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The cardiac tumors: Etiology, epidemiology and diagnosis

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Abstract
Primary cardiac tumors are a rare entity whose incidence is 0.3% to 0.7% of all cardiac tumors. The aim of this study was review of modern data of heart tumors epidemiology, etiology, pathogenesis and diagnostic procedures. Symptom presentation for cardiac tumors is quite varied, but it is dependent upon their location and size, rather than upon histological characteristics. The diagnosis of cardiac tumors relies heavily on the use of multiple imaging techniques. Surgical excision in combination with systemic chemotherapy are the best treatment for malignant cardiac tumors.

Keywords: Cardiac tumors, epidemiology, treatment

1. Introduction
According to surgery and autopsy reports, primary cardiac tumors are a rare entity whose incidence is 0.3% to 0.7% of all cardiac tumors [1]. Metastasis to the heart from other primary cancers is 30 times more common. Only 25% of primary cardiac tumors are malignant, and, of these, 75% are sarcomas. Malignant primary cardiac sarcomas are usually located in the right atrium and are most commonly angiosarcomas. In the left atrium, the most common malignant tumors are pleomorphic sarcoma (also known as malignant fibrous histiocytoma) and leiomyosarcoma [2].
Malignant primary cardiac tumors, which often strike a young patient population, have a dismal prognosis: without surgical resection, the survival rate at 9 to 12 months is only 10% [1]. In addition, it has been reported that patients with benign cardiac tumors are at increased risk of first ischemic stroke, particularly patients younger than 50 years [3]. Progress in imaging and cardiac surgery have considerably improved the prognosis in general population. However, cardiac sarcomas are still life-threatening diseases.

2. The aim of this study is review of modern data of heart tumors epidemiology, etiology, pathogenesis and diagnostic procedures.

3. Results and Discussion
Due to the low frequency, there is no specific grading scheme for cardiac tumors. This volume largely follows the principles of classification and grading detailed in the WHO Classification of Tumors of Soft Tissue and Bone (table 1) [4].

Table 1: Histological classification of heart tumors

Benign tumors and tumors-like lesions	Malignant tumors	Pericardial tumors
Rhabdomyoma	Angiosarcoma	Solitary fibrous tumors
Histiocytoid cardiomyopathy	Epithelioid hemangioendothelioma	Malignant mesothelioma
Hamartoma of mature cardiac myocytes	Malignant pleomorphic fibrous histiocytoma	Germ cell tumors
Adult cellular rhabdomyoma	Undifferentiated pleomorphic sarcoma	Metastatic pericardial tumors
Cardiac myxoma	Fibrosarcoma and myxoid fibrosarcoma	
Papillary fibroelastoma	Rhabdomyosarcoma	
Haemangioma	Leiomyosarcoma	
Cardiac fibroma	Synovial sarcoma	
Inflammatory myofibroblastic tumor	Liposarcoma	
Lipoma	Cardiac lymphomas	
Cystic tumor of atrio-ventricular node	Metastatic tumors	

The genetics of primary cardiac tumors is poorly understood. Although several complexes with genetic links (such as Carney complex) have been associated with benign primary cardiac myxomas, there are no demonstrable associations with malignant sarcomas [1]. There is, however, some small amount of information on sarcoma overall. Sarcomas generally reside in 2 groups: specific gene alterations with simple karyotypes or nonspecific gene alterations with complex karyotypes. Most simple alterations—such as rhabdomyosarcomas and Ewing, synovial, and gastrointestinal stromal tumors—are the result of chromosomal translocations. Pleomorphic sarcomas and leiomyosarcomas are probably the result of nonspecific translocations [5-7].

Symptom presentation for cardiac tumors is quite varied, but it is dependent upon their location and size, rather than upon histological characteristics. Large tumors may be relatively silent, whereas small tumors in a critical location may give rise to devastating clinic consequences. Presentation includes congestive heart failure from intracardiac obstruction, systemic embolization, constitutional symptoms, and arrhythmias.

Left atrial tumors, especially those that are mobile or pedunculated, may lead to

Systemic embolism involving the coronary, cerebral and peripheral circulations

Resulting in myocardial infarction, stroke etc. These tumors may also interfere with mitral valve function resulting in mitral stenosis or regurgitation. Cardiac murmurs and a characteristic tumor “plop” may be auscultated. Valve dysfunction manifests as left-sided heart failure with typical symptoms such as shortness of breath, orthopnea, paroxysmal nocturnal dyspnoea, pulmonary edema, fatigue, cough, and chest pain [8].

Intramural left ventricular tumors may be asymptomatic or present with a mass effect. With protrusion into the cavity, hemodynamic compromise may result. Local extension of the tumor may cause conduction or coronary artery compromise with chest pain, myocardial infarction, arrhythmia, heart block or sudden death [9].

Right atrial or right ventricular tumors may result in right heart failure from atrioventricular or pulmonary outflow obstruction, resulting in peripheral edema, hepatomegaly, ascites, shortness of breath, syncope and sometimes, sudden death. If the tumours interfere with valve function they may result in regurgitation or stenosis. Right-sided cardiac tumors may embolize to the lungs and present as pulmonary emboli with typical signs of pulmonary embolism. Chronic embolization may also mimic chronic thromboembolic disease with signs and symptoms of pulmonary hypertension [8].

Pericardial tumors may cause chest pain typical of pericarditis [10], sometimes they may be haemorrhagic and cause pericardial effusion and tamponade.

The diagnosis of cardiac tumors relies heavily on the use of multiple imaging techniques, including cardiac computed tomography, cardiovascular magnetic resonance, and echocardiography. Important imaging data to collect include information on the size of the intracardiac mass, the mobility of the mass (an important predictor of prognosis and embolic potential), myocardial invasion, and cardiac chamber location. These factors will provide the means to diagnosis and prognosis. Other important data to collect include the mechanism of tumor implantation, the relationship of the tumor with adjacent structures, the surgeon's route of access to the heart, left ventricular ejection fraction, and the dimensions of the affected chamber [1]. The modern diagnostic techniques such as positron emission tomography also can be used in heart tumors verification.

Surgical excision in combination with systemic chemotherapy are the best treatment for malignant cardiac tumors. The principal problem with surgical resection of primary cardiac tumors has been the tumor's extensive involvement of cardiac structures, which makes access difficult—the involvement of left-sided posterior structures especially impedes adequate resection [11, 12]. Orthotopic heart transplantation is an option if tumor resection and reconstruction would be expected to cause irreparable damage to essential cardiac structures [11].

The prognosis is excellent, including disease-free survival, if the tumor is completely resected. Incomplete resections predispose patients to further tumor growth and recurrence of symptoms; risk is undefined [13].

4. Conclusion

Despite primary cardiac tumors are a rare entity, the malignant ones have poor prognosis. The usage of multiple imaging techniques is the key method of early diagnosis. The aggressive and early treatment with early surgical intervention and systemic chemotherapy is the main position of their management.

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