Primary cardiac tumors are rare. We report 9 patients with primary cardiac tumors (six benign, three malignant) over a 6-year period from 1985 to 1991. Six (54.5%) of these occurred in children less than 15 years of age. There were four myxomas (two right, two left atrial), one mesothelioma of atrioventricular node, and one rhabdomyoma. All the benign tumors were excised with the exception of the rhabdomyoma. Of the three primary malignant tumors (two schwannomas, one rhabdomyosarcoma), one underwent resection of a schwannoma and is alive and well, another patient with schwannoma had the tumor debulked and died from local recurrence, while the patient with rhabdomyosarcoma developed recurrence and died of systemic spread.

TUMORS OF THE HEART, despite the potential for more frequent recognition with the advent of echocardiography, are rare. The incidence of cardiac tumors ranges from 0.0017% to 0.28% in the general population.1,2 Primary tumors are much less common than metastatic tumors of the heart. Of the primary tumors over 75% are benign,3 of which the majority are made up of myxomas, rhabdomyomas, and fibromas. The common malignant tumors are rhabdomyosarcoma and angiosarcoma.2 We report herein our experience with primary cardiac tumors.

Materials and Methods

Over a 6-year period from 1985 to 1991, 9 patients with a diagnosis of a primary cardiac tumor, both benign and malignant, underwent surgical treatment. There were four myxomas, two benign non-myxomatous tumors, and three primary malignant tumors of the heart (Tables 1 and 2). The patients comprised of 3 males and 6 females, with the age ranging from 13 days to 54 years. Of these, there were 5 children under the age of 15 years.

Cardiac Myxomas

Four patients presented with solitary atrial myxomas, two in the left atrium and two in the right atrium. Both patients with left atrial myxoma presented with congestive cardiac failure, one in functional class II while the other presented with acute pulmonary edema. One patient with right atrial myxoma presented with abdominal pain and ascites, and the other with a history of hemoptysis and syncopal attacks.

The diagnosis was established by echocardiography. The tumors were excised successfully with neighboring tissue. One of the patients with left atrial myxoma had mitral valve regurgitation undiagnosed preoperatively and required a modified Kay commissuroplasty to restore competence of the mitral valve.

Non-myxomatous Benign Tumors

The first patient, a 13-day-old neonate, presented with cardiorespiratory failure requiring ventilation. On examination, she had hepatomegaly and was found to have a biventricular rhabdomyoma that was large and multifocal, involving the left ventricle more than the right. She was found to be inoperable at sternotomy, and following confirmation of her
Table 1. Patients with benign non-myxomatous tumors of the heart.

<table>
<thead>
<tr>
<th>Patient Age (Sex)</th>
<th>Site</th>
<th>Diagnosis</th>
<th>Presenting features</th>
<th>Surgery</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>13 d (F)</td>
<td>Right ventricle, left ventricle</td>
<td>Rhabdomyoma</td>
<td>Cardiorespiratory failure</td>
<td>Inoperable</td>
<td>1 mo</td>
</tr>
<tr>
<td>2</td>
<td>21 Y (F)</td>
<td>Atrioventricular Mesothelioma</td>
<td>node</td>
<td>Cardiac failure, atrial septal defect</td>
<td>Complete excision</td>
<td>25mo</td>
</tr>
</tbody>
</table>

Table 2. Patients with malignant tumors of the heart.

<table>
<thead>
<tr>
<th>Patient Age (Sex)</th>
<th>Site</th>
<th>Diagnosis</th>
<th>Presenting features</th>
<th>Surgery</th>
<th>Follow-up</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>41 y (F)</td>
<td>Left superior pulmonary vein</td>
<td>Rhabdomyoma</td>
<td>Syncope, left hemiparesis</td>
<td>Complete excision</td>
<td>6mo</td>
</tr>
<tr>
<td>2</td>
<td>2 Y (M)</td>
<td>Right atrioventricular groove</td>
<td>Schwanoma</td>
<td>Cough, cardiac failure</td>
<td>Complete excision</td>
<td>24mo</td>
</tr>
<tr>
<td>3</td>
<td>12 Y (F)</td>
<td>Right atrioventricular groove</td>
<td>Schwanoma</td>
<td>Cough, cardiac failure</td>
<td>Incomplete excision</td>
<td>2mo</td>
</tr>
</tbody>
</table>

diagnosis she was returned to the referring hospital.

The second, a 21-year-old female, presented with shortness of breath and was diagnosed as having an atrial septal defect. At surgery to correct this congenital anomaly, a 1-em tumor was found in the upper part of the triangle of Koch and was excised. Histology showed mesothelioma of the atrioventricular node. Postoperatively, but for transient atrioventricular nodal dissociation, she made normal recovery and remains well at followup of over two years.

Malignant Tumors

There were 3 patients with malignant tumors, of varied presentation. A 41-year-old female presented with syncopal attacks and left hemiparesis. Examination was normal, with the exception of a diastolic murmur at the apex. Echocardiography revealed a frond-like tumor involving all chambers of the heart. At surgery a rhabdomyoma was found, originating from the left superior pulmonary vein and extending into the left atrium and through the mitral valve into the ventricle. This tumor was unusual in that it extended through a patent foramen ovale into the right atrium and through the tricuspid valve into the right ventricle. The tumor was completely excised by decorticating the atria and reconstructing the left superior pulmonary vein and interatrial septum. She, however, developed recurrence of a tumor in the left ventricle and sustained an embolus in the left parieto-occipital region from which she succumbed 3 months after surgery.

The other two malignant tumors occurred in children. They presented with cough and shortness of breath. Both of them underwent surgery. The younger of the two, a 2-year-old child, had a thoracotomy for recurrent pericardial effusions at another hospital. At thoracotomy she was found to have a cardiac tumor and was then referred to us. This patient had a tumor involving the right atrioventricular groove and the free wall of the right atrium. This tumor was excised and resuspension of the tricuspid valve, along with pericardial reconstruction of the right atrium and a small part of the right ventricle, was carried out. There was no recurrence in the follow-up at two years. Histology showed a slow growing schwannoma of the vagus nerve sheath. The older child, a 13-year-old girl, had a much larger tumor arising from the right ventricle extending to the right atrium, superior and inferior vena cavae, and both great arteries. She underwent debulking of the tumor. It recurred with extensive local...
spread in 3 months and she died 6 months later. Histology again showed schwannoma of the vagus nerve sheath but with grade II malignancy.

Discussion

Myxomas usually form 75% of all primary cardiac tumors. The present series is unusual in that the myxomas formed only 36% of the tumors. The series is also unusual in that three of the nine tumors (33%) were malignant. The varied histology is also surprising. Constitutional symptoms, shortness of breath, syncope, and embolic episodes are the common manifestations of cardiac tumors. Two of the patients in the present series had syncopal attacks from obstruction of the atrioventricular valves. Two patients gave a history of embolic manifestations. One had a right myxoma who had a pulmonary embolism and the other a rhabdomyosarcoma of the left superior pulmonary vein extending into the left atrium with a right cerebral infarction.

Auscultatory signs are usually unhelpful. Five of the patients had soft murmurs, four of them systolic and one diastolic.

Echocardiography is the mainstay in the diagnosis of cardiac tumors and is the reason for greater recognition by the primary care centers who have gained access to this investigative modality. Seven of the nine tumors were detected by echocardiography and two were found incidentally at surgery. One patient was found at the referring hospital to have a tumor, at thoracotomy, for recurrent pericardial effusion. An atrioventricular nodal tumor was found as an incidental finding in a patient with an atrial septal defect.

Primary cardiac tumors are less frequent in children than in adults. However, in this series we found five of the nine tumors (54.5%) in children below the age of 15 years. In a 62-year review on childhood cardiac tumors, the Hospital for Sick Children in Toronto reported only 16 children with primary cardiac tumors; of these 15 were benign.5 In the series collected by the association of European pediatric cardiologists, there were 21 benign and no malignant tumors.6 The mortality rate for the pediatric age group, in our experience, has been two out of five. Of the 16 patients from the Toronto group, four underwent surgery; three of them are long-term survivors.

Treatment of primary cardiac tumors is complete wide excision. This is not always possible and thus partial excision,7 substitute myoplasty,8 or transplantation9 is sometimes carried out. In a patient with right ventricular fibroma where excision was not possible, the tumor was bypassed using a Fontan-type correction to obtain symptomatic relief.10

In our series, complete excision was possible in all myxomatous tumors. One benign nonmyxomatous tumor, a rhabdomyoma, in a neonate was considered inoperable because of the extent of myocardial involvement. Complete removal was not possible, and de bulking was achieved in a malignant schwannoma occurring in the atrioventricular groove.

Though there was no early mortality in our group, 3 patients died late (two had malignant tumors and one had an inoperable benign lesion). Death usually occurs within 6 months in malignant tumors.11

In a review of reports on a large series9,12,13 of cardiac tumors, it is rare to find any in association with conduction or nervous tissue. In a review of 133 cardiac tumors over a 25-year period, there were 13 Purkinje cell tumors.13 There were 12 mesotheliomas of the atrioventricular node in a review of 533 tumors and cysts of the heart.2 It is the smallest tumor of the heart and can cause sudden death in children.14 Reports of preoperative recognition and successful management are rare.15

Schwannomas have been described in other parts of the body but rarely in the heart.15 Their malignant potential is highly variable as evidenced by our experience with 2 patients. One had a recurrence in less than 2 months following incomplete resection. The other patient is well on follow-up at 2 years; histologically, it was a tumor of low-grade malignancy.

In conclusion, the prognosis in benign tumors is good if complete local excision is obtained. In malignant tumors the outcome is determined by the grade of malignancy, spread at presentation, and extent of resection. Overall, the prognosis remains poor.
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Acknowledgment

We are grateful to Alison Silkstone and Virginia Crosiar for their help in the preparation of this manuscript.

References