

A CASE REPORT: A RARE FORM OF RIGHT VENTRICULAR FIBROMA

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Primary tumors of the heart are rare. As one of them, fibromas are benign connective tissue tumors which usually localize in the ventricles. Macroscopically, fibromas are typically solid tumors. Fibromas are not encapsulated and they extend into the surrounding myocardium. Also, neovascularisation of the tumor is rarely observed.

We present a case with an encapsulated cardiac fibroma adhering to the chorda tendinea of the tricuspid septal leaflet with a single pedicle and showing cystic degeneration and neovascularization, which is uncommonly seen in cardiac fibromas.

Key Words: Cardiac tumor, fibroma, right ventricle, neovascularization

Tumors of the heart, while uncommon, present in protean ways and have challenged the physicians since, 1774. Primary tumors of the heart are rare with a frequency of 0,001 to 0,28 percent in reported postmortem series. Most of the primary tumors of the heart manifest themselves as intracavitary lesions(1). Cardiac tumors may present themselves primarily with rhythm disorders, contractility abnormalities, obstructions, valvular regurgitations, embolic events, or pericardial effusion with cardiac tamponade. Because of the crucial nature of the various cardiac structures involved, the consequences can be fatal. The complications related to the tumors depend on their location, size, mobility, friability, and invasiveness. Fibromas are benign connective tissue tumors the majority of which are encountered before the age of 10. Males and females are equally affected (2). The frequency of cardiac fibromas which are usually diagnosed in infants and children is reported to be 4 percent in different postmortem histopathological series (3). Sudden death occurring in nearly one-third of the patients is thought to be likely due to the involvement of the conduction system, inducing arrhythmias, or the obstruction of the left ventricular outflow tract (4).

Almost all cardiac fibromas occur within the ventricular myocardium, most frequently within the anterior free wall of the left ventricle or the interventricular septum and much less often in the posterior ventricular wall or right ventricle. Macroscopically, fibromas are typically solid tumors. Fibromas are not encapsulated and they extend into the surrounding myocardium(5). Microscopically, cardiac fibromas consist of elongated fibroblasts admixed with fibrous tissue being composed mostly of collagen. This cellularity of the tumor is variable and mitotic figures are seen rarely. Calcification and bone formation may be seen and neovascularisation of the tumor is rarely observed.

We present a case with an encapsulated cardiac fibroma adhering to the chorda tendinea of tricuspid septal leaflet with a single pedicle and showing cystic degeneration and neovascularization, which is uncommonly seen in cardiac fibromas.

CASE

A 20 year-old-male patient was referred to the outpatient clinic due to an auscultation of grade II systolic murmur in his left lower sternal area. He was entirely asymptomatic. There were no cardiac or any other organic diseases in his medical history. In the patient's family history, his father was hypertensive and has used anti-hypertensive drugs for 5 years regularly. His mother and two brothers had no cardiac disease.

In physical examination, vital signs were normal. His arterial blood pressure and heart rate values were 110/70 mmHg, 76 beat per minute, consecutively. An early, soft, low pitched systolic murmur was heard in the left lower sternal area. The other system's examinations were normal. His resting surface electrocardiography and chest X-Ray revealed no abnormality.

Two dimensional transthoracic echocardiography showed a mobile, threelobulated, pedicled hyperechogenic mass lesion located beneath the tricuspid septal leaflet in the right ventricular cavity (Figure 1). Additionally, a mild tricuspid regurgitation was found.



Fig 1: Apical Four Chamber Image Of The Tumor by Transthoracic Echocardiography.

Transesophageal echocardiography (TEE) helped us to delineate the accurate location of the tumor in the right ventricle. Under the septal leaflet of the tricuspid valve, a mobile trilobulated, pedicled, hyperechogenic mass, which did not obstruct the right ventricular outflow tract, along with a mild tricuspid regurgitation were seen in TEE (Figure 2).



Fig 2: The Visualization Of The Tumor In The 82 0 Transesophageal Image Plane.

Cardiac spiral computed tomography (CT) revealed a suspicious smooth walled filling defect with a diameter of 3 cm in the right ventricle cavity (Figure 3). Cardiac magnetic resonance imaging (MRI) informed us about the tumor's anatomic boundaries and tissue characterization.



Fig 3: Filling Defect In Right Ventricle Cavity In Contrast Enhanced Spiral CT.

A right ventricular mass lesion with a diameter of 2,5x2 cm and becoming isointense with the myocardium after contrast enhancement was found in cardiac MRI (Figure 4).



Fig 4: Postcontrast Short Axis Axial Plane Images Of The Tumor In Cardiac MRI.

It was decided to excise the tumor after taken written informed consent from the patient. A cardiologist was ready in the operating room. The tumor was located in the right ventricle cavity area and didn't obstruct any crucial cardiac structures involving the right ventricular outflow tract. The encapsulated tumor resem-

bled clusters of grapes in colour of purple (Figure 5). The tumor adjoined to the underside of the septal leaflet of the tricuspid valve with a single pedicle.



Fig 5: Intraoperative View Of The Tumor At The Operating Room.

In histopathologic examination, it was reported that this lesion was a right ventricular cardiac fibroma which showed neovascularization and complete cystic degeneration with calcification. The patient's hospital course was uneventful after excision of the tumor and was discharged without complication after ten days followup.

DISCUSSION

Primary tumors of the heart are far less common than metastatic tumors to the heart (6). Benign primary cardiac tumors occur more frequently than malignant ones. The most commonly seen cardiac tumor is reported to be the myxoma(1). During the past decade, major advances in noninvasive cardiovascular diagnostic techniques, especially echocardiography(7), CT(8) and MRI(9) have permitted the diagnosis of these rarely seen tumors in clinical cardiologic practice.

Fibromas are benign connective tissue tumors which usually localize in the ventricles(7).

Although fibromas are mostly detected as incidental findings during the postmortem examination, approximately 70 percent of the diagnosed ones can cause mechanical interference with intracardiac flow, ventricular contraction abnormalities, or conduction disturbances(6). Clinical manifestations are protean and vary from cardiac murmurs, atypical chest pain to congestive heart failure, signs of subaortic stenosis, valvular or infundibular pulmonic stenosis with right ventricular hypertrophy, tricuspid stenosis, conduction disturbances, ventricular tachycardia, and sudden death. Our patient was referred to the cardiology department due to an auscultation of a soft low pitched early murmur at the lower left sternal area with no symptoms. Importantly, not only did the tumor not obstruct the right ventricular outflow tract, but also didn't invade the conduction system. Additionally, there were no signs of pulmonary embolism.

Two dimensional echocardiography is one of the mostly used modalities to diagnose the cardiac tumors. In this case, a transthoracic and followed by a transesophageal echocardiography showed the location, size and mobility of the tumor moderately. CT and MRI added detailed information about the tumor's accurate location, size, tissue characterization and the relation to the other crucial cardiac structures. Macroscopically, the tumor, which is localized under the septal leaflet of the tricuspid valve as a freely moving mass lesion, look liked a cluster of grapes in purple color. More importantly, the tumor was encapsulated and not solid which is not usually encountered in this type of tumors.

In the histopathology, extensive neovascularization and complete cystic degeneration, which is rarely seen in cardiac fibromas, were seen.

In conclusion, we presented a rare form of right ventricular cardiac fibroma localized under the septal leaflet of tricuspid valve and showed extensive cystic degeneration and neovascularization. So, the cardiac fibromas should be remembered in the diagnosis of the masses showing the features like those of our

patient's lesion.

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