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A Large Cardiac Fibroma Lied in the Interventricular Septum of an Adult with Exertional Pain and Palpitation: A Case Report

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ABSTRACT

Introduction: Cardiac fibromas are rare primary tumors that mainly affect children and teenagers and are rarely found in adults. The patients may be asymptomatic or may present their disease with arrhythmia or even heart failure. However, bizarre presentations can be found. The present study aimed to report a case of cardiac fibroma presented with exertional dyspnea and palpitation.

Case Presentation: A 29-year-old Caucasian male presented with the symptoms of exertional dyspnea and palpitation in the emergency room. Tachycardia was evident in physical examination and electrocardiography. On the echocardiographic assessment, a hyperechogenic mass was found in the proximal part of the anterior interventricular septum with calcified spots and without defined capsules around the mass suggestive of lipoma or other infiltrative mesenchymal tumors. Cardiac magnetic resonance assessment with fat suppression mode ruled out lipoma and bolded fibroma diagnosis. The patient underwent excisional surgery and survived the condition. On pathology and immunohistochemistry evaluation, the diagnosis of fibroma was confirmed. The patient survived the surgery with no morbidities. Hemodynamic study revealed no findings suggestive of heart failure.

Conclusions: Cardiac fibroma is a rare tumor in adults and usually happens in children under five years old. This condition can be diagnosed in echocardiography and the subsequent cardiac magnetic resonance imaging. The tumor is usually featured with central calcification.

1. Introduction

Cardiac fibroma is a benign fibroblastic tumor with unknown etiology, comprising 12 - 16% of cardiac tumors. It has been mentioned as the second most prevalent benign heart tumor after rhabdomyoma (1). The tumor commonly occurs in children under five years old; however, there are reports of its occurrence in adults (1, 2). This rare condition occurs in 0.0017 - 0.019% of the common population (3). Some of the cases are a part of the Gorlin syndrome or the basal cell nevus syndrome (4). One-third of the patients are found accidentally without any symptoms (5). However, some others may present with arrhythmias and heart failure. Other symptoms include syncope, chest pain, dyspnea, cyanosis, and even sudden cardiac death (6, 7). Electrocardiography (ECG) may show Atrioventricular (AV) block or ventricular arrhythmias (5, 8). Yet, the hallmark of the diagnosis is the central calcification of a relatively avascular tumor in the Computed Tomography (CT) scan or Magnetic Resonance Imaging (MRI) (9). MRI may also show a heterogeneous mass with lower central intensity (10). The tumor is immobile in echocardiography (9).

Patients can be treated by the total resection of the tumor and recurrence is uncommon. If the tumor remains untreated, no regression or progression will occur in the mass (11). The current study aims to report a case of cardiac fibroma in a young man presenting with palpitation and chest pain.

2. Case Presentation

A 29-year-old man with the weight of 158.7 lbs and the height of 72 inches presented with recurrent episodes of

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palpitation and exertional pain within the last two months. The physical examination of the patient showed no notable points, except for tachycardia, with a heart rate of 118 beats per minute. The patient also had no past medical or surgical history. On the emergency admission, the patient had a normal electrocardiogram and chest X-ray. Thus, the patient underwent echocardiography. The report of echocardiography revealed a 5 × 4.7 cm localized homogenous and hyperechogenic mass in the proximal part of the anterior interventricular septum with calcified spots and without defined capsules around the mass suggestive of lipoma or other infiltrative mesenchymal tumors. The echocardiography report also showed reduced septal strain, mild tricuspid regurgitation, and mild pulmonary regurgitation. Other echocardiographic measurements were normal. The measured ejection fraction was 60%.

Further assessment was done using Fast Imaging Employing Steady-state Acquisition (FIESTA) Cardiac Magnetic Resonance (CMR) imaging. The 4-chamber, 2-chamber, coronal, Right Ventricular Outflow Tract (RVOT), and short views as well as tagging sequences showed a considerably large mass occupying a considerable proportion of the left ventricle and even the right ventricular outflow. However, no change in the length of cuboids, no systolic thickening based on the radius of the RCL curvature, no C and L shortening, no twisting and torsion of the mass, no difference in the radius of L and C curvature, and no segmental deformation were found, and the mass was detected as an immobile tumor (Figure 1A). Moreover, pre-contrast T1 and T2 DIR FSE revealed heterogeneous, iso-intense, and intra-mass hypo-intense foci. However, fat suppression showed no evidence of fat in the mass and, consequently, lipoma was ruled out. Furthermore, T1 DIR FSE fat suppression imaging ten minutes after gadolinium injection presented a heterogeneously enhanced mass composed of central areas with lower intensity and a peripheral hyper-intense rim that corresponded to a previous intra-mass necrosis secondary to a poorly vascularized fibrous region (Figure 1B). Myocardial Delayed Enhancement (MDE) 15 minutes after gadolinium injection showed a heterogeneously enhanced mass with a lower intensity central area (Figure 1C). Myocardial Perfusion Imaging (MPI) first pass study showed no first pass filling and vascular lesion (Figure 1D). Moreover, hemodynamic study showed that the end diastolic volume indices were 103.7 and 72.9 for the left and the right ventricle, respectively. The end systolic volume indices were also 53.2 and 31.3 for the left and the right ventricle, respectively. The left and right ventricles' ejection fractions were 49% and 57.1%, respectively (Figure 2).

The patient underwent surgery and survived the total excision of the tumor with no morbidities. The excised tumor was a solitary huge well-circumcised intra-cardiac soft tissue mass with sharply defined borders and a size of 7.4 × 6.2 cm, a volume of 140.7 cc, and a weight of 115.2 g, occupying 63% of the left ventricle. Pathological assessment reported the hypocellular proliferation of the band slender

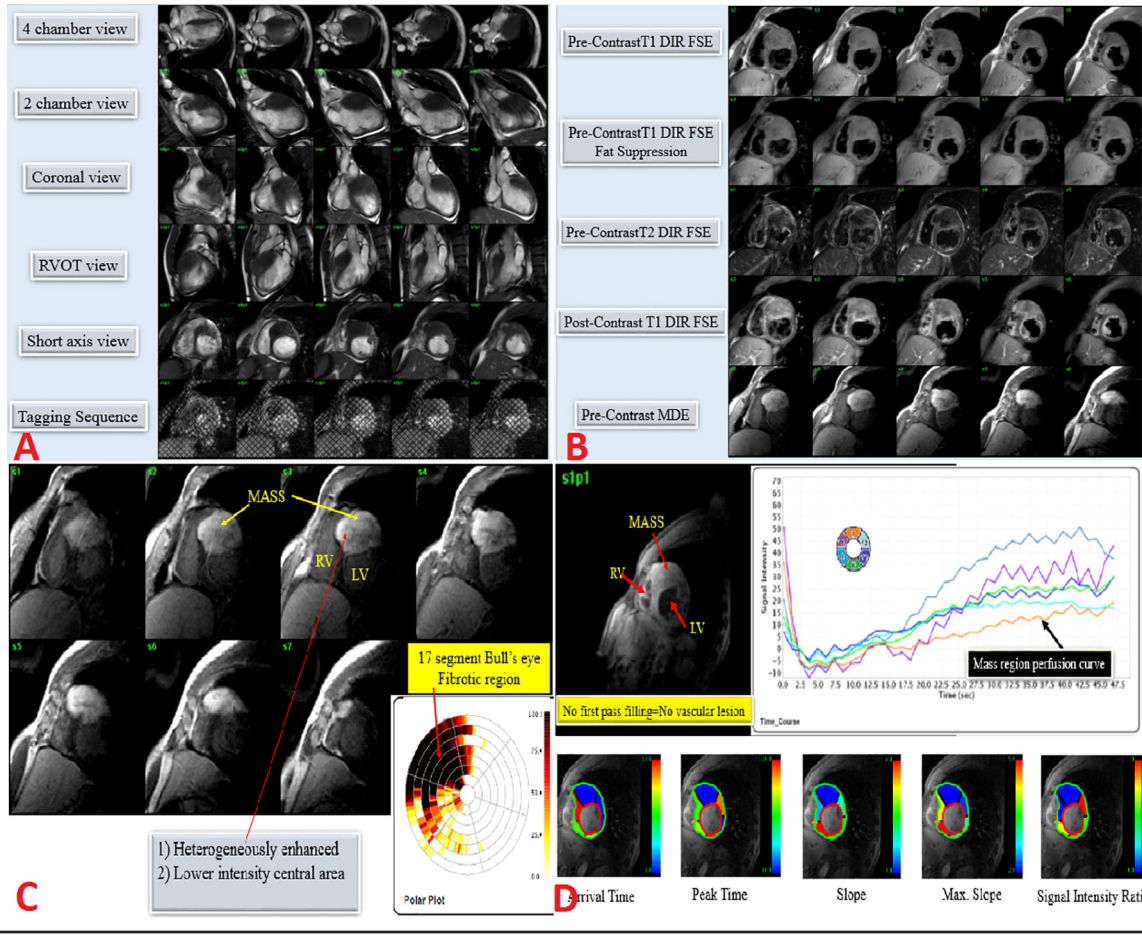

Figure 1 (A). FIESTA Different Views Showing a Hypointense Mass in the Interventricular Septum; **(B).** Different T1 and T2 MRI Views with or without Contrast Injection; **(C).** Myocardial Delayed Enhancement 15 Min after Gadolinium Injection; **(D).** Myocardial Perfusion Imaging, First Pass Study

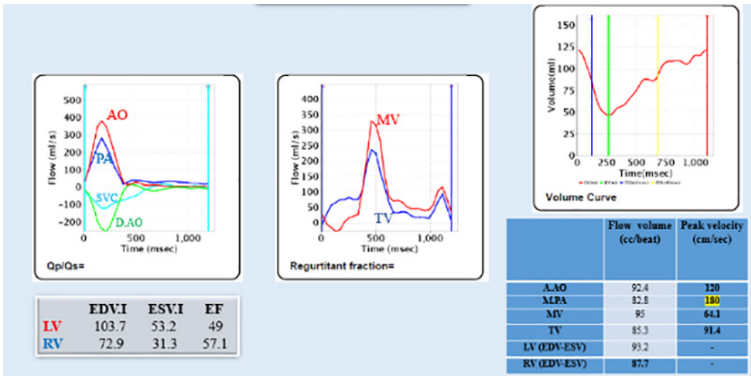

Figure 2. The Hemodynamic Study Data

spindle cells arranging in interlacing fascicles embedding in a densely collagenized stroma extending superficially to the myocardium, which was indicative of cardiac fibroma. Besides, the immunohistochemistry study showed that the tumor was negative for Ki67, B-CATENIN, SMA, Desmin, S100, Calretinin, and CK markers, which further confirmed the diagnosis of fibroma. In order to publish the records, the patient's written informed consent was obtained.

3. Discussion

Cardiac tumors are classified into primary and secondary categories. According to autopsy studies, primary tumors are rare with a frequency of 0.001 - 0.030%, affecting three out of every four patients (12). Cardiac fibromas are very rare primary tumors in adults. Around 170 cases of cardiac fibroma in different heart chambers were found during the past 50 years, most of which sized around 5 cm (13). This tumor usually presents itself in teenagers and around 33% of the patients are younger than one year. However, adult cases have been found, as well (14). The most commonly affected chambers have been reported to be the left ventricle, right ventricle, and interatrial septum. Around three out of four fibromas occur in the left ventricle and one-third in the right ventricle, while other parts of the heart may also be affected. The highest rate of mortality (60%) belongs to the interseptal tumors (15). Furthermore, younger patients have been found to have poorer prognosis (16).

In the current study, the case presented in an uncommon age with uncommon presentations. He had palpitation and exertional chest pain. Generally, patients with fibroma can present with arrhythmia or heart failure. However, some may have a vague pain in the chest (17). The most common symptom of fibroma is progressive heart failure due to the obstruction of the inflow or outflow of the heart. However, the presentation of the tumor depends on its location. In minority of the cases, tumor embolism may be seen (17). In the present case, the site of the tumor could be also listed as rare as it was found in the inter-ventricular septum. However, the mass occupied a considerable portion of the left ventricle. A recently published systematic review reported that 57.3% of the fibromas were located in the left ventricle, 27.5% in the right ventricle, 17% in the inter-ventricular septum, 5.3% in the right atrium, and 1.8% in the left atrium (14).

The first diagnostic procedure in these patients is echocardiography. This modality shows a large, solid, hyperechogenic, and immobile mass in the heart (9). Further validation of the diagnosis should be done by CMR imaging, which shows central calcification and fat suppression can rule out lipoma as a differential diagnosis of fibroma (10). This procedure was followed in the present investigation, revealing a mass suggestive of fibroma. However, the definite diagnosis had to be made via biopsy as the tumor was excised and studied by an expert pathologist who reported a mere diagnosis of fibroma. The case survived after the total excision of the tumor. However, there were similar cases in other studies who could not survive the condition or developed heart failure (18, 19).

3.1. Conclusion

In conclusion, cardiac fibroma is a rare condition, especially in adults. The diagnosis can be made with non-invasive modalities, such as echocardiography and CMR imaging. The hallmark of the diagnosis is central calcification. Patients can have desirable outcomes after the surgical removal of the tumor. However, some may have heart failure at the time of diagnosis, which requires heart transplantation.

3.2. Ethical Approval

Not applicable.

3.3. Informed Consent

Written informed consent form was obtained.

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Authors' Contribution

Study concept and design: A.F. and A.D.; Analysis and interpretation of data: A.F. and A.K.; Drafting of the manuscript: A. G.; Critical revision of the manuscript for important intellectual content: A.F., A.G., and A.O.

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