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Left Ventricular Fibroma: An Exceptional Case of Primary Tumor Involving Left Ventricle of Heart

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Abstract: Incidence of primary tumor of the Heart is approximately 0.02 percent in autopsy series, among this Fibroma is the second most common type of primary cardiac tumor occurring in the pediatric age group. Most occur before 10 year of age and around one third occur before 1 year of age and during infancy.
Keywords: Primary Tumor of the Heart, Levt Ventricular Tumor, Fibroma of Left Ventricle

INTRODUCTION

Primary tumors of heart has incidence of 0.02 percent and among this, relative incidence of Fibromais around 17 percent. They can present with Heart failure, arrhythmia or embolic phenomena. They are benign connective tissue tumors derived from fibroblast which occur predominantly in children and constitute the second most common type of primary cardiac tumor occurring in pediatric age group.Males and females are equally affected. Cardiac fibromas are typically large tumors ranging from 3-10 cm in diameter. They usually occur within ventricular myocardium much more frequently within the anterior free wall of the left ventricle or the interventricular septum [3]. Approximately 70 percent of fibromas are symptomatic causing mechanical interference with intra cardiac flow (usually with bulky intracavitary tumors), ventricular systolic function or conduction disturbance.

The most common clinical manifestations are congestive cardiac failure (21 percent), ventricular tachyarrhythmia's (13 percent), and atypical chest pain (3.5 percent) [4]. Sudden cardiac deaths occur in 14 percent of patients. Most symptomatic patients have cardiomegaly on chest X-ray and tumor calcification is seen in 25% of cases.

CASE REPORT

In this exceptional case, a baby boy of 2 day age born through normal delivery at a government hospital Bhopal. It was full term normal delivery second issue,presented with history of respiratory distress of the past two day duration along with failure to thrive and poor feeding history, bluish discoloration of lips, fingers and tongue. The patient was admitted on 15 august 2013 for above complaints.

Respiratory rate was 78 per minute,Heart rate was 156 per minute central cyanosis was present, blood pressure was 70/50mmHg, and his oxygen saturation was 92%.

On auscultation he had mid systolic murmur. Second heart sound was loud and single. There was presence of third heart S3 sound. Basal crepetations were presented suggestive of left ventricular failure.

His chest X–Ray was suggestive of cardiomegaly with bilateral basalnon homogenous opacity.

Echocardiography shows evidence of large homogenous intramural mass measuring 7cm by 9 cm arising from the lateral wall of left ventricle which was obscuring the left ventricular cavity significantly. It was not mobile and broad based. The mass was obstructing Aortic valve too. Apart from this, the patient had large ventricular septal defect with left to right shunt (restrictive), septum secundam atrial septal defect, of around 5 mm with left to right shunt, and paraductal discreat Coarctation of aorta with left to right shunt and peak gradient of 22mmHg. There were no evidence of satellite lesions, no evidence of pericardial effusion and pulmonary artery hypertension. There was no evidence of other congenital heart disease. Left ventricular functions were moderately compromised with left ventricular ejection fraction of 30%.

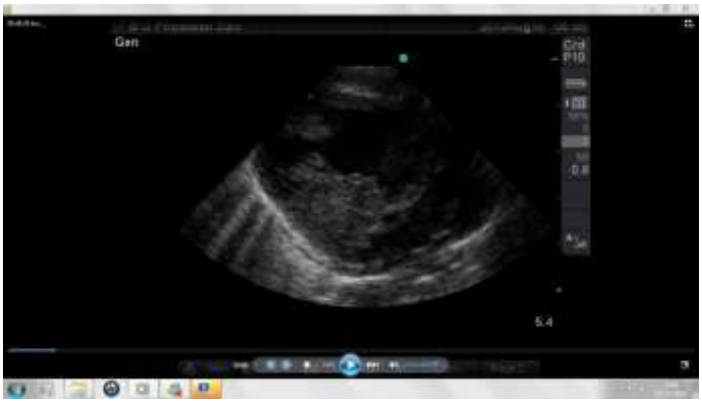

Fig. 1: Large Fibroma with Large Muscular VSD in Long Axis Echocardiography View

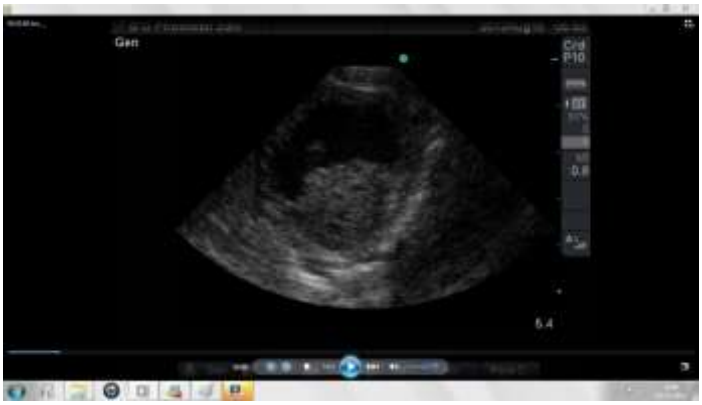

Fig. 1: Fibroma In Short Axis View

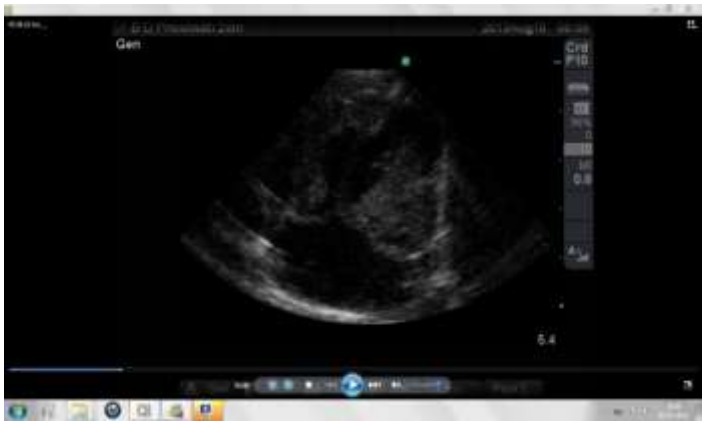

Fig. 3: LV Fibroma in Apical Four Chamber View

Fig. 4: Coarctation of Aorta (Paraductal Discreat) in Patient with LV Fibroma

Management

Baby was put on assisted ventilation, dopamine and dobutaminewas started, along with supportive measures.Surgical excision is challenging but possible. Although they lack capsule and may have extension or satellites. Given the risk of fatal arrhythmia, complete resection is always recommended. As the patient's condition was not appropriate so he could not be taken for surgery and expired after 3 day.

CONCLUSION

Primary tumors of heart has incidence of 0.02 percent and among these, relative incidence of fibroma is around 17 percent. They can present with Heart failure, arrhythmia or embolic phenomena. The most common clinical manifestations are congestive cardiac failure (21percent), ventriculartachy-arrythmias (13 percent), and atypical chest pain (3.5 percent) [4]. Sudden cardiac death occurs in 14% of infants. Intracavatiry tumors are more likely to cause heart failure and embolic phenomena, whereas intramural tumors are more likely to cause arrhythmias. They can be misdiagnosed as rheumatic valve disease, Cardiomyopathy, endocarditis, myocarditis, pulmonary emboli, pulmonary hypertension, cerebrovascular disease and vasculitis. Advances incardiovascular imaging techniques-especially echocardiography, computed tomography and magnetic resonance imaging –have greatly facilitated the diagnostic evaluation and permit the rapid identification of intracardic masses.

Treatment

Surgical excision is challenging but possible. Although they lack capsule and may have extension or satellites. Given the risk of fatal arrhythmia, complete resection is always recommended.

Diagnostic Techniques

Echocardiography usually reveals intramural large solid homogenous echogenic mass with central hyperechoic foci. CT scan revels homogenous mass with low attenuation calcification. CT scan currently appears to be most useful in the evaluation of tumors of the Heart either to provide additional information when the echocardiographic data are equivocal or to determine the degree of myocardial invasion and the involvement of pericardial and extra cardiac structures [5].

MRI Fibroma is usually homogenous and isointense to hyperintense on T1, and hypointense on T2 with minimal enhancement with gadolinium [5, 6]. Gorlin syndrome; it is an autosomal dominant syndrome characterized by cardiac fibroma associated with nevoid basal cell carcinomas, Medulloblastomas and fibrous histiocytomas [7].

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