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Review

Cardiac tumors in children- A review

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Primary cardiac tumors 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. Primary cardiac tumors are much less common than metastatic tumors of the heart; and of the primary cardiac tumors, over 75% are benign, 3 of which are made up of myxomas, rhabdomyomas and fibromas. The most common primary sites are the lung, breast and cutaneous melanoma. The common malignant tumors are rhabdomyosarcoma and angiosarcoma.

Key words: Tumors, children, myxomas, rhabdomyomas, fibroma, lipoma.

INTRODUCTION

Cardiac tumors are benign or malignant neoplasms arising primarily in the inner lining, muscle layer, or the surrounding pericardium of the heart. Cardiac tumors can be primary or metastatic. Primary cardiac tumors are rare in paediatric practise with a prevalence of 0.0017% to 0.28% in autopsy series. In contrast, the incidence of cardiac tumors during fetal life has been reported to be approximately 0.14% (McAllister, 1979; Nadas and Ellison, 1968; Holley et al., 1995). The vast majority of primary cardiac tumors in children are benign, while approximately 10% are malignant. Secondary malignant tumors are 10-20 times more prevalent than primary malignant tumors (Lam et al., 1993).

Rhabdomyomas are the most common cardiac tumor during fetal life and childhood (Holley et al., 1995) (Table 1). This is usually followed by teratomas, fibroma and hemangioma (Holley et al., 1995). Cardiac tumors may present in fetal or post-natal life. The presenting features depend on the size and location of the mass. The manifestations of a cardiac tumor in fetal life include arrhythmia, congestive heart failure, hydrops and rarely stillbirth. In postnatal life, cardiac tumors may affect the integrity and function of the adjacent cardiac structures leading to severely compromised blood flow due to inflow or outflow tract obstruction, cyanosis, murmur, respiratory distress, myocardial dysfunction, valvular insufficiency, arrhythmias and sudden death (Nadas and Ellison, 1968; Groves et al., 1992).

The diagnosis of cardiac tumors can be established in symptomatic patients, but in rare cases sudden death is the presenting feature. Echocardiography or Magnetic Resonance Imaging is usually adequate to facilitate the diagnosis of cardiac tumors. Tumor biopsy, with histological assessment, remains as the gold standard for confirmation of the diagnosis.

Due to the progressive nature of pregnancy, fetal cardiac tumors are expected to grow antenatally and it is not unusual for cardiac lesions to be missed at an early obstetric scan. Some tumors can be detected from 20 weeks onwards but the majority will develop later in the course of the pregnancy. Most fetal cardiac tumors will be readily detectable in the late second or third trimester.[3, 5]

EPIDEMIOLOGY AND NOMENCLATURE

1. Primary cardiac tumors are rare
2. The most common primary cardiac tumor is the

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Table 1. Approximate incidence of benign tumors of the heart in children.

Incidence (%)	Children
Myxoma	15
Lipoma	-
Papillary fibroelastoma	-
Angioma	5
Fibroma	15
Haemangioma	5
Rhabdomyoma	45
teratoma	15

Source: Holley et al. (1995); Primary cardiac tumors. In: Goldhauber S, Braunwald E, eds. Atlas of heart diseases. Philadelphia: Current Medicine 1995.

rhabdomyoma followed by atrial myxomas
3. A quarter of primary cardiac tumors are malignant, the vast majority being sarcomas
4. Embolization, obstruction, and arrhthmogenesis are the chief modes of presentation
5. Sudden death is not common

General clinical features

Cardiac tumors are diverse in clinical presentation and atrial myxomas in particular may cause systemic symptoms mimicking collagen vascular disease, malignancy or infective endocarditis. There are several clinical features that are seen commonly with cardiac tumors:

1. Embolization: This occurs frequently. Either the tumor itself, or adherent thrombus may dislodge and migrate; multiple small emboli may mimic vasculitis or endocarditis, while larger fragments may lead to cerebrovascular events. Right sided tumors naturally embolise to the lungs producing pleuritic symptoms and possibly right heart failure.
2. Obstruction: Atrial tumors, once they are large enough, may result in obstruction of atriventricular valvar flow, and in particular, may mimic valvar stenosis. Symptoms are markedly paradoxical and may relate to body positions; frequent chest pain, breathlessness and syncope.
3. Arrhythmias: Intramyocardial and intracavity tumors may both affect cardiac rhythm, either through direct infiltration of the conduction tissue, or through irritation of the myocardium itself. The presence of serious ventricular arrhythmias should always lead to a search for structural heart disease and very infrequently a tumor may be found.

BENIGN TUMORS WITH MYOCYTE DIFFERENTIATION

Rhabdomyoma

Rhabdomyomas are benign tumors of the cardiac myocyte, which can be solitary or multiple. The cells typically contain large glycogen filled vacuoles. Rhabdomyomas are the most common tumors in the paediatric age group. They are also the tumors most commonly diagnosed during the prenatal period by foetal echocardiography; intrauterine as well as sudden death after birth has been attributed to these tumors. Rhabdomyomas may cause infant respiratory distress, congestive heart failure, or low cardiac output. The majority of children with cardiac rhabdomyomas also have tuberous sclerosis (Webb et al., 1993). Spontaneous tumor resolution is common and treatment is therefore usually conservative. Life threatening complications are unusual, but occasionally surgical resection is necessary (Black et al., 1998).

Histiocytoid cardiomyopathy

This is a rare but distinctive arrhythmogenic disorder caused by a neoplastic or hamartomatous proliferation of cardiac cells with some Purkinje cell characteristics. The synonyms of histiocytoid cardiomyopathy are *Purkinje cell hamartoma, infantile cardiomyopathy, oncocytic cardiomyopathy, isolated cardiac lipidosis, myocardial* or *conduction system hamartoma, foamy myocardial transformation* and *congenital cardiomyopathy*. Histiocytoid cardiomyopathy occurs predominantly in the first two years of life; 20% of cases are diagnosed in the first month, 60% in the first year and less than 3% after two years of life.

Histiocytoid cardiomyopathy is an arrhythmogenic disorder; over 70% of patients present with a spectrum of arrhythmias and electrical disturbances including: paroxysmal atrial tachycardia, atrial fibrillation, premature ventricular contractions, Wolff-Parkinson-White syndrome and right or left bundle branch block. Approximately 20% of patients present as sudden death and often such cases have been misclassified as Sudden Infant Death Syndrome (SIDS).

BENIGN TUMORS OF PLURIPOTENT MESENCHYME

Cardiac myxoma

Myxoma is a neoplasm composed of stellate to plump cytologically bland mesenchymal cells set in a myxoid stroma. Myxoma represents one of the most common benign cardiac tumors; are usually solitary and develop in the atria, 75% originating in the left atrium and 15-20% in the right atrium. They characteristically arise from or near the interatrial septum at the border of the fossa ovalis membrane (Bruce, 2007). Multiple acronyms for this condition are LAMB (*lentigines*, *atrial myxoma, mucocutaneous myxoma and blue naevi*) and NAME (*naevi, atrial myxoma, myxoid neurofibromata and ephilides*).

Recent nomenclature, however, suggests that they should be brought together under a broader cate-gory of Carney complex, named after the physician who first described the familial nature of this disorder (Carney et al., 1986).

Clinical manifestations are legion, both cardiac and systemic. Symptoms include breathlessness, fever, weight loss, syncope, haemoptysis and sudden death. Murmurs are frequently present, as is evidence of pulmonary hypertension, right sided cardiac failure and pulmonary embolization. Anemia, erythrocyte sedimentation rate and less frequently, the characteristic "tumor-plop" may be detected. This is heard as a loud but rather dull sound as the tumor prolapses into the left ventricle, and may be confused with a third heart sound. The method of choice for treatment is surgical resection on cardiopulmonary bypass.

Papillary fibroelastoma

This is an endocardial based papilloma lined by endothelial cells with proteoglycan rich avascular stroma, usually rich in elastin. This is a rare and benign tumor representing less than 10% of primary cardiac tumors and is the most common tumor of cardiac valves. Until recently, these tumors were considered to be benign and insignificant, but recent autopsy studies have demonstrated a high incidence of embolization; and the "*sea anemone*" appearance, with a short attaching pedicle, is typical. The clinical diagnosis of papillary fibroelastoma can be difficult because embolic complications can mimic a variety of underlying diseases.

HAEMANGIOMA

Haemangiomas are benign tumors composed predominantly of blood vessels. The histologic classification includes three types; the cavernous, capillary and arterio-venous haemangioma. Most cardiac haemangiomas are discovered incidentally but patients may present with dyspnoea on exertion, arrhythmias, right-sided heart failure, pericarditis, pericardial effusion and failure to thrive. This affects all age groups and accounts for 5% to 10% of all benign tumors. Haemangiomas undergo spontaneous regression with a good prognosis. However, their clinical course may be unfavourable in infants due to high-output cardiac failure, haemorrhage from ruptured vessels and thrombocytopenia.

BENIGN TUMORS WITH MYOFIBROBLASTIC DIFFERENTIATION

Cardiac fibroma

Cardiac fibroma is a rare primary heart tumor composed of fibroblasts or myofibroblasts with a matrix containing collagen. It almost exclusively occurs within the myocardium of the ventricles or ventricular septum. It is unclear whether it is a hamartoma or a true neoplasm. Because most cases occur in infants and children, it is likely to be congenital; with size varying from 1 to 10 cm. Cardiac fibroma may invade the ventricular muscle, replace the working myocardium and may result in intractable congestive heart failure or cyanosis; and rarely, a cardiac fibroma may extend into the ventricular conduction system causing ventricular arrhythmias (Isaacs, 2004; Becker, 2000; Marlin-Garcia et al., 1984).

Cardiac fibromas usually remain dormant and spontaneous regression rarely occurs, therefore total surgical resection is normally recommended. Large tumors can be resected subtotally and heart transplantation is done if there is progressive loss of working myocardial fibres (Burke et al., 1994; Geha et al., 1967).

Inflammatory myofibroblastic tumor

Inflammatory myofibroblastic tumor is composed of myofibroblasts accompanied by a variable number of inflammatory cells including lymphocytes, macrophages, plasma cells and eosinophils. The synonyms are plasma cell granuloma, inflammatory pseudotumor and possibly inflammatory fibrosarcoma. This tumor is characterized by fibroinflammatory and pseudomembranous appearance (Coffin et al., 1995; Demirkan et al., 2001); mostly occurring in children and young adults with a slight female gender predilection (male-to-female ratio being 3:4). Common site of occurrence is the lungs. Extrapulmonary sites have also been reported and they include the mesentery, genitourinary tract, gastrointestinal tract, retroperitoneum, pelvis, head and neck, trunk and extremities (Coffin et al., 1995; Ko et al., 2005).

CARDIAC LIPOMA

Cardiac lipomas are benign tumors composed of mature, white adipocytes. This occurs exclusively in adults; but when it occurs in children, accounts for less than 2% of heart tumors. Cardiac lipomas may occur anywhere in the heart but there is a predilection for the pericardium and epicardial surfaces. Other sites include the ventricular septum and cardiac valves. They are commonly silent but may rarely cause arrhythmias and atrioventricular block (Val-Bernal et al., 2000; Ashar and van Hoeven, 1992).

OTHER BENIGN TUMORS

Angioma

These tumors are extremely rare, occurring principally in the interventricular septum. They are visualised as

subendocardial nodules, having 2 to 4 cm diameter. Coronary angiography reveals a characteristic “tumor-blush”. Total surgical excision is not feasible due to the highly vascular nature of the tumor (Leonard, 2001).

Teratoma

Cardiac teratoma is a rare tumor of the heart and pericardium (Ali et al., 1994). Teratomas are the second most common tumor in the fetus and neonate after rhabdomyoma (Flyer, 1980; Isaacs, 1997; Isaacs, 1997). Most commonly, these tumors are detected in the pericardial activity attached to the pulmonary artery and aorta (Uzon et al., 1996). The tumor size within the heart varies from 2 to 9 cm in diameter and intrapericardial tumors as large as 15 cm have been reported (Carter et al., 1982; Roberts, 1997). Cardiac and pericardial teratomas are easily detected in the fetus and neonate by echocardiography as heterogeneous and encapsulated cystic masses. Treatment is by dissecting the teratoma from the great vessels.

MALIGNANT TUMORS

Primary malignant cardiac tumors

Up to a quarter of all cardiac tumors may exhibit some features of malignancy. 95% of these primary malignancies are sarcomas, 5% being lymphomas (Roberts, 1997). Sarcomas are more common in adults and most commonly located in the right atrium. The clinical course is usually aggressive with extensive local infiltration, intracavity obstruction and death.

Angiosarcoma

Angiosarcomas are the most common primary cardiac malignancy and more common in males. 80% of these tumors originate in the right atrium or pericardium (Roberts, 1997; Herrmann et al., 1992). Clinical picture includes right-sided heart failure, pericardial disease, pleuritic chest pain, dyspnoea and pericardial effusion. Some patients also present with fever, weight loss, and lassitude appear before signs of cardiac involvement (Leonard, 2001).

Rhabdomyosarcoma

Rhabdomyosarcomas are the second most common primary malignancy of the heart; originating from the striated muscle. These malignancies mostly occurs in adults, affecting the children rarely. Most common presenting symptoms are fever, anorexia, malaise and weight loss and the prognosis is usually poor (Leonard, 2001; Isaacs, 1997).

Fibrosarcoma

Fibrosarcomas are mesenchymal tumors with fibroblastic origin. These tumors can be seen within the left or right heart chambers (Leonard, 2001; Isaacs, 1997). The clinical findings include heart murmurs, chest pain, fever and malaise and the prognosis is poor (Leonard, 2001; Isaacs, 1997).

Lymphoma

Primary lymphomas involve only the heart and/or pericardium. They are rare and are sometimes seen as part of acquired immunodeficiency syndrome (Holladay et al., 1992) and in transplant recipients on immunosuppressive regimes. Lymphomas may go unrecognised as the chief presentation is with intractable heart failure, cardiomegaly or pericardial effusion (Holladay et al., 1992).

SECONDARY MALIGNANT (METASTATIC) CARDIAC TUMORS

Secondary cardiac tumors may be epicardial, myocardial or endocardial, but the vast majority are epicardial. Metastasis is rarely limited solely to the heart. The development of tachycardia, arrhythmias, cardiomegaly or heart failure in a patient with carcinoma should raise the suspicion of cardiac metastases. Rarely, cardiac involvement may be the first clinical feature of malignancy.

Macroscopically, carcinomatous metastatic tumors are multiple small, discrete and firm nodules. Carcinomas are more frequent than sarcomatous infiltrations.

Melanoma shows a special affinity to spread to heart with equal distribution to all four chambers (Gibbs et al., 1999). Leukemias and lymphomas may cause intramyocardial infiltration, haemorrhagic pericardial effusion, but occasionally they remain asymptomatic (Isaacs, 1997; Carter et al., 1982). The majority of metastatic tumors remain silent but some may present with arrhythmias, cardiac failure or pericardial effusion.

DIAGNOSTIC EVALUATION OF CARDIAC TUMORS

Diagnosis and differential diagnosis of cardiac tumors often presents a challenge for the physician. Cardiac tumors, either benign or malignant, are difficult to diagnose due to their rarity, variety and nonspecificity of the symptoms that they may cause. Patient’s history, clinical examination and blood tests rarely lead to an immediate diagnosis of the tumor; therefore, suspicion of this condition is critical for the correct and timely diagnosis of a cardiac tumor. Furthermore, beyond the

performance of imaging techniques, histological evaluation via biopsy is essential for the final diagnosis to be established.

MANAGEMENT

Therapy of benign malignant tumors is surgical resection and the urgency to intervene is determined by the symptoms of the patient and the type of the tumor.

1. Myxomas are indicated for immediate surgical resection regardless of symptoms, because of the high risk of embolic and cardiac complications.
2. Papillary fibroelastomas are surgically removed if the tumors are larger than 1 cm.
3. For small, immobile tumors in the left ventricles, conservative management and close follow-up is advocated.
4. Lipomas and lipomatous hypertrophy are surgically managed.
5. Rhabdomyomas usually do not require surgical management, since they tend to regress spontaneously.
6. Sarcomas are managed by surgical resection and chemotherapy is used as an adjuvant to help decrease tumor.

Finally, regarding cardiac manifestations due to metastatic extra cardiac cancer, priority is given to the management of the primary focus of the disease and the cardiovascular complications that are manifested (that is, percutaneous balloon pericardiotomy in cases of cardiac tamponade, radiotherapy and chemotherapy in cases of tumors that obstruct flow in the venae cavae).

CONCLUSION

Although cardiac tumors are rare, they are increasingly recognised ante mortem, permitting earlier diagnosis and treatment. The most likely etiology of a cardiac mass is a thrombus or vegetation. If a cardiac mass represents a tumor, its etiology can be determined by considering the histology based likelihood, the age of the patient at time of presentation, tumor location and tissue characterization by non-invasive imaging. Echocardiography is a vital non-invasive method of detecting and diagnosing cardiac tumors since it provides the precise anatomic location and hemodynamic impact of most cardiac tumors. In the current era, for benign cardiac tumors, an early diagnosis and appropriate treatment is not only possible, but often curative. Unfortunately the outcome for malignant primary tumors, even despite early diagnosis and aggressive treatment, remains dismal. Fortunately these tumors are exceedingly rare and seldom encountered in clinical practise.

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