pattern (clockwise from top left).

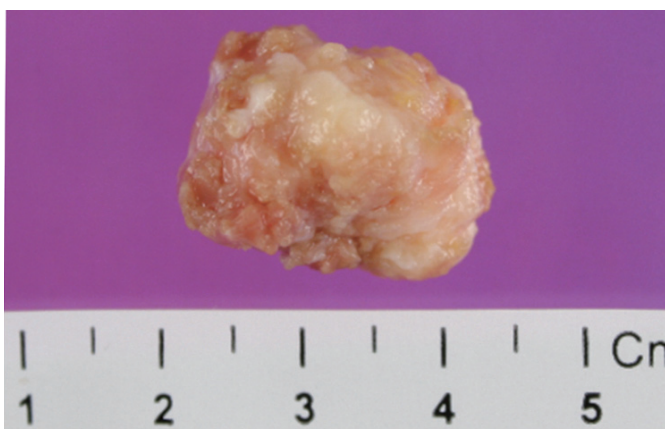


Fig. 3. On gross examination, a whitish, solid mass is detached from the base of the anterolateral papillary muscle.

Cardiac hemangioma is a benign cardiac neoplasm which has an abundant vascular component. It shows a more rapid enhancement than angiofibroma during the infusion of the contrast agent.

In immunohistochemistry, CD31 is used primarily to demonstrate the presence of endothelial cells in the histological tissue sections. Desmin is used to demonstrate the smooth muscle cells. In fibroma, the staining for CD 31 and desmin should be negative.

In this case, angiofibroma showed benign vascular proliferations associated with the surrounding collagen deposition. The immunohistochemical staining for CD31 and desmin were positive and negative, respectively.

In summary, we described the MRI and pathologic findings of a cardiac angiofibroma of the LV. These features were considered to be associated with the fibrous and vascular component of the tumor.

References

1. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumours: diagnosis and management. *Lancet Oncol* 2005;6:219-28.
2. Funari M, Fujita N, Peck WW, Higgins CB. Cardiac tumors: assessment with Gd-DTPA enhanced MR imaging. *J Comput Assist Tomogr* 1991;

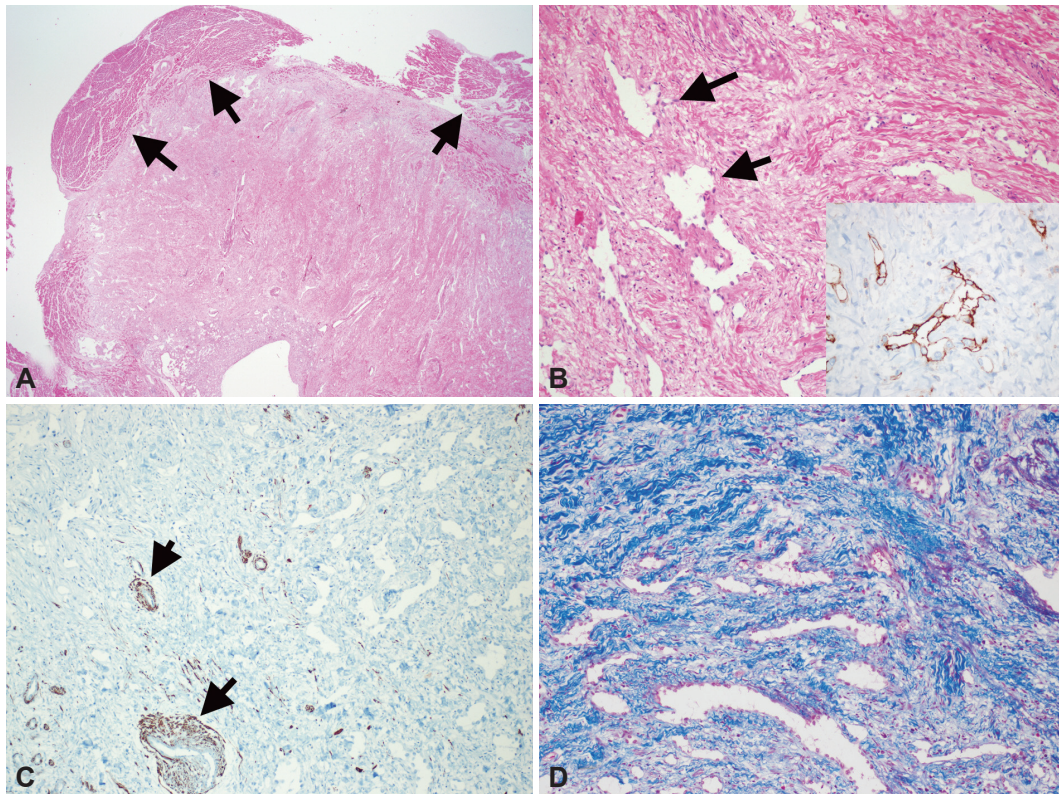


Fig. 4. A: on lower power view, the mass is somewhat well demarcated. And the mass seems to be originated from myocardium (arrows) (H-E, $\times 12$). B: high power view shows multiple irregular vascular spaces (arrows) with intervening wavy eosinophilic collagen deposition (H-E, $\times 100$). The vascular spaces express CD31 immunoreactivity (CD31 $\times 400$ inset). C: desmin immunohistochemical staining reveals that most of stroma do not express desmin except smooth muscles around vessel (arrows) (Desmin $\times 100$). D: most of the stroma shows bluish staining on trichrome (trichrome $\times 100$). Desmin and trichrome staining confirms that most of stroma consists of collagen, not smooth muscle.

15:953-8.

3. Brechtel K, Reddy GP, Higgins CB. Cardiac fibroma in an infant: magnetic resonance imaging characteristics. *J Cardiovasc Magn Reson* 1999;1:159-61.
4. Jutley RS, Janas R, Matuszewski M, Suvarna K, Locke TJ. Angiofibroma of the tricuspid valve: a rare presentation of the tuberous sclerosis complex? *J Cardiovasc Surg (Torino)* 2006;47:481-2.
5. Satgé D, Vidalo E, Desfarges F, de Geeter B. A third case of cardiac neo-

plasm in a fetus with Beckwith-Wiedemann syndrome: epicardial angiofibroma. *Fetal Diagn Ther* 2005;20:44-7.

6. Cao DB, Yang SR, Gao YS, Hua SC. Angiofibroma in the right atrium and inferior vena cava. *Eur J Cardiothorac Surg* 2012;41:962.
7. Kirillov MM, Arkhangel'skiĭ AV, Ivanovskii GI, Kuz'mina NIu. [Endocardial angiofibroma simulating a mitral heart defect]. *Klin Med (Mosk)* 1986; 64:120-3.