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Primary cardiac tumours in a paediatric population: An experience from a tertiary centre with a review of literature

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ABSTRACT

Background: To observe the histopathological spectrum of primary cardiac tumours in paediatric population those came in Pathology Department over a period of last 16 years. **Materials and Methods:** During the time period of 16 years (1995-2010), we had received 16 cases of primary cardiac tumours in paediatric patients. The tumour diagnosis and subtyping was done by histopathological examination along with cytochemistry and immunohistochemistry. **Results:** Benign cardiac tumours were much more common (15 cases) than the malignant tumours. Among these, myxoma was the most frequent (13 cases). The other benign cardiac tumours were rhabdomyoma (one case) and fibroma (one case). A primary malignant cardiac tumour was diagnosed in one case and was labelled as undifferentiated sarcoma. **Conclusions:** The present study reveals the pathological spectrum of surgically excised cardiac tumours in the paediatric population in northern India. Although the diagnosis of cardiac masses can be made by routine imaging techniques, but the role of pathologist is important for exact characterisation of tumour subtype.

Key words: Fibroma, myxoma, primary cardiac tumour, primary malignant cardiac tumour, undifferentiated sarcoma

INTRODUCTION

Primary cardiac tumours are rare in paediatric population with a prevalence of 0.0017-0.28 in autopsy series. The incidence of cardiac tumours during foetal life has been reported to be approximately 0.14%.[1-3]

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The vast majority of primary cardiac tumours in children are benign, while approximately 10% are malignant. Secondary malignant tumours are 10-20 times more prevalent than primary malignant tumours.[4] The frequency and type of cardiac tumours in adults differ from children with 75% being benign and 25% being malignant. Rhabdomyoma is the most common cardiac tumour during foetal life and childhood. This is usually followed by teratoma, fibroma, myxoma and haemangioma.[3] Myxoma is exceedingly rare in foetuses and neonates.[4-7] The symptoms are more often determined by location than by histological type and any cardiovascular symptom can be a clinical manifestation of a cardiac tumour.[8] Echocardiography or Magnetic Resonance Imaging (MRI) is usually needed to make a clinical diagnosis of cardiac tumours. Cardiac catheterisation is rarely necessary. Histopathological examination of excised tumour of biopsy remains as the gold standard for confirmation of the diagnosis.[9] Here we report a series of paediatric cardiac tumours that came in the Department of Pathology in the last 16 years (1995-2010).

MATERIALS AND METHODS

During the time period of 16 years (1995-2010), we received 16 surgically excised cardiac tumours in paediatric patients for histopathological examination.

All the specimens were fixed in formalin, routinely processed for light microscopy and stained with haematoxylin and Eosin. Special histochemical stains like Periodic acid Schiff (PAS), Periodic acid Schiff with diastase (PAS-D), alcian blue and Masson trichrome (MT) were performed wherever was required. The selected cases were additionally stained with immunohistochemistry using a large panel of antibodies or markers through avidin-biotin peroxidase method (ABC) against vimentin (Diagnostic biosystem 1:200), desmin (Dako 1:100), myoglobin (Novacastra 1:75), smooth muscle actin (SMA) (Novacastra 1:400),

S100-protein (Dako 1:800), synaptophysin (thermo scientific 1:400), chromogranin (Thermo scientific 1:800), Ki-67 protein (MIB-1) (Dako 1:400) and cluster of differentiation (CD) 99 (MIC-2) (Dako 1:100).

RESULTS

A total of 16 paediatric cases specimens were received in the Department of Pathology as primary cardiac tumours. Out of these 16 cases of primary cardiac tumours, 15 were benign and one was malignant. The incidence of primary benign neoplasm 93.75% (15/16) was much higher than malignant one 6.25% (1/16).

In the cases with paediatric cardiac tumours (16 cases), the age ranges from 2 months-14 years (mean 9.5 years). The male to female ratio was 1.3:1.

Of the 15 benign paediatric cardiac tumours cases, the most common site of involvement was the left atrium (LA) (11/15, 73.33%), followed by right atrium (RA) (3/15, 20%) and right ventricle (RV) (1/15, 0.06%). The malignant one was involving the left atrium (LA). Left ventricle (LV) was not involved in any case [Table 1].

The most common histopathologic subtype among primary benign tumours was myxoma (13/15, 86.66%). The other benign tumours in this series were fibroma (1/15) and rhabdomyoma (1/15). The only malignant case was undifferentiated sarcoma. The detail histopathological findings are described below respectively.

Table 1: Histopathology and location of paediatric cardiac tumours

Location		LA	RA	LV	RV	N
Benign	Myxoma	12	1			13
	Fibroma		1			1
	Rhabdomyoma				1	1
Malignant	Sarcoma, undifferentiated	1				1
N		13	2	0	1	16

There were 13 paediatric cases diagnosed as myxoma. Seven of them were male patients and six were female. The age ranged from 6 year-14 years (mean age 10.53 years). Eleven of them arose from LA and two from RA. The tumour size ranged from 3 × 2 × 2 cm to 7 × 5 × 4 cm. Cut surface of the most of the tumours was grey-white and gelatinous with focal areas of calcification. The histopathological examination showed a hypo-cellular tumour comprising of stellate or lepidic cells in a myxomatous background. There were many foci of old haemorrhage and calcification. These cells had scant to moderate amount of eosinophilic cytoplasm and elongated to star shaped nuclei. Interspersed few thin as well as thick wall blood vessels were also identified [Figure 1].

A case of fibroma occurred in a 3-year-old male child who presented with dyspnoea and breathlessness. The echocardiographic finding showed a right atrial mass measuring 2.5 × 1.5 × 1 cm. Gross examination showed a firm white lobulated mass and the histopathologic examination showed a benign spindle cell tumour arranged in short fascicles along with areas of hyalinisation. No mitoses or cellular atypia were found. Foci of dystrophic calcification were also identified. The diagnosis of fibroma was made [Figure 2].

Rhabdomyoma was seen in a 2-month-old male child. The echocardiography showed a huge mass measuring 5 × 5 × 4 cm. The mass was excised and sent for histopathological examination. The histopathology of mass showed a tumour with features of a rhabdomyoma. The tumour was well circumscribed and was composed of vacuolated cells along with varying numbers of myofibres. Some spider cells were also seen.

Undifferentiated sarcoma was diagnosed in a 12-year-old female who presented with a huge mass in the RV

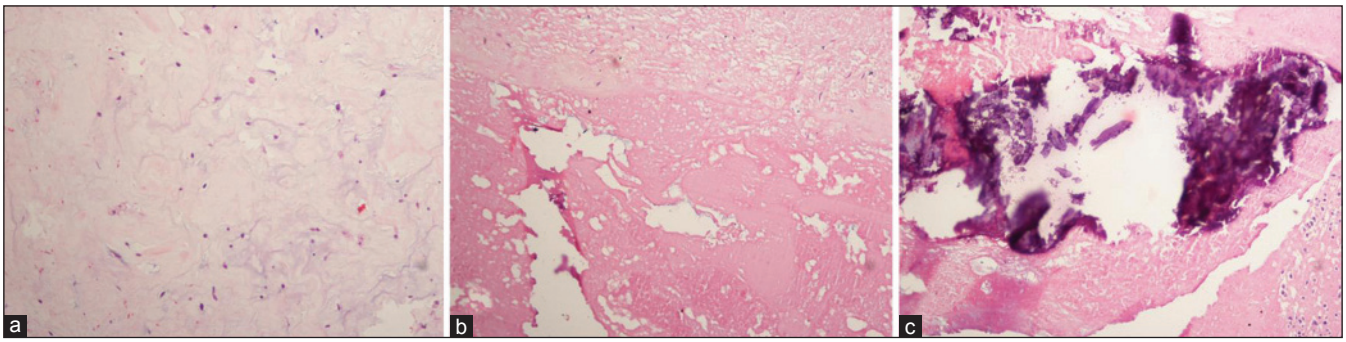


Figure 1: Photographs show features of a myxoma. There are areas of myxoid change with stellate cells or lepidic cells lying in-between (a) (H & E 100×). There are foci of fibrin insudation (b) (H & E 100×) and calcification (c) (H & E 100×)

measuring 5 × 4 × 2 cm. The mass was excised and sent to pathology department. The gross examination of mass showed that it was a well circumscribed and shows gelatinous changes. The histopathological examination the tumour showed large areas of myxomatous change along with fibrin insudation. At the periphery, there were many clusters of atypical tumour cells. These tumour cells were polygonal to spindle in shape with high nuclear cytoplasmic ratio and frequent presence of mitosis. The tumour cells were immunopositive for vimentin and MIB-1 labelling index was high. The other immunohistochemical stains (desmin, myogenin, leukocyte common antigen, C99, synaptophysin, chromogranin and pan-cytokeratin) were negative. So the diagnosis of an undifferentiated sarcoma was given [Figure 3].

DISCUSSION

Primary paediatric cardiac tumours are rare.[1] The vast majority of primary cardiac tumours in children are benign, whilst approximately 10% are malignant.[4] In our tertiary care centre, we had seen a total 16 cases of primary paediatric cardiac tumours over a time period of 16 years.

Myxoma is a benign neoplasm derived from cardiac multipotential mesenchymal cells. This is the most common primary cardiac tumour in adults (65%) with particular predilection to females. In children, it account for only 2-4% of all tumours.[9] The LA is the most common location (90%), but they can be seen in the RA as well.[5] In the present series, myxoma was the most common tumour and was 81.25% (13/16) of all paediatric cardiac tumours.

According to the western literature, the most common primary cardiac tumours in infants and children are rhabdomyoma. It accounts for more than 60% of all primary cardiac tumours. Rhabdomyomas are usually located within the ventricles.[6,7] Rhabdomyoma cells usually lose the ability to divide and hence, regression of the tumour in infancy is an expected outcome. Complete resolutions in more than 80% of the tumours occur during early childhood.[10] Our series had only a single case of rhabdomyoma in a 2-month-old child that was located in RV. Most of the cases in our series were relatively older in age as rhabdomyoma is much more common in infants and neonates.

Fibromas are the solitary benign tumours that account for 12-16% of all primary cardiac tumours in children.[9]

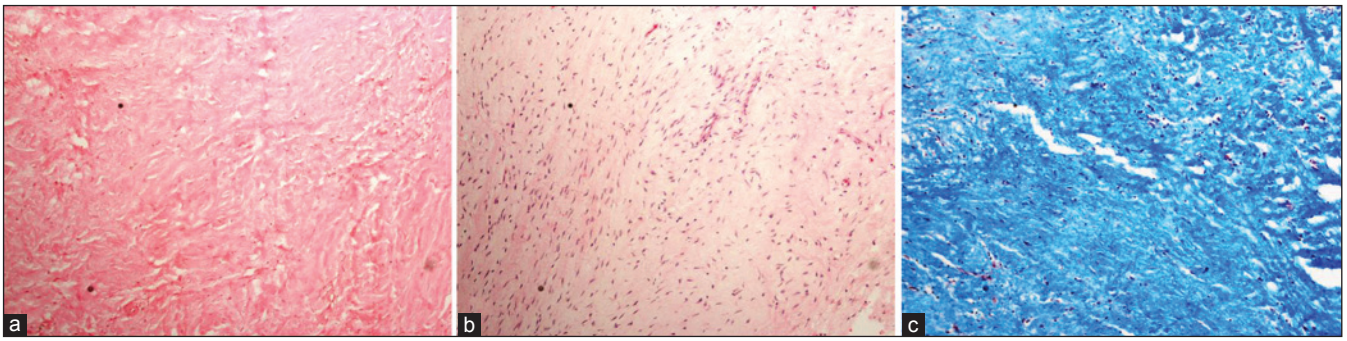


Figure 2: Photomicrographs show features of a fibroma. There are hypo cellular areas demonstrating extensive collagen deposition (a) (H & E 100×) along with cellular areas showing benign spindle cells without any atypia (b) (H & E 100×). Special stain Masson's trichrome show blue colour due to collagen (c) (MT 100×)

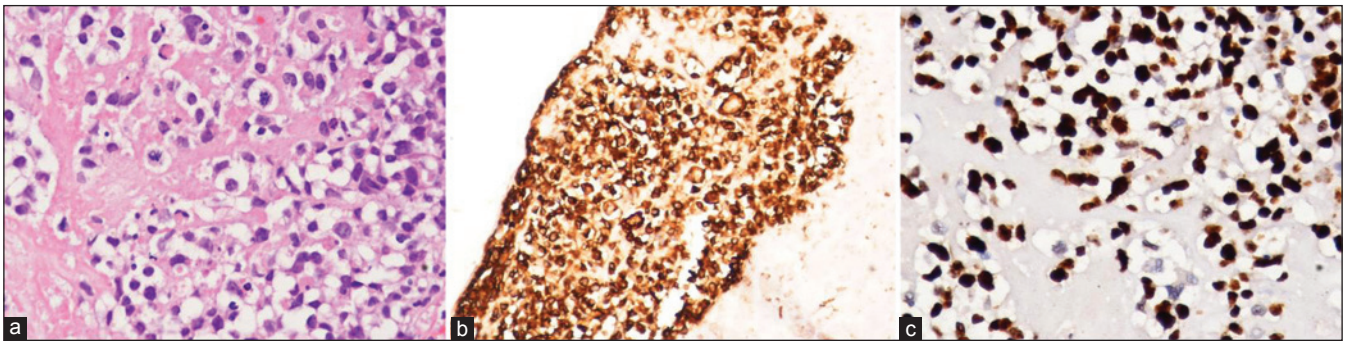


Figure 3: Photomicrographs show an undifferentiated sarcoma. The tumour cells show high nuclear-cytoplasmic ratio and pleomorphism (a) (H & E 400×). Tumour cells are immunopositive for vimentin (b) (IHC (vimentin) 200×) and have high MIB-1 labelling index (c) (IHC (MIB-1) 200×]

Fibroma is derived from connective tissue fibroblasts. Their size may vary from 1 cm – 10 cm. These are mainly located in the ventricular septum. Cardiac fibromas usually remain dormant and spontaneous regression rarely occurs, therefore, total surgical resection is normally recommended.[11] Our series had a single case of fibroma and that was located in the RA.

Malignant cardiac tumours are less common in children and account for 4% of all cardiac tumours.[9] Ninety-five percent of these primary malignancies are sarcomas and the other 5% are lymphomas. Sarcomas are more common in adults as compared to children and most commonly located in the RA. The clinical course is usually aggressive with extensive local infiltration, intracavity obstruction and death.[12] In our series, we had a single case of malignant cardiac tumour (undifferentiated sarcoma) with a prevalence of 6.25% (1/16) and that was located in the LA.

CONCLUSION

Primary cardiac tumours are uncommon in children. In the present series, we have analysed the histopathological spectrum of surgically excised primary cardiac tumours in paediatric population. In contrast to the available literature, myxoma is the most common cardiac tumour in our study. The disparity is probably due to the age of the patients, which is much higher in our cases. Though echocardiography and MRI are helpful, tumour tissue biopsy or the surgically excised specimens examined by a trained pathologist is crucial in making the accurate diagnosis.

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