Primary Left Ventricular Angiomatosis

First Description of a Rare Vascular Tumor in the Left Heart

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SUMMARY

A 24 year-old male who presented with palpitations and presyncopal attacks had monomorphic ventricular tachycardia and a well-defined huge mass within the inferoposterior wall of the left ventricle proved by transthoracic echocardiography. The mass was completely resected and shown to be proliferative angiomatosis by histopathologic examination. This is the first reported case of primary left ventricular angiomatosis in the literature. (Int Heart J 2006; 47: 469-474)

Key words: Cardiac angiomatosis, Left ventricular tumor, Histopathological study

PRIMARY tumors of the heart are rare, with an incidence of 0.002% in reported or collected autopsy series.1,2) Approximately 75% of all cardiac tumors are histologically benign. Most benign cardiac tumors are myxomas, whereas almost all malignant tumors are sarcomas. Hemangiomas and lymphangiomas are major vascular cardiac tumors and classified according to the predominant type of proliferating vascular channel.3)

Angiomatosis is an extremely rare vascular tumor and is usually observed during childhood. We present a patient with primary cardiac angiomatosis arising within the left ventricular inferoposterior wall and invading the left ventricular myocardium, who presented with ventricular tachycardia and presyncopal attacks. The clinical, radiological, and pathological features of the case are discussed.

CASE REPORT

A 24-year-old white male presented with palpitations and dizziness which had progressed during the previous month. His cardiac examination was normal. A baseline ECG revealed symmetric negative T waves in inferior and lateral pre-
During an attack of palpitations, telemetry revealed monomorphic ventricular tachycardia at a rate of 280/min (Figure 1). Transthoracic echocardiography showed a huge (45 × 56 × 61 mm), nonhomogeneous mass within the inferoposterior wall of the left ventricle (Figure 2). The first-pass transit of injected GdDTPA on sagittal T2-weighted MRI image showed vascular nature of the mass (D). AO indicates aorta; LA, left atrium; LV, left ventricle; RV, right ventricle; and tm, tumor.

Figure 1. Baseline ECG (left panel) and ECG during palpitations (right panel).

Figure 2. Transthoracic echocardiography using parasternal long (A) and short axis (B) views demonstrated a huge mass within the inferoposterior wall of the left ventricle (arrows). On T1-weighted horizontal magnetic resonance imaging, an intramural nonhomogeneous mass was seen (C). The first-pass transit of injected GdDTPA on sagittal T2-weighted MRI image showed vascular nature of the mass (D). AO indicates aorta; LA, left atrium; LV, left ventricle; RV, right ventricle; and tm, tumor.
The mass was protruding into the left ventricle without obstructing the mitral inflow. The posteromedial papillary muscle was not involved. Magnetic resonance imaging confirmed the localization of the mass shown by the echocardiography. After IV administration of GdDTPA, the first-pass transit of contrast material showed the vascular nature of the mass (Figure 2C-D). Biochemical tests, hemogram results, tumor marker serology, and hydatid cyst were unremarkable. There were no other masses in the thorax, abdomen, or extremities.

Preoperative coronary angiography revealed that the tumor was fed mainly by the left circumflex coronary artery, and additionally to some extent by the right coronary artery (Figure 3). The patient was then referred to surgery. The capsulated, yellowish, rubbery tumor was completely resected under cardiopulmonary bypass using a left ventriculotomy approach after the benign nature of the tumor was proved by a frozen section histopathologic examination (Figure 4).

Figure 3. Coronary angiography (right anterior oblique projection) demonstrated that the mass was mainly fed by the left circumflex artery (left panel, arrows) and was protruding into the left ventricular cavity (right panel, arrows).

Figure 4. Macroscopic view of the excised yellowish, rubbery tumor during surgery.
Histologically, there were vessels of various sizes, adipose tissue, and striated muscle cells. Most of the vessels were small and thin-walled with flattened endothelium, however, some vessels were medium-sized and thick-walled. In some focal areas, there were small vessels arising adjacent to or within the wall of a larger vessel. There was a prominent amount of adipose tissue and striated muscle cells with large, hyperchromatic nucleoli and cross-striations (Figure 5). The diagnosis was proliferative angiomatosis, which is one of the rarest forms of vascular tumor. The patient was discharged without any complications and prescribed metoprolol 100 mg/bid to suppress the recurrence of arrhythmia.

**DISCUSSION**

Left ventricular tumors are rarely observed. They can appear as fixed intracavitary masses or mobile masses arising from the endocardium. Rhabdomyomas, fibromas, lipomas, and metastasis of malignant melanomas, as well as breast, bronchogenic, or esophageal cancers are the most common tumors of the left ventricle.4)

Angiomatosis is a rare, benign but clinically extensive vascular lesion and most cases are observed in childhood or adolescence. It can either be congenital or acquired. The congenital form of angiomatosis may be seen sporadically or accompanying certain types of syndromes such as Klippel-Trenaunay-Weber syndrome,5) Sneddon’s syndrome,6) or Gorham disease.7) In these syndromes, the lesions may be observed in any tissue and are clinically extensive, covering large parts of the body in a continuous pattern. However, in sporadic cases, the tumors are classically seen in extremities.
The acquired form of angiomatosis may be infectious or iatrogenic. Bartonella henselae is a recently recognized pathogenic bacterium associated with cat-scratch disease. In humans, bartonellosis can result in angioproliferative lesions that are potentially life threatening to the patient, such as angiomatosis. This type of angioproliferative lesion is seen especially in immunocompromised patients, transplantation recipients, or HIV patients. Iatrogenic cases occur after AV fistula formation or traumas. The treatment of choice in extensive angiomatosis is either radiotherapy or interferon α-2a treatment. In localized cases, complete resection is preferred, but there is a risk of local recurrence.

Two involvement types of angiomatosi are observed; extensive vertical involvement of multiple tissue planes or involvement of the same type of soft tissue. Such lesions usually present in the first 2 decades of life and have highly characteristic but not totally specific histologic patterns. The most common pattern consists of a haphazard proliferation of varying sized vessels, particularly large veins. However, the most distinctive feature is the clusters of capillary vessels residing within or adjacent to the vein walls. A second but uncommon pattern is the clusters of capillary-sized vessels infiltrating the soft tissues. Both types are typically associated with large amounts of adipose tissue and hence these lesions have led previous authors to use the term infiltrating angiolipoma, suggesting that these lesions are more generalized mesenchymal proliferations rather than exclusive vascular lesions.

Hemangioma and angiomylipoma should be kept in mind in the differential diagnosis of cardiac angiomatosis. Irregular venous channels with clustered small vessels in their walls are a characteristic of angiomatosis and this finding is not observed in hemangiomas. A basic component of an angiomylipoma is mature adipose tissue and thick-walled, medium-sized vascular channels surrounded by smooth muscle cells, which is not a basic component of angiomatosis. In our case, histopathologically there was striated muscle of the heart, infiltrated by the tumor. Although metastatic pericardial and cardiac angiomatoses were previously reported, to the best of our knowledge this is the first primary angiomatosis case of the left ventricle. After the operation, the patient was completely asymptomatic and is being closely followed-up for local recurrence.

REFERENCES