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Case Report

A Case Report of an Extensively Ossified Left Atrial Myxoma

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ABSTRACT

A cardiac myxoma, a rare but relatively common cardiac tumor, was surgically excised. Histopathological examination of the lesion revealed an extensive internal ossification. Ossification of the interior of myxomas is rarely reported, and most are large tumors; however, this is a rare case of a small tumor. The cause of ossification has not yet been clearly reported.

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Introduction

Cardiac tumors are relatively rare, accounting for 0.0017-0.33% of all autopsy cases. Approximately 70% of cardiac tumors are benign, and 30% are malignant [1]. The most common benign tumor is myxoma, which accounts for approximately half of all benign tumors and more than 30% of all cardiac tumors. Therefore, when considering cardiac tumors, we first consider myxomas. This is a tumor with an abundance of mucous-like substrates, which are reddish brown and jelly-like in appearance (Figure 1). It is two to three times more common in women than in men, is arbuscular, and can occur anywhere in the heart; however, three-fourths of cases occur in the left atrium. Approximately 5% of cases are familial, in which young males, multiple, and recurrent cases are common. Tumors vary in size from 1 to 15 cm but are usually detected at approximately 5 cm.

The main symptoms are impaired blood flow and embolism (thrombosis) associated with tumor occupation. In the case of myxomas, the tumor often occurs in the left atrium. As the tumor grows and begins to obstruct blood flow, it blocks the mitral valve, the door between the left atrium and left ventricle, causing symptoms similar to those of mitral stenosis (soporific valve stenosis). The symptoms include syncope, dizziness, and shortness of breath. In the upright position, the myxoma is pulled toward the mitral valve by gravity, obstructing blood flow through the mitral valve and causing symptoms. In some cases, the tumor remains stuck in the mitral valve opening, blocking blood flow, and causing sudden death. Apart from symptoms associated with impaired blood flow, 30–50% of myxomas break off, and a portion of the tumor and fly into the bloodstream, causing an embolism. In approximately half of these cases, an embolism of the central nervous system occurs. The possibility of embolism also exists in tumors other than myxomas.

Histologically, it is mainly composed of PAS stain-positive myxoma cells, with some fibrosis and calcification. Its origin is unclear, but it is thought to be due to remnant mesenchymal cells from the embryonic period. Benign tumors other than myxomas include lipomas, papillary elastic fibromas, rhabdomyomas, fibromas, hemangiomas, atrioventricular nodular mesotheliomas, and teratomas. Each has its own unique morphological characteristics. Papillary elastoma, for example, has a chorionic structure that resembles the sea anemone in the body. However, depending on the location of the tumor, it may affect the cardiac function or break off a part of the tumor, which may cause embolism. Therefore, in principle, the tumor should be removed.

Malignant tumors in the heart can be primary or metastatic. Most primary malignant tumors are mesotheliomas, sarcomas, and lymphomas. All the patients had a poor prognosis. Malignant mesothelioma often develops in young adults and leads to death within a year. Sarcomas and malignant lymphomas are most common in the

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right atrium and develop in middle age or later, with a high rate of metastasis to the lungs and mediastinum. Treatment consists of surgical resection, radiation therapy, and chemotherapy with anticancer drugs; however, none of these are very effective, and there are few reports of therapies that have improved prognosis.

Metastatic cardiac tumors were also observed. This refers to the metastasis of malignant tumors to the heart, primarily to organs other than the heart. Of all malignant tumors, 10–20% are said to metastasize to the heart. Primary tumors include lung cancer, breast cancer, malignant lymphoma, and leukemia. Looking at the rate of cardiac and pericardial metastasis of primary malignancies, the question is, for example, what percentage of lung cancer metastasizes to the heart, while leukemia and malignant melanoma account for 40–50%, thyroid cancer, lung cancer, and sarcoma for 30%; and breast cancer, malignant lymphoma, esophageal cancer, and kidney cancer for 20%.

Case Presentation

The patient was in 80s. He had a history of atrial fibrillation and aortic regurgitation and was under observation at an internal medicine clinic. During follow-up, an echocardiogram revealed a tumor attached to the left atrial septum. Cardiac catheterization revealed significant stenosis in the anterior descending branch of the coronary artery, and the patient underwent tumor resection and coronary artery bypass surgery. He had no family history of cardiac tumors. No physical examination findings were obtained. No significant inflammatory findings or elevated tumor markers were observed in biochemical blood tests.

Preoperative cardiac MRI revealed a well-defined mass lesion, 30 × 27 mm in diameter, in the left atrium, bordering the atrial septum. T1WI and T2WI images showed iso- to high-signal areas, while T2WI and T2WI images showed uneven strong high-signal areas. The atrial septum was widely contiguous with the atrial septum, was not mobile, and had no evidence of a stalk. No fatty components or calcifications were observed. Preoperative echocardiography revealed a tumor attached to the left atrial septal wall. The tumor was approximately 3 × 2 cm in size with no stalk. It was not located in a position that would interfere with mitral valve motion, and no effect on valve motion was noted (Figure 1).

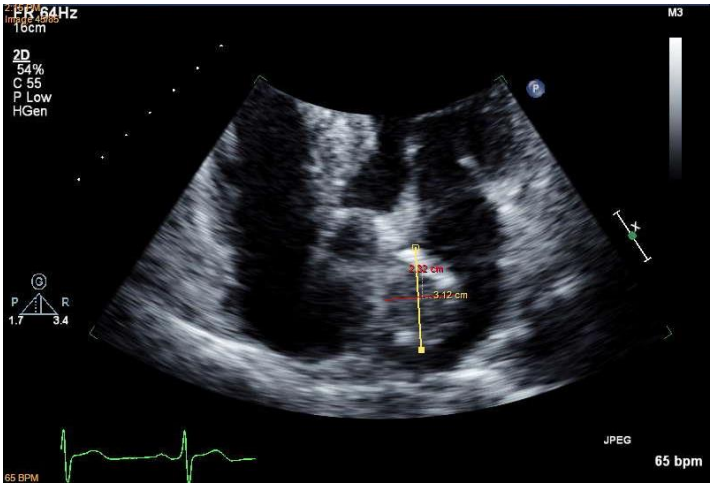

Figure 1: Preoperative echocardiography. The tumor was approximately 3 × 2 cm in size with no stalk.

During surgery, an incision was made on the right side of the left atrium with a margin of approximately 5–8 mm from the tumor, and the entire atrial septum was removed. The tumor was smooth, hard, and dense. It extensively adhered to the atrial septum. Gross examination of the resected specimen revealed a tumor measuring 3.5 × 2.7 × 2.3 cm in diameter with a stalk at the base of the tumor (Figure 2).

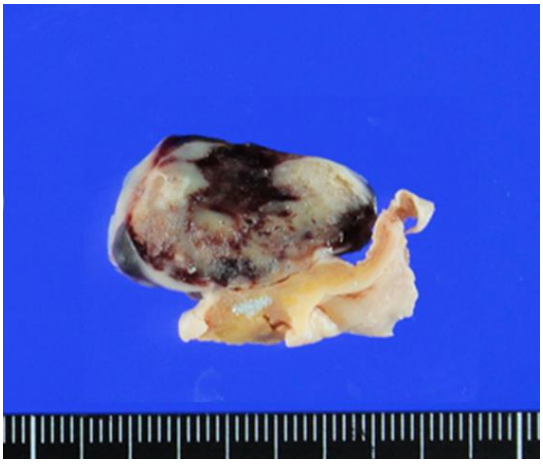

Figure 2: The surface was a mixture of smooth and non-smooth areas. The split surface was a mosaic of yellowish brown and dark red.

The surface was a mixture of smooth and non-smooth areas. The split surface was a mosaic of yellowish brown and dark red. Histologically, fibrous tissue with myxoid changes was observed. Ossification was observed extensively (approximately 35%) in the central part of the lesion, and some of the bone tissue resembling layered mature bone was observed. Fatty marrow was also present (Figures 3 & 4). The immunostaining results are shown in (Table 1). Vascular endothelial markers and calretinin were positive, whereas the other markers were negative.

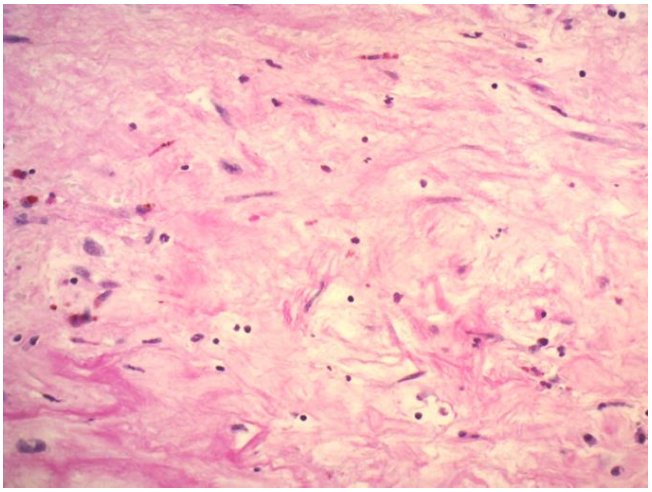

Figure 3: Fibrous tissue showing myxoid change (H.E. stain).

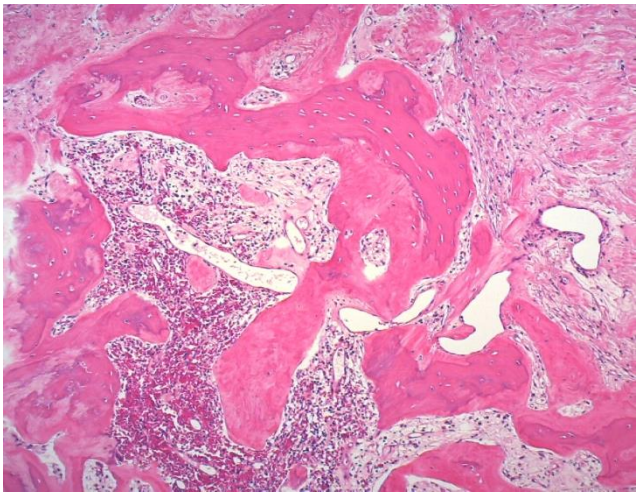

Figure 4: Ossification was observed extensively.

Table 1: Results of immunostaining.

Antibody	Results
CD34	Positive
CD31	Positive
Factor VIII	Positive
Calretinin	Positive
β-catenin	Negative
Ki-67	Less than 1%
CD117	Negative
S100 protein	Negative
D2-40	Negative

Vascular endothelial markers and calretinin were positive.

Differential diagnoses included thrombus, intimal fibroplasia, myxoid sarcoma, calcified cardiac amorphous tumor, mesothelioma, fibroma, and hemangioma. However, the presence of myxoid cells positive for calretinin, the absence of pleomorphic spindle cells, no conspicuous calcified lesions, no conspicuous collagenous stroma, no vascular lobular, the absence of pleomorphic spindle cells, no conspicuous calcified lesions, no conspicuous collagenous stroma, and no vascular lobular changes was observed. Based on these findings, a diagnosis of cardiac myxoma with extensive ossification was made.

Discussion

Cardiac myxomas are the most common primary tumors of the heart; however, bone formation and myelopoiesis are extremely rare. Although the origin of myxoma has not yet been established, myxoma cells have been reported to take a variety of forms, including smooth muscle myoblasts, fibroblasts, endothelial cells, and chondroblast [2]. Myxomas are thought to appear during the process of differentiation into various tissues, and the theory of embryonic cell remnants is considered to be the most likely explanation. The atrial septum, especially in the fossa ovalis, is subjected to blood flow stress during the embryonic period, and it is believed that mesenchymal cells with pluripotency capacity may

remain. Therefore, the presence of bone and bone marrow of mesenchymal origin in myxomas is an embryologically understandable phenomenon.

This case is considered rare, as there have been only four reports of cardiac myxomas with ossification [3-6]. The size of the myxoma is relatively small compared with previously reported ones, which were 120 mm × 100 mm, 77 mm × 55 mm, 70 mm × 30 mm, and 43 mm × 30 mm, respectively. Considering the origin of cardiac myxomas, it is understandable that even small myxomas can ossify. However, because many cases do not show ossification, it is possible that some trigger or remnant cells may cause ossification. The origin of the disease should be clarified in the future when more cases are reported and molecular biological studies are conducted.

Conclusion

Herein, we report a case of cardiac myxoma with extensive ossification. The exact mechanism of bone formation in myxomas is unclear, and further histological and molecular biological studies and case series are needed.

Conflicts of Interest

None.

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