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Large intra-cardiac fibroma identified on cardiac MRI — a case report and review of literature

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Abstract

Primary cardiac tumours are rare. Cardiac fibroma is a benign tumour of the heart. It is fairly common among children and adolescents and is rarely encountered in adults. We present the case of a thirty-eight year old lady who presented with shortness of breath and was found to have a very large intra-cardiac mass that had cardiac magnetic resonance (CMR) features consistent with cardiac fibroma. The patient was referred for tumour resection, however could not survive the surgery.

Keywords: Fibroma, Cardiac MRI, Cardiac tumours, Cardiac fibroma, CMR.

Introduction

Cardiac fibromas are rare benign tumours of fibroblasts. They are the second most common primary cardiac tumours among children; however they are rarely seen in adults.¹ Their clinical presentation depends upon their location and size. Rarely they may be asymptomatic and found on routine imaging. Treatment is with complete or partial resection of the tumour. We present here the case report of a patient who was found to have a cardiac fibroma on Cardiac Magnetic Resonance Imaging (CMR). To the best of our knowledge this is the first case report from Pakistan that describes the CMR features of a cardiac fibroma.

Case Report

A thirty-eight year old lady with no known comorbid conditions presented with three months history of progressively worsening shortness of breath on exertion. She had an echocardiogram done at a local hospital that revealed a cardiac mass. The patient was referred to the Aga Khan University Hospital for Cardiac Magnetic Resonance Imaging (CMR) in December 2014 for further evaluation of the mass.

After taking informed consent from the patient for carrying out the cardiac MRI and telling her that the study may be used for academic writing or educational

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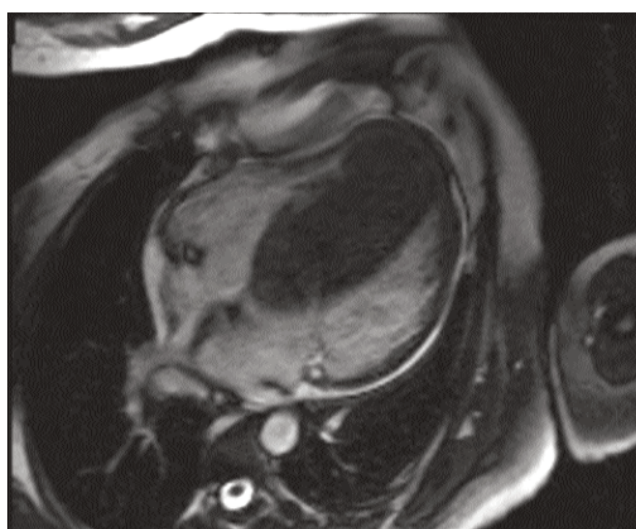


Figure-1: SSFP 4-chamber view showing a hypointense mass in the interventricular septum, occupying most of left ventricular cavity.

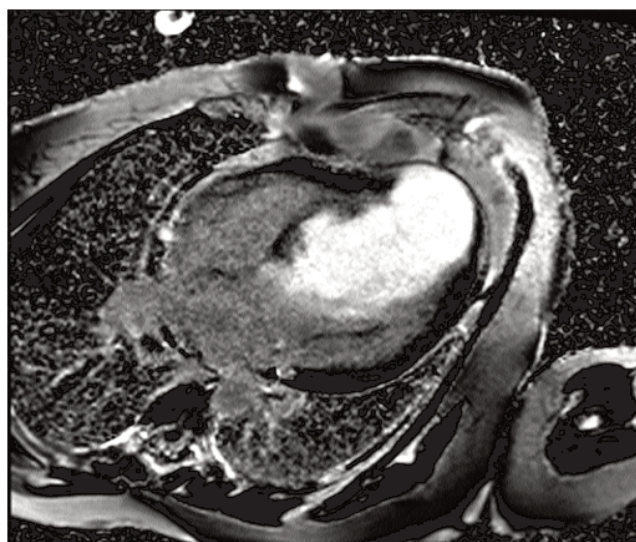


Figure-2: Delayed enhanced imaging (4-chamber view) showing marked late hyper-enhancement of the mass with gadolinium, which is a hallmark of cardiac fibroma.

purposes, CMR was done on Siemens MAGNETOM AVANTO 1.5 Tesla system. The CMR revealed a large intra-cardiac mass (128x75 mm) embedded in the

interventricular septum. It was hypo-intense on Steady State Free Precision (SSFP) images, occupying major part of the left ventricular cavity causing left ventricular outflow tract (LVOT) and left ventricular inflow tract obstruction. Figure-1 shows the SSFP 4-chamber view with the tumour in the interventricular septum and occupying most of the left ventricular cavity. The left ventricular systolic function was also mildly reduced with an ejection fraction of 45%. On Turbo Spin Echo (TSE) T1 weighted images the mass appeared iso-intense to slightly hyper-intense, while on TSE T2 weighted images it appeared hypo-intense. Delayed enhanced imaging with gadolinium showed marked hyper-enhancement of the whole mass as seen in the 4-chamber view in Figure-2. These findings were consistent with cardiac fibroma. The patient was referred for surgical resection of the tumour however; she could not survive the operation.

Discussion

Primary cardiac tumours are rare. They have an autopsy frequency of between 0.001-0.030%, with three-fourth of them being benign.² Almost half of the benign cardiac tumours are myxomas and the rest are papillary fibroelastomas, rhabdomyomas, fibromas and lipomas.²

Cardiac fibroma is the second most common paediatric tumour of the heart.¹ The usual age at presentation is 13 years³ with almost one-third patients presenting at less than one year of age.⁴ Only 15% patients present in adulthood.⁴ Our patient presented quite late at the age of thirty-eight years which is uncommon for cardiac fibromas.

Clinical presentation of patients with cardiac fibroma depends on the location and size of the tumour. Our patient presented with symptoms of progressively worsening shortness of breath. Patients with cardiac fibroma can present with symptoms of heart failure, arrhythmias, or sudden cardiac death. Quite a few cases are discovered incidentally on imaging, whereas a few patients may present with vague symptoms of chest pain. Tumour embolization is rarely seen in cardiac fibroma. The most common presenting symptom of cardiac fibroma is progressively worsening heart failure that occurs due to outflow or inflow tract obstruction, and obliteration of the left ventricular cavity as was seen in our patient.⁵

In a recent systemic review and literature search, the left ventricle was found to be the most common site of cardiac fibroma (57.3%), followed by the right ventricle (27.5%), inter-ventricular septum (17%), right atrium (5.3%) and left atrium (1.8%).⁶ In our case although the tumour was embedded in the inter-ventricular septum, it

was bulging into the left ventricular cavity occupying major part of the left ventricle. Younger age at the time of diagnosis and involvement of the inter-ventricular septum was associated with a greater incidence of arrhythmias, conduction abnormalities and generally a poor prognosis.⁶

A transthoracic echocardiogram is usually the first investigation, whenever a cardiac tumour is suspected. In our patient, the intra-cardiac mass was first identified on a transthoracic echocardiogram. Echocardiography offers excellent information with regards to the location and functional impact of a cardiac tumour; however it has its limitations in terms of tissue characterization. CMR, although time consuming and has limited availability, offers excellent tissue characterization. Unlike computed tomography (CT) it does not expose the patient to ionizing radiation. Hoffmann et al⁷ compared histology of cardiac tumours with CMR using a multi-parametric MR protocol to evaluate signal properties, morphologic characteristics and contrast enhancement of cardiac tumours in 55 patients. MR imaging had a diagnostic accuracy of 0.92 (area under the curve) in determining whether the mass is malignant.

Features on CMR that allow us to differentiate between benign and malignant cardiac tumours include borders, size, location, and pericardial effusion. Benign tumours are well defined with no irregularities or infiltration.⁸ Malignant tumours are more likely to be ill-defined, lobular with invasive borders and may already be invading the pericardium at the time of diagnosis.⁹

Several studies have been done on CMR features of cardiac tumours. The features that have consistently been shown to favour a diagnosis of cardiac fibroma include the following:¹⁰ an intra-myocardial location, especially the interventricular septum, well-defined borders with a thin rim of myocardium, hypo-intense on SSFP images, iso-intense to hyper-intense on T1-weighted images and intense homogenous enhancement on late gadolinium images. Similar findings were observed in our patient.

A number of case reports have now been published that describe the CMR features of cardiac fibroma; however, to the best of our knowledge, this is the first case report of its kind from Pakistan. Availability of histopathology would have provided us more insight into this case but unfortunately it could not be acquired. Our patient was appropriately sent for surgery, which is the only available treatment option for symptomatic patients with cardiac fibroma. Unfortunately the patient did not survive the surgery and died on the table.

Conclusion

Primary cardiac tumours are rare and require a high level of suspicion for their diagnosis. They are lethal if left untreated. An echocardiogram should always be done for patients who present with signs and symptoms of heart failure to identify possible causes of heart failure. Cardiac MRI is an excellent tool for evaluation of patients with heart failure and suspected cardiac masses as they provide excellent tissue characterization and may give information about the possible etiology of cardiac mass thus foregoing the need for myocardial biopsy.

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Conflict of Interest: None to declare.

Funding Disclosure: None to declare.

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