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Other Cardiac Topics

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CARDIAC TUMORS

Primary cardiac tumors are extremely rare, with a prevalence of less than 0.3% in most pathologic series (Table 11-1). Myxoma, the most common primary tumor of the heart, is usually benign. Myxomas are frequently isolated lesions, arising most often in the left atrium in the region of the fossa ovalis. Less commonly, they may be detected in the right atrium, in the right or left ventricle, or in multiple sites within the heart. A familial pattern of myxomas can occur and is transmitted in an autosomal dominant manner. In these patients, multiple cardiac myxomas may be present in association with a constellation of extracardiac abnormalities including pigmented nevi, cutaneous myxomas, breast fibroadenomas, and pituitary and adrenal gland disease. In addition, patients with familial myxoma may have recurrence of the tumors after surgical excision. Whether sporadic or familial, fewer than 10% of myxomas are malignant.

Symptoms associated with myxoma are usually related to embolization of tumor fragments and obstruction of the mitral valve. Tumor involvement of the conduction system may manifest as sick sinus syndrome and dysrhythmias. In addition, patients may exhibit a constellation of nonspecific symptoms and laboratory abnormalities including fever, malaise, weight loss, anemia, and elevated erythrocyte sedimentation rate. The diagnosis is usually made with echocardiography; the transesophageal approach is the most sensitive method for detecting small left atrial tumors. Considering the propensity for embolization, most myxomas are surgically removed when diagnosed. Tumors may recur, so follow-up echocardiograms should be performed. Because of the low incidence right-sided tumors and subsequent low index of suspicion by physicians, these tumors are often misdiagnosed as thrombus on echocardiography. This leads to delayed diagnosis or inappropriate long-term commitment to anticoagulation therapy. Cardiac magnetic resonance imaging is often warranted to differentiate cardiac thrombus from a mass when the initial findings are not consistent with the clinical signs and symptoms (Fig. 11-1).

Other, less common benign tumors include papillary fibroelastomas, fibromas, and rhabdomyomas. Fibroelastomas are pedunculated tumors with frondlike attachments that usually arise from the surface of the mitral and aortic valve leaflets. These tumors do not result in valve dysfunction but may be a source of systemic embolization. Fibromas most often arise within the interventricular septum and may be associated with arrhythmias or conduction disturbances. Rhabdomyomas are the most common cardiac tumors found in children and are often associated with tuberous sclerosis.

Cardiac lipomas may occur throughout the heart and pericardium. Pericardial lipomas can be large, whereas intramyocardial lipomas are small and often encapsulated. Surgical excision is the treatment of choice. Lipomatous hypertrophy of the interatrial septum should be considered in the differential diagnosis of atrial masses. This lesion is a consequence of nonencapsulated adipose tissue hyperplasia. Although it is occasionally found incidentally at autopsy, it may be associated with supraventricular arrhythmias, conduction disturbances, and, in rare cases, sudden cardiac death.

About one fourth of all primary cardiac tumors are malignant, and most of these are sarcomas. These tumors grow rapidly and often result in chamber obliteration and obstruction of blood flow. If there is involvement of the pericardium, a hemorrhagic effusion with pericardial tamponade may develop. The prognosis in affected individuals is poor; surgical excision is possible in rare cases. Irradiation and chemotherapy may provide palliative relief.

In contrast to primary cardiac tumors, metastatic disease involving the heart is common, occurring in up to one in five patients dying with malignancy. The most common tumors to metastasize to the heart are carcinomas of the lung, breast, and kidney; melanoma and lymphoma may also have cardiac involvement. Metastasis to the pericardium is common and is often complicated by a hemorrhagic effusion and pericardial tamponade. Infiltration of the myocardium may result in conduction disturbances and arrhythmias. Intracavitary masses are unusual but may result from local tumor invasion or direct extension of the malignancy through the venous system (e.g., renal cell carcinoma may metastasize to the heart through the inferior vena cava). Treatment is directed at the underlying malignancy. If pericardial tamponade is present, immediate drainage will help stabilize the patient. A pericardotomy is often necessary to prevent reaccumulation of fluid within the pericardial sac. Surgical excision of an obstructing tumor mass is usually palliative.

TABLE 11-1 EXAMPLES OF TUMORS OF THE HEART AND PERICARDIUM

PRIMARY	Mesothelioma
Benign	Fibrosarcoma
Myxoma lipoma	**METASTATIC**
Papillary fibroelastoma	Melanoma
Rhabdomyoma	Lung
Fibroma	Breast
Malignant	Lymphoma
Angiosarcoma	Renal cell
Rhabdomyosarcoma	