DOI: 10.7860/JCDR/2018/37667.12292

Case Report

Surgery Section

Successful Surgical Management of Cardiac Fibroma with Recurrent Ventricular Tachycardia: A Case Report

AVISHEK SAMADDAR[1], HARKANT SINGH[2], MANOJ KUMAR ROHIT[3], BHALINDER SINGH DHALIWAL[4], VIVEK JASWAL[5]

ABSTRACT

Cardiac fibromas are rare tumours of the heart. We present a case of cardiac fibroma in a four-year-old child in view of rarity of the case and success achieved in its management. The child had presented with a history of intermittent palpitations. Electrocardiography (ECG) showed monomorphic Ventricular Tachycardia (VT). She was initially managed with intravenous Metoprolol and Amiodarone infusion but had recurrent VT. Echocardiography revealed a mass in the apical septum. A Contrast Enhanced Computed Tomography (CECT) showed a homogeneous mass involving the anterolateral and apical aspect of interventicular septum causing expansion of the septum and indentation and compression of both ventricles. The tumour was approached through median sternotomy under Cardiopulmonary Bypass (CPB). The adventitia over the exposed part of the tumour was incised and the tumour was dissected out of the interventricular septum. A bovine pericardial patch was used to close the septal defect. Most of the small tumours dispersed around Left Anterior Descending Artery (LAD) were excised. The patient was weaned off CPB easily.

In the postoperative period, Amiodarone infusion was continued with no recurrence of arrhythmias. Histopathology revealed classical features of fibroma. The child is stable and is on outpatient follow-up.

Our patient had a huge fibroma and we could successfully resect it avoiding need for transplant. We hereby conclude that in children with a huge intracardiac fibroma involving almost a complete cardiac chamber, success can be achieved through resection and reconstruction in selected cases and the need for transplant may be obviated.

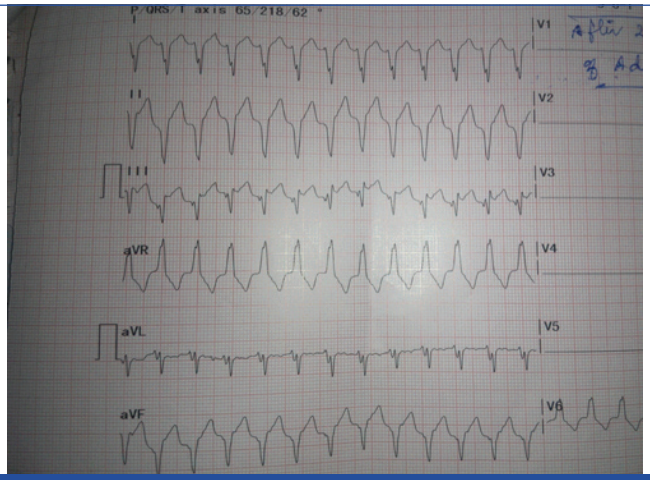
Keywords: Arrhythmia, Cardiac tumour, Fibroma

CASE REPORT

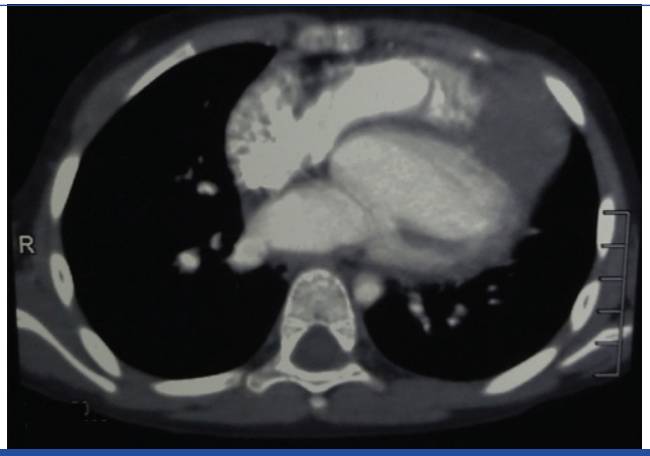
A four-year-old girl was referred to our institute with intermittent palpitations and loss of appetite for one month. The intermittent bouts of palpitations had sudden onset and offset with specific precipitating or relieving factors. There was no other significant history and there was no family history of cardiac diseases or arrhythmias.

On physical examination, the child had low grade fever along with irregular pulses. There was no murmur and the other general and systemic physical examination findings were unremarkable. Initial ECG on admission showed monomorphic VT [Table/Fig-1]. She was managed with intravenous Metoprolol followed by Amiodarone infusion which corrected the arrhythmia initially for the first two days but was followed by recurrences of VT. A transthoracic echocardiography performed on the third day following admission, revealed a homogeneous mass in the apical septum [Video-1]. A CECT scan done subsequently on the next day showed a well defined homogeneous mass involving the anterolateral and apical aspect of interventricular septum measuring 4.2×4.1×4.5cm causing expansion of the septum and indentation and compressing both ventricles. A small exophytic component was seen arising from the right ventricular aspect, extending into pericardial space anterosuperiorly [Table/Fig-2].

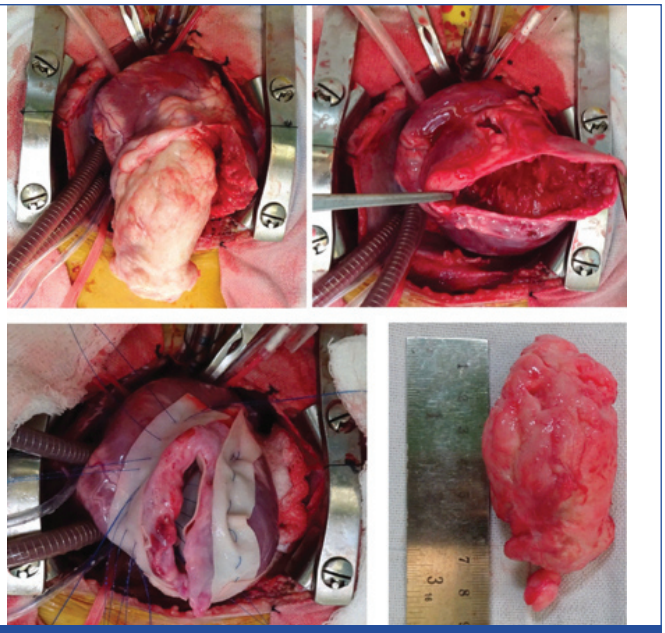
In view of recurrent episodes of VT, surgical excision was planned and the child was taken up for surgery on the seventh day following admission. CPB was initiated with distal ascending aortic arterial cannulation along with single dual-stage right atrial venous cannulation. Antegrade and retrograde routes were used for cardioplegia. Intraoperatively a firm mass of 5×5×5 cm was found arising from the apex of the interventricular septum and protruding into the pericardial space across the ventricular musculature. Resection of the mass resulted in multiple small rents in the ventricular free wall which were repaired using pericardial patch (St. Jude Bovine Pericardial Patch) [Table/Fig-3].



[Table/Fig-1]: ECG of the patient showing monomorphic ventricular tachycardia.



[Table/Fig-2]: CECT image of the patient showing well defined homogeneous mass involving the anterolateral and apical aspect of interventricular septum.



[Table/Fig-3]: The tumour showing globular fibroma and the result of resection and reconstruction of the interventricular septum and the ventricular free wall.

Pathological examination of the mass showed a large, globular fibroma [Table/Fig-3] with microscopic evidence of bland fibroblastic proliferation and monomorphic cells with elongated nucleus, fine chromatin and scanty cytoplasm arranged in fascicles. So the final diagnosis for the child was a case of large intracardiac fibroma involving both the ventricles and the interventricular septum. Postoperatively, patient had an uneventful course with no recurrence of VT and transthoracic echocardiography showed mild Mitral Regurgitation (MR) with normal biventricular function. The patient was discharged on the eighth postoperative day. Patient is at present in our outpatient follow-up and is doing well till date with about a year of follow-up ever since the surgery.

DISCUSSION

A cardiac fibroma is a collection of fibroblasts interspersed among large amounts of collagen. It is the 2nd most common benign cardiac tumour in children [1] occurring rarely in adults [2]. Typically the free wall or interventricular septum of the Left Ventricle (LV) is involved [3], but they can involve any cardiac chamber and even the pulmonary artery [4].

The most common cardiac tumour in children and infants after the rhabdomyoma is a fibroma [5]. Fibroma has been the second most common benign primary cardiac tumour after myxoma in different surgical series reported across all age groups. They are also seen in patients with Gorlin's syndrome with associated features of odontogenic cysts, epidermal cysts, multiple naevi and cutaneous basal cell carcinomas [6]. Diagnosis of these tumours are still rare with medical literature containing mainly case reports and a few small series, inspite of hugely improved imaging modalities.

Cardiac fibromas are connective tissue tumours derived from fibroblasts. Macroscopically they are almost always single arising from the ventricular myocardium or septum. These tumours are firm, gray/white having a whorled appearance and often become very large. Central calcification is common and may be appreciated on radiography, a feature which helps to distinguish them from rhabdomyomas. On microscopy, fibromas appear as nonencapsulated tumours extending into the surrounding myocardium. Hyalinized fibrous tissue with multiple calcification foci and myxoid cystic degeneration often occupy the central portion of the tumours [5].

Echocardiography is the most common initial diagnostic modality. It can provide information about the tumour's extent besides giving an idea about ventricular and valvular function. Cardiac Computed Tomography (CT) or Magnetic Resonance (MR) helps to delineate the mass anatomically and reveals its interaction with adjoining vital structures. The diagnosis on CT is clenched by feature of homogeneous soft-tissue mass that may be either sharply marginated or infiltrative. Presence of dystrophic calcification is common. On Magnetic Resonance Imaging (MRI) the tumours seem homogeneous and hypointense on T2-weighted images and on T1-weighted images they are isointense relative to muscle. Cardiac fibromas often demonstrate little or no contrast enhancement [7].

The natural history of cardiac fibromas is unpredictable. The clinical presentation varies widely: one third of patients are asymptomatic with incidental diagnosis on imaging, another 23% can present with ventricular arrhythmia resulting in sudden demise [2]. The remainder present with chest pain, shortness of breath, fatigue, or syncope [3]. The major concern with asymptomatic tumours is the risk of sudden malignant ventricular arrhythmias. It is henceforth considered prudent to resect asymptomatic fibromas which are large or growing [3].

In 1962, resection of a cardiac fibroma was first performed [8]. Resection may be difficult in view of large size and the anatomic location of fibromas, especially when they encroach upon the coronary arteries or into the ventricular cavity. Postoperative mitral regurgitation can occur secondary to distortion or involvement of papillary muscles. Orthotopic cardiac transplantation has been necessary in cases where the tumour has been extensively large and involve vital, irreparable areas of the heart [9]. However, subtotal excision has also resulted in prolonged survival [3]. Recurrence of cardiac fibromas is unusual, and post-resection, long-term survival rates have been excellent. Gualis J et al., reported a similar case in a 57-year-old male who had presented with VT and post resection was symptom free for a period of nine month follow-up [10].

The patient belonged to the typical age group and presented with non specific symptoms. She had recurrent VT and echocardiography had revealed a cardiac mass. On further imaging with CT scan, the features of homogeneous hypodense mass also confirmed with the common features of fibroma as described in literature. However, there was no calcification detectable on imaging although the histopathology did reveal numerous areas of calcification apart from the classical described features of hyalinised fibrous tissue and gross firm whorled morphology. We hope this case will add to the existing literature pertaining to this rare neoplasm and will aid in the management of similar cases in the days to come.

CONCLUSION

Our patient presented with recurrent VT. On diagnostic imaging of 2D echocardiography and CECT, she was diagnosed with a large intracardiac fibroma involving both the ventricles as well as the interventricular septum. We could successfully resect the mass along with reconstruction of the septum as well as protecting the coronaries. We hence conclude that successful resections of large intracardiac fibromas can be performed in selected patients with proper surgical planning and if resection is adequate, symptomatic relief as well as reverting of arrhythmias can be achieved.

REFERRENCES

[1] Gasparovic H, Coric V, Milicic D, Rajsman G, Burcar I, Stern-Padovan R, et al. Left ventricular fibroma mimicking an acute coronary syndrome. Ann Thorac Surg. 2006;82(5):1891-92.
[2] Darwazah AK, Shoeb J, Eissa SS. Pedunculated endocardial left ventricular fibroma presenting with cerebral and bilateral peripheral embolization. Ann Thorac Surg. 2010;89(3):965-67.
[3] Cho JM, Danielson GK, Puga FJ, Dearani JA, McGregor CG, Tazelaar HD, et al. Surgical resection of ventricular cardiac fibromas: early and late results. Ann Thorac Surg. 2003;76(6):1929-34.

[4] Schroeder JK, Srinivasan V. Intraluminal pulmonary artery fibroma in a 7-year-old boy. Pediatr Cardiol. 2000;21(5):480-82.
[5] Sheppard MN, Mohiaddin R. Tumours of the heart. Future Cardiol. 2010;6(2):181-93.
[6] Yan AT, Coffey DM, Li Y, Chan WS, Shayne AJ, Luu TM, et al. Myocardial fibroma in gorlin syndrome by cardiac magnetic resonance imaging. Circulation. 2006;114:e376-79.
[7] Leja MJ, Perryman L, Reardon MJ. Resection of left ventricular fibroma with subacute papillary muscle rupture. Tex Heart Inst J. 2011;38(3):279-81.
[8] Parks FR Jr, Adams F, Longmire WP Jr. Successful excision of a left ventricular hamartoma. Report of a case. Circulation. 1962;26(6):1316-20.
[9] Burke AP, Rosado-de-Christenson M, Templeton PA, Virmani R. Cardiac fibroma: clinicopathologic correlates and surgical treatment. J Thorac Cardiovasc Surg. 1994;108(5):862-70.
[10] Gualis J, Castaño M, Gómez-Plana J, Martín C, Alonso D. Surgical treatment of giant intramural left ventricular fibroma in an adult patient with refractory ventricular tachycardia. J Card Surg. 2010;25(6):656-58.

PARTICULARS OF CONTRIBUTORS:
1. Senior Resident, Department of CTVS, Post Graduate Institute of Medical Education and Research, Chandigarh, India.
2. Additional Professor, Department of CTVS, Post Graduate Institute of Medical Education and Research, Chandigarh, India.
3. Professor, Department of Cardiology, Post Graduate Institute of Medical Education and Research, Chandigarh, India.
4. Senior Resident, Department of CTVS, Post Graduate Institute of Medical Education and Research, Chandigarh, India.
5. Senior Resident, Department of CTVS, Post Graduate Institute of Medical Education and Research, Chandigarh, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Avishek Samaddar,
House Number 822, Sector 16d, Chandigarh-160015, India.
E-mail: see_avishek@yahoo.co.in

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Jul 22, 2018**
Date of Peer Review: **Aug 08, 2018**
Date of Acceptance: **Aug 13, 2018**
Date of Publishing: **Nov 01, 2018**