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Original Article

Myxoma in Pediatric Patients: A Single Centre Experience

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Key Words : *Myxoma, Primary Cardiac Tumor, Paediatric Patients.*

Abstract

Background: *Pediatric cardiac tumor is a very rare entity with an autopsy frequency 0.0017-0.28%. This study aims to analyze the characteristics and outcome of pediatric patients with myxoma.*

Methods: *Nine Patients with myxoma treated between January 2008 & December 2019 were included into this retrospective study. The patients' age ranged from 4 years to 11 years and weight from 10-20 kg. All patients were diagnosed by transthoracic echocardiography.*

Results: *We did complete resection of tumor in all patients under cardiopulmonary bypass. Mean cardiopulmonary bypass time was 55 minutes & mean cross clamp time was 34 means. Mean ICU stay was 3 days. Six (66.33%) tumors found in left atrium, while 2 (22.11%) found in right atrium and 1(11.06%) in right ventricle. Histopathological report of all 9 tumors revealed myxoma. 1 patient developed left sided hemiparesis and another 1 patient developed wound infection. No mortality or recurrence in follow up period.*

Conclusion: *Myxoma is the commonest primary cardiac tumor. Surgery of cardiac tumor can result in good early and long-term outcome.*

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Introduction:

Primary cardiac tumors are rare in pediatric population, with a prevalence ranging from 0.0017 up to 0.28% in different autopsy series. Furthermore, the incidence of cardiac tumors during fetal life has been reported to be 0.14% in autopsy series.[1,2] The vast majority of primary cardiac tumors in children are benign, whereas 10% are malignant. The frequency and type of cardiac tumors in pediatric population differ from those of adults. In the adult population, primary cardiac tumors represent up to 75% of heart neoplasm, while malignant cardiac tumors represent only 25%.[3]

Moreover, myxomas are the most common primary tumors in adults, accounting for 40% of benign tumors. While on the contrary they are exceedingly rare in fetuses and neonates. Rhabdomyoma is the most common cardiac tumors in fetal life and childhood, representing nearly 70% of primary tumor, followed by teratoma, fibroma and hemangioma.[3,4]

Clinical presentation depends on the age of the patient, as well as size and location of the cardiac tumors.[4] Children with cardiac tumor can be asymptomatic and may present with symptoms such as- murmur, arrhythmia, heart failure or sudden death.[4-6]

The main diagnostic tool in the initial evaluation of cardiac tumor is echocardiography. Magnetic resonance imaging (MRI) is the most sensitive examination[3]. Biopsy is the diagnostic gold standard[7].

Treatment modalities include conservative management, surgical resection and transplant

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but treatment choice varies according to the diagnosis and patient symptoms.[4] Surgery is the main approach for patients with symptomatic cardiac tumour.[5,6]

This study aimed to analyze the characteristics and outcome of pediatric patient with myxoma treated in a specialized cardiac center.

Methods:
Nine patients (5 male and 4 female) with primary cardiac tumor treated in National Institute of Cardiovascular Disease (NICVD) under pediatric cardiac surgery department between January 2008 and December 2019 were included into this retrospective review. The most clinical symptom was palpitation and dyspnea. Chest X ray showed mild moderate cardiac enlargement in 6 cases. Echocardiography revealed mass inside the cardiac chamber. Written informed consent was obtained from the patients' family.

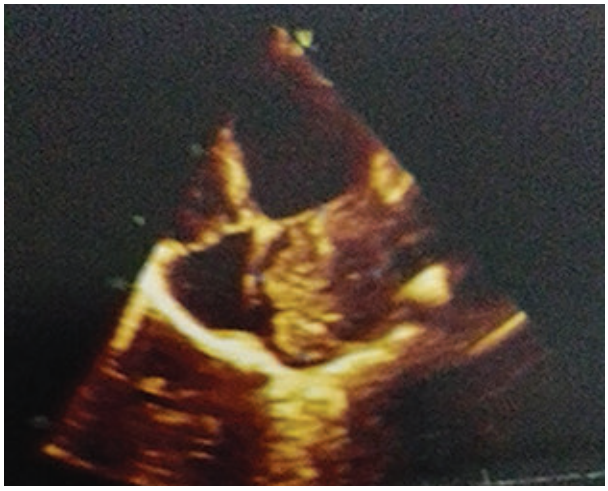

Fig.-1: *Myxoma in left atrium*

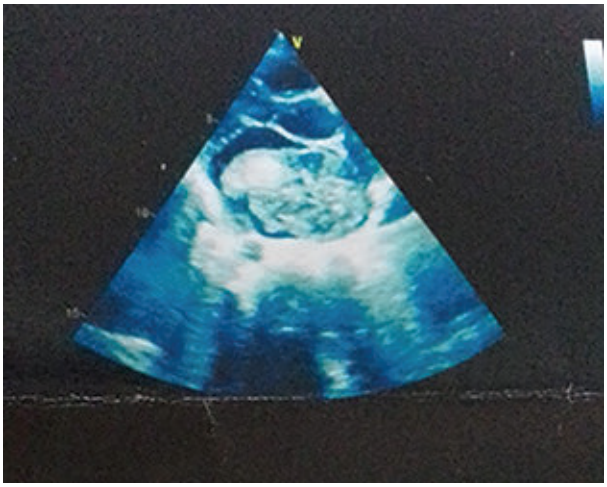

Fig.-2: *Myxoma in left atrium*

Surgical procedure: All surgeries in this study were performed in a median sternotomy. The goal of surgery is to resect the entire tumour mass with sufficient margins.

Cardiopulmonary bypass is instituted in a standard fashion with bi-caval cannulation and heart is protected with cardioplegic solution. Right atriotomy done, incision over the interatrial septum was made where needed. Complete resection of the mass done and all four chamber was checked. Interatrial septum was repaired with autologous pericardial patch in 3 patients.

Patients were not discharged from intensive care unit until circulation and breathing were stable. Electrocardiogram, chest x-ray and echocardiography were done before patient left hospital. Al patients were followed up by phone interview and regular check after operation to assess late functional status.

Results:
The patients age ranges from 4 years to 11 years with weight ranging from 10 kg to 22 kg. In this study patients' characteristic, as well as tumour locations and histological type of tumour, are listed in table I. On admission most of patients were asymptomatic with cardiac murmur.

All patients operated under cardiopulmonary bypass with cardiopulmonary bypass time 45 min-80 min (mean time 55 min) and cross clamp time was 21 min-45 min (mean time 34 min). Overall median ICU stay was 3 days with a median mechanical positive pressure ventilation was 12 hours (6 hours-18 hours).

All the patients were successfully discharged from the hospital. Postoperative complication includes left sided hemiparesis in one patient who recovered within week, one patient had wound infection and others were healthy.

Histopathological examination revealed myxoma in all patients. Left atrium was the most common tumour location found in 6 patients (66.6%), right atrium stood second found in 2 patients (22.22%) and lastly right ventricle with one patient (11.11%).

At a mean follow up of 5 years all patients are alive and recovered well. There was no tumour recurrence or any other complication like valve regurgitation.

Table-I

Clinical data of patients with myxoma.

----Gender	Age (years)	Weight (Kg)	Presenting Symptoms	Location	Tumor size in Echocardiography (cm^2)	Histopathology report
Male	8	17	Dyspnea	RA roof	8.01 cm^2	Myxoma
Female	11	22	Dyspnea	RV at septal leaflet of tricuspid valve	1.2×1.7 cm^2	Myxoma
Female	8	15	Palpitation	LA	2×2.5 cm^2	Myxoma
Male	5	14	Murmur	LA, stalk attached to IAS	3×2.5 cm^2	Myxoma
Male	4	12	Murmur	LA, Stalk at lower portion of IAS	4×3 cm^2	Myxoma
Male	7	15	Murmur	LA, IAS	4.8×3.6 cm^2	Myxoma
Female	4	10	Asymptomatic	LA	2.5×1.5 cm^2	Myxoma
Male	6	13	Palpitation	RA	3×2.5 cm^2	Myxoma
Male	5	15	Asymptomatic	LA	1.7×1.9 cm^2	Myxoma

Table-II

Peri-operative characteristics of study population.

Characteristics	Myxoma
Approach through RA-tomy (number of patients)	03
Approach through Bi-atriotomy (number of patients)	06
CPB time (minutes)	55 (45-80)
Cross clamp time (minutes)	34 (21-45)
Mechanical ventilation time (hours)	12 (6-18)
ICU stay (days)	3 (2-5)

Discussion:

Cardiac neoplasms are rare both in adult and children. The majority of tumours in adult are secondary, arising from non-cardiac tissue, and either invading or metastasis to the heart. While the majority of tumors in the pediatric population are primary, arising from the heart itself.

Primary cardiac tumor may be asymptomatic with up to 12% of case found incidentally during evaluation of a related medical condition.[9] The clinical manifestation of the primary tumours may be related to tumor's size and location, as they enlarge they can progressively obstruct cardiac chambers mimicking valve disease. So, dyspnea may be followed by symptoms of heart failure. In our study 22.22% patients were asymptomatic, 33.33% patients present with murmur, 22.22% with dyspnea and 22.22% patients with palpitation. Liyang et al. reported 43.75% patients present with asymptomatic and 25% present with murmur when undergoing a routine heart check, 16.18% present with dyspnea in whom tumor compress the heart tightly.[10]

Echocardiography plays a very important role in the cardiac tumor diagnosis, which can evaluate the morphology, location and range of the tumour, and also assess the degree of blood flow. Magnetic resonance imaging (MRI) and computed tomography (CT) scan were also necessary.[11,12]

The goal of surgery is to resect the entire mass with sufficient margins, which is almost always achievable with benign cardiac tumor.[13] Most of the primary tumors resection in children require CPB especially when the mass is intracardiac. Surgical approach such as trans atrial, trans ventricular and trans aortic depends on the mass position. In these series, we use only trans atrial approach, surgical resection in the operation needs gentle manipulation of the heart to prevent mass fragmentation and embolization.[10]

The pathological examination of excised cardiac masses in our series had showed myxomas in all patients. Three patients require autologous pericardial patch for repair of interatrial septum.

Rhabdomyoma is the most frequent tumor in the pediatric age group, followed by myxoma, fibroma, hemangioma, whereas malignant tumor is very rare. Our results were not consistent with literature.[14,15] Though rhabdomyomas and fibromas were reported to be higher in the infancy, while myxomas are most frequent in older children.[16] In these series most patients are older age so, myxomas were the most frequent findings.

Myxoma are usually seen single, pedunculated, and adherent to the fossa ovalis. The left atrium in the most common location (90%) but they can be seen on right atrium as well.[17] However, in this series myxoma were found 66.66% in the left atrium and 22.22% in right atrium and interestingly in the right ventricle.

We had a family history of myxomas in one case. Familial occurrence of myxomas has been reported usually seen in younger patients in 7% cases. No history of mortality or recurrence in follow up period.

Conclusion:

The majority of primary cardiac tumors in children are benign and patients presents in a variety of ways. Common presentations are asymptomatic murmur and dyspnea. Echocardiography plays a major role in diagnosis. Surgery is the main modality of treatment with good perioperative outcomes.

Conflict of Interest - None.

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