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Original Article / Özgün Makale

Rare operations in pediatric heart surgery: Cardiac tumors in childhood

Pediatrik kalp cerrahisinde nadir ameliyatlar: Çocukluk çağı kalp tümörleri

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ABSTRACT

Background: In this study, we present our 12-year experience in the surgical treatment of primary cardiac tumors in childhood.

Methods: Thirteen pediatric patients (8 males, 5 females; mean age 1.3±1.9 years; range, 3 days to 6 years) who were operated for a primary cardiac tumor in our center between January 2005 and December 2017 were included in this study. The data were evaluated retrospectively based on our medical records.

Results: All of the masses resected were benign. However, the most common tumor was rhabdomyoma (n=7), followed by fibroma (n=3), myxoma (n=2), and pericardial teratoma (n=1). The mortality rate was 15.4%, as two patients died in the early postoperative period. No residual mass or tumor recurrence was observed in the early and late postoperative period in the remaining patients.

Conclusion: Although primary cardiac tumors in childhood are usually benign, they may cause clinically significant problems depending on the localization and size of the tumor. Surgical tumor excision is often associated with good long-term outcomes.

Keywords: Childhood; heart surgery; primary cardiac tumors.

ÖZ

Amaç: Bu çalışmada çocukluk çağı primer kalp tümörlerinin cerrahi tedavisine ilişkin 12 yıllık deneyimimiz sunuldu.

Çalışma planı: Ocak 2005 - Aralık 2017 tarihleri arasında merkezimizde primer kalp tümörü nedeni ile ameliyat edilen 13 pediatrik hasta (8 erkek, 5 kız; ort. yaş 1.3±1.9 yıl; dağılım, 3 gün-6 yıl) bu çalışmaya alındı. Veriler hastanemiz tıbbi kayıtlarından retrospektif olarak incelendi.

Bulgular: Rezeke edilen kitlelerin tümü benign idi. En sık görülen tümör rabdomiyomayı takiben (n=7), fibroma (n=3), miksoma (n=2) ve perikardiyal teratoma (n=1) idi. İki hasta ameliyat sonrası erken dönemde kaybedildiği için mortalite oranı %15.4 idi. Diğer hastalarda ameliyat sonrası erken ve geç dönemde rezidüel kitle veya tümör nüksü görülmedi.

Sonuç: Çocukluk çağı primer kalp tümörleri genellikle benign olmakla birlikte, tümörün yerleşimi ve büyüklüğüne bağlı olarak klinik açıdan önemli sorunlara yol açabilmektedirler. Cerrahi tümör rezeksiyonu genellikle uzun dönem iyi sonuçlar ile ilişkilidir.

Anahtar sözcükler: Çocukluk çağı; kalp cerrahisi; primer kalp tümörleri.

The primary cardiac tumor is an extremely rare disease with an incidence ranging between 0.001% and 0.3% based upon the data of autopsy series.[1] With the advancements in the in imaging modalities, these tumors have been increasingly diagnosed with their prominent distinctive features.[2]

The majority of the primary cardiac tumors during childhood are benign, while only about 10% are malignant.[1] While myxomas are the most common benign tumors in adults, the most common benign tumors in children are rhabdomyomas, usually followed by teratomas, fibromas, and hemangiomas.[3-5]

Cardiac tumors may be symptomatic during the fetal or post-natal period. Their symptoms vary depending on the size and localization of the tumor. The most common symptoms during the fetal period include arrhythmias, congestive heart failure, fetal hydrops, and albeit rare, stillbirth. Inflow or outflow obstruction symptoms are often prominent depending on the tumor localization in the post-natal period. The

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other symptoms include cyanosis, murmur, respiratory distress, myocardial dysfunction, valve failure, and sudden death.[6]

The diagnosis can be often made with echocardiographic examination or magnetic resonance imaging.[7,8] Cardiac catheterization is rarely required. Tumor biopsy and histopathological assessment still remain gold standards for the definitive diagnosis.[1]

Surgical resection of the tumoral masses can be performed with low intra- and postoperative risks. The main goal of total excision of the tumoral mass is to prevent postoperative complications, such as embolism or arrhythmia.

In the present study, we present our 12-year surgical experience in primary heart tumors which are usually published as case reports or multicenter reviews in the literature.

PATIENTS AND METHODS

A total of 13 patients (8 males, 5 females; mean age 1.3±1.9 years; range, 3 days to 6 years) with primary cardiac tumors were operated in our center between January 2005 and December 2017. The mean body weight was 7.0±5.2 kg (range, 2.4 to 18.1 kg). These patients constituted only 0.6% of all open-heart surgeries performed within the same period in our center. Medical records were retrospectively reviewed with the permission of the hospital's Medical Specialty Training Committee. Non-tumor cardiac masses were excluded from the study. A written informed consent was obtained from each parent. The study protocol was approved by the Dr. Sami Ulus Maternity and Children's Training and Research Hospital Medical Specialty Training Committee. The study was conducted in accordance with the principles of the Declaration of Helsinki.

For all patients, electrocardiographic, telecardiographic, and echocardiographic findings, surgical techniques, and follow-up echocardiographic findings were evaluated. Echocardiography was performed preoperatively, and at one week, one month, and six months after surgery. The volume and systolic function of the heart cavities, the size and location of the preoperative mass, and postoperative residual recurrence rates were recorded.

All operations were performed using median sternotomy. The patients with intrapericardial masses underwent off-pump surgery, while the remaining patients underwent cardiopulmonary bypass with aortic and bicaval venous cannulation. Myocardial protection was maintained with moderate systemic hypothermia, and St. Thomas' cold crystalloid cardioplegia. Right and/or left atriotomy or ventriculotomy was performed, depending on the tumor location. Concomitant procedures were tricuspid valvuloplasty in three patients, ventricular septal defect closure in two patients, and atrial septal defect or patent foramen ovale closure in five patients. The resected masses were placed into formaldehyde solution and transferred to the pathology laboratory. The patients operated were followed every six months during the first postoperative year and every 12 months, thereafter.

Statistical analysis

Statistical analysis was performed using the SPSS version 11.0 software (SPSS Inc., Chicago, IL, USA). Descriptive statistics were expressed in mean ± standard deviation (SD), median, number (n), and

Table 1. Demographic data (n=13)

Demographics	n	Mean±SD	Median	Min-Max
Age (year)		1.3±1.9	8 month	3 days-6 years
Gender				
Male	8			
Female	5			
Weight (kg)		7.3±1.2	4.1	2400-18.1
Length of hospital stay (days)		11.3±8.2	11	1-20
Localization				
Left atrium	2			
Right ventricle	6			
Left ventricle	4			
Pericardium	1			

SD: Standard deviation; Min: Minimum; Max: Maximum.

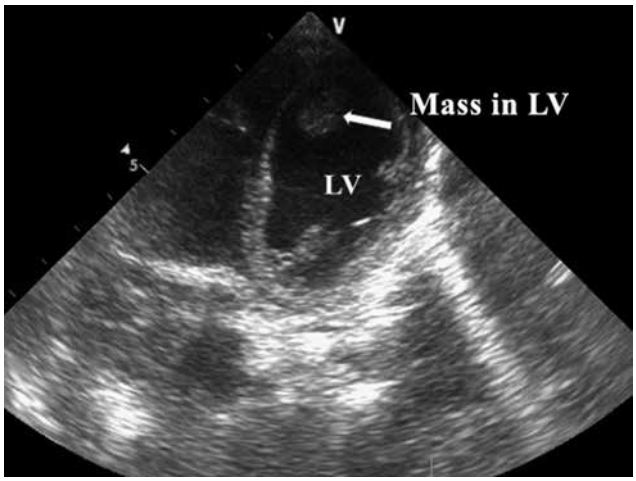

Figure 1. Echocardiographic image of left ventricular mass.
LV: Left ventricle.

frequency (%). A *p* value of p<0.5 was considered statistically significant.

RESULTS

The patients were referred to our clinic for surgery due to symptoms of mechanical obstruction or signs of severe heart failure or uncontrolled arrhythmias. All patients with rhabdomyomas received everolimus treatment, although the masses did not reduce in size with a hemodynamically significant obstruction. The demographic data of the patients are shown in Table 1.

The most common clinical symptoms were a heart murmur, cyanosis, feeding intolerance, palpitations, and dyspnea. Chest X-ray showed mild-to-moderate cardiac enlargement and pulmonary oligemia in two patients. Echocardiography revealed masses inside three cardiac chambers and in the intrapericardial cavity. Preoperative echocardiographic evaluations revealed the presence of a mass lesion with a reduced ventricular volume either in the right or left ventricle in seven patients (Figures 1 and 2). Two patients had a left atrial mass with the stalk (Figure 3). Three patients had a motile mass attached to the papillary muscles and tricuspid valve located in the right ventricle. In one patient, the pressure from the pericardial mass located on the aorta and pulmonary artery caused anterior cardiac tamponade. Localization of the masses and associated heart defects are given in Table 2.

Two tumors located in the left atrium were resected from the left atrium after opening the septum secundum. In these patients, the interatrial septum was initially repaired. In two patients, tumors located in the right ventricle and obstructed the pulmonary outflow tract were resected via pulmonary arteriotomy. In one of these patients, the incision was extended through the right ventricle.

In patients with right ventricular tumors, right ventriculotomy was performed, and the tumors extending into the ventricular cavity were resected via ventriculotomy. The tumors attached to the papillary muscle and tricuspid valve in three patients were resected through the right atrium via the transvalvular route. In all three patients, tricuspid valvuloplasty was performed, after the resection and the leaflet coaptation was achieved. Resection in four patients was made by aortotomy due to the obstruction of the left ventricular

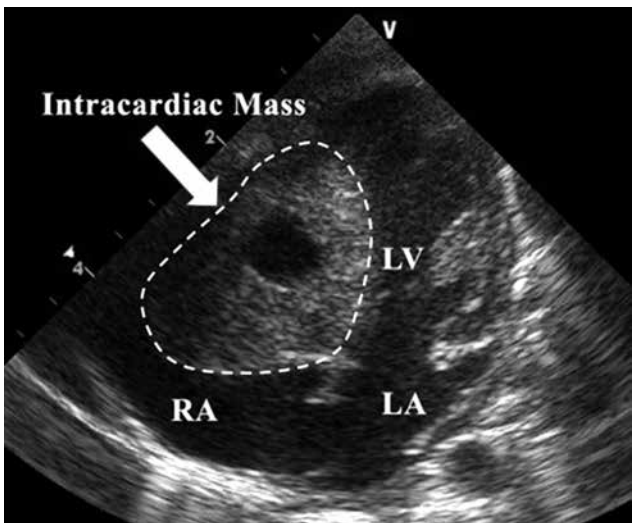

Figure 2. Echocardiographic image of right ventricular mass.
LV: Left ventricle; RA: Right atrium; LA: Left atrium.

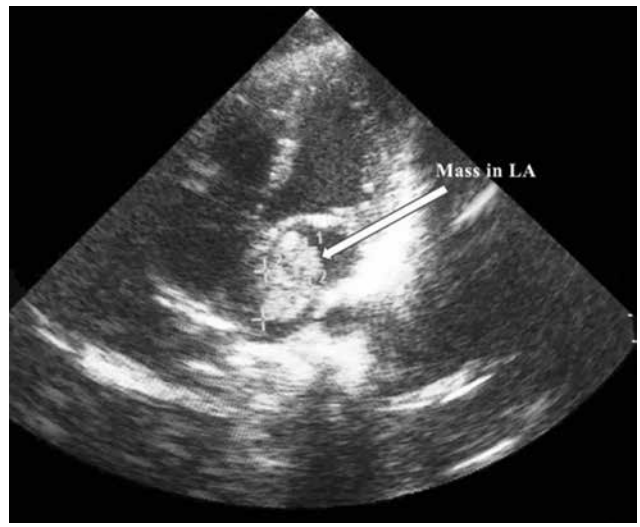

Figure 3. Echocardiographic image of left atrial mass.
LA: Left atrium.

Table 2. Tumor localization, operation, and postoperative status

Number	Age	Weight	Gender	Tumor	Site	Associated procedure	Excision	CPB	Survival status	Cause of dead
1	6 years	18.1	M	Myxoma	LA		Complete	Yes	Alive	
2	18 months	10.3	F	Myxoma	LA	VSD	Complete	Yes	Alive	
3	5 years	15.2	M	Fibroma	RV	VSD	Complete	Yes	Alive	
4	18 days	3.1	F	Teratoma	Pericardium	PFO	Complete	No	Alive	
5	13 days	3.2	F	Rhabdomyoma	RV	PFO	Complete	Yes	Exitus	LCO
6	9 months	6.5	M	Rhabdomyoma	LV		Complete	Yes	Alive	
7	13 months	8.3	M	Fibroma	RV		Complete	Yes	Alive	
8	4 days	2.7	M	Rhabdomyoma	RV	PFO	Complete	Yes	Alive	
9	5 days	2.4	F	Rhabdomyoma	LV	ASD	Complete	Yes	Exitus	LCO/CA
10	4 months	3.7	M	Rhabdomyoma	RV	PFO	Complete	Yes	Alive	
11	8 months	4.1	M	Rhabdomyoma	LV		Complete	Yes	Alive	
12	40 days	2.8	M	Rhabdomyoma	LV	PFO	Complete	Yes	Alive	
13	16 months	11.0	F	Fibroma	RV		Complete	Yes	Alive	

CPB: Cardiopulmonary bypass; LA: Left atrium; VSD: Ventricular septal defect; RV: Right ventricle; PFO: Patent foramen ovale; LCO: Low cardiac output; LV: Left ventricle; ASD: Atrial septal defect; CA: Cardiac arrhythmias.

outflow tract. The patient who had a pericardial mass was also operated without cardiopulmonary bypass, and the mass was completely resected after sternotomy (Figures 4 and 5).

The pathological diagnosis of three masses resected from the right ventricle was reported as rhabdomyomas. Three tumors attached to the papillary muscle and tricuspid valve were fibromas. The pathological diagnosis of two masses resected from the left atrium was reported as myxomas. All of the masses resected from the left ventricle were diagnosed as rhabdomyomas (Figure 6), and the pathological diagnosis of the pericardial tumor was a teratoma.

All patients were followed in the intensive care unit after the operation. The mean duration of mechanical ventilation was 35.8±31.7 h (median: 29.5 h) and the mean duration of stay in the intensive care unit was 5.5±6.0 days (median: 3 days). The mean inotropic score (IS) in the intensive care unit was 21.0±24.6 (median: 10) [IS= dopamine (μg/kg/min) + dobutamine (μg/kg/min) + 100 × adrenaline (μg/kg/min)], and the mean vasoactive inotropic score (VIS) was 21.1±24.6 (median: 10.3) [VIS= IS + 10 × milrinone (μg/kg/min) + 10.000 × vasopressin (unit/kg/min) + 100 × norepinephrine (μg/kg/min)].[9] The mean duration of hospital stay was 11.4±8.2 days (median: 11 days).

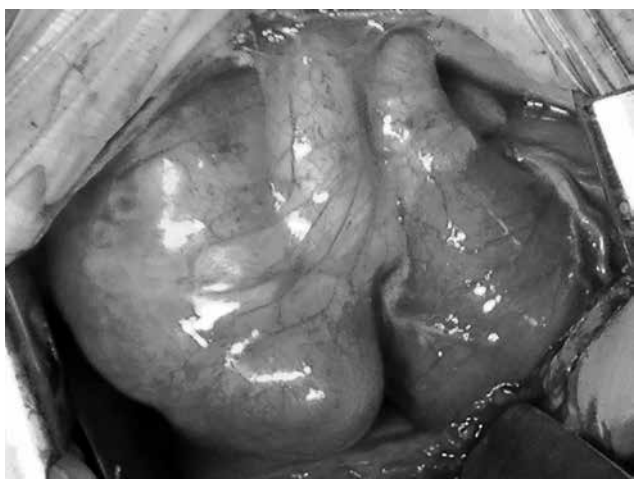

Figure 4. Pericardial teratoma.

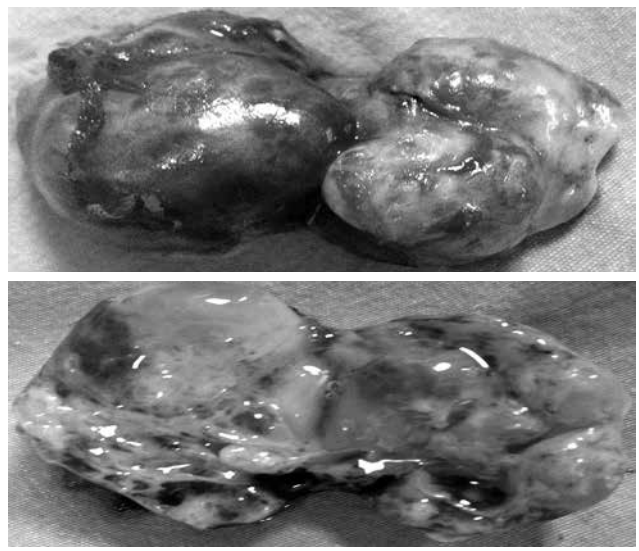

Figure 5. Teratoma.

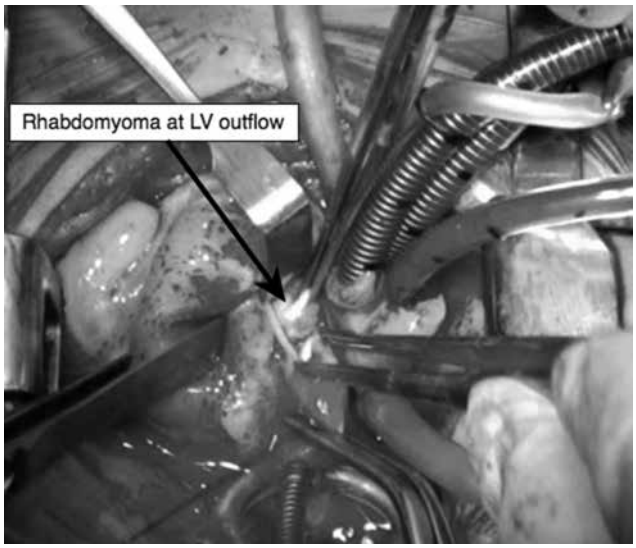

Figure 6. Transaortic resection of left ventricular rhabdomyoma. LV: Left ventricular.

In-hospital mortality was seen in two patients (15.4%) who were five days old and 13 years old (Table 2). Due to the presence of severe heart failure, respiratory distress and cyanosis in the preoperative period, these patients were under mechanical ventilatory support. Their preoperative inotropic scores were 70 and 80. Frequent recurrent supraventricular tachycardia and ventricular tachycardia were also present. Tumoral tissue attached to the interventricular septum in both patients constituted 70 to 80% of the right ventricular cavity. Early postoperative echocardiographic examinations revealed no residual tumor tissue in these patients, but biventricular dysfunctions. The five-day-old patient died due to uncontrolled ventricular fibrillation and cardiac arrest following low cardiac output on the postoperative second day. The 13-year-old patient died due to low cardiac output and multiple organ failure on the postoperative third day. There were no complications in the rest of the patients during the early postoperative period and long-term follow-up.

The mean duration of follow-up was 64.0±47.7 (range: 5.1 to 135) months in surviving patients. Echocardiographic examinations showed no recurrence or residual mass.

DISCUSSION

In the present study, we present our findings in patients operated due to primary cardiac tumors over a period of 12 years and compare our results in the light of literature data. The results of pathological diagnosis showed that all patients had benign tumors, and rhabdomyoma was the most common tumor (53%, n=7). Our results are consistent with the literature in terms of the pathological diagnosis.[5,10,11]

Although rhabdomyomas respond very well to medical therapy today, surgical resection still plays an important role in rhabdomyomas' treatment.[12] Since spontaneous regression and response to medical treatment are seen frequently, the need for surgical resection is reported as 16 to 25% in the literature.[13,14] Resection was required for benign tumors due to cardiac dysfunction caused by their size and localization. Surgically resected rhabdomyomas originated from the right ventricle in three patients and the left ventricle in four patients. In their study, Nir et al.[15] reported a 60 to 80% association between rhabdomyomas and tuberous sclerosis, although tuberous sclerosis was not detected in our patients.

Tumoral tissue resected from the left atrium was pathologically diagnosed as a myxoma in two patients. While myxomas are the most common primary cardiac tumors in adults, they are extremely rare in childhood. Although myxomas are resectable tumors without an aggressive course in adults, they may grow aggressively and require resection in infants.[16] Therefore, myxomas often have a fatal progression in newborns and infants.[17] Mortality was not observed in our patients with a myxoma.

Fibromas were the second most common cardiac tumor in our study, accounting for three patients. As fibromas have a non-encapsulated nature, they often progress with myocardial invasion and, therefore, frequently require resection.[18] Thomas-de-Montpréville et al.[19] reported that fibromas were non-malignant tumors with a high mortality rate of 33%. Myocardial invasion of the tumoral tissue was not seen in our patient group, and in all of our patients, the tumoral tissue was found to be a pedunculated papillary lesion associated with the tricuspid valve. No recurrence and mortality was also observed after resection.

One patient underwent resection of a pericardial teratoma. In this patient, consistent with the literature, teratoma had an extracardiac origin.[16,20] The patient was operated due to the symptoms of tamponade and the tumor was resected. No recurrence or hemodynamic problems were observed during follow-up. In addition, no residual tumor tissue and need for reoperation were seen in any patient.

Childhood cardiac tumors have good outcomes after resection; however, mortality occurs due to insufficient myocardial tissue, which enables normal cardiac function after the resection of the tumoral tissue. In our patient group, two patients were lost in

the early postoperative period due to heart failure and low cardiac output. In these patients, the tumoral tissue, which was pathologically diagnosed as a rhabdomyoma, invaded a large part of the right ventricular cavity, and myocardial tissue was highly disrupted. The remaining patients had normal heart functions in the short- and long-term follow-up.

On the other hand, this study has several limitations. A small number of patients and the limitations inherent to the surgical therapy of cardiac tumors in infants and children are the main limitations. In addition, inclusion of interpretations of the echocardiographic studies from a number clinicians would improve the evaluation, which was unable to be performed in our study. Also, including the interpretations of the echocardiographic studies from a number of clinicians would improve the evaluation, which was unable to be performed in our study. Finally, advanced diagnostic techniques were unable to be used throughout the study, which adds to the list of our limitations.

In conclusion, cardiac tumors during childhood are extremely rare and mostly benign. It should be kept in mind that the patient groups requiring surgery are usually the newborns and the infants, and there can be an additional risk of mortality depending on the localization and the size of the tumor tissue. Nevertheless, tumor resection in the pediatric age group is associated with good long-term outcomes.

Declaration of conflicting interests

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