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Malignant tumors of the heart

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ABSTRACT

Primary malignant cardiac tumors are rare, and mostly manifest as sarcomas in various types. As non-invasive diagnostic modalities, e.g. echocardiography and magnetic resonance imaging, have become more sensitive, there is a marked increase in the number of patients diagnosed. Nevertheless, most patients die within one year of initial diagnosis, either because of the often asymptomatic presentation of cardiac tumors until advanced disease, or a low index of suspicion on the part of the physician. The presenting symptoms, treatment options and, indeed, prognosis are largely controlled by the tumor's anatomic location. Cardiac sarcomas may present with a variety of symptoms and are known to be great mimickers. A quick diagnosis facilitates the initiation of a proper treatment (surgical resection, adjuvant chemotherapy), which may in turn improve the prognosis. Metastases to the heart are far more common, unfortunately, clinical manifestations are mainly dominated by generalized tumor spread. The article summarizes epidemiology, symptoms, diagnostic modalities, and possible treatment options.

1.

The diagnosis of cardiac tumors has been reported as early as the 16th century.[1] However, due to their rarity, diagnosis and treatment still poses a serious challenge for the treating physician. Primary cardiac tumors represent only a small fraction of all cardiac masses (Fig. 1). The most prevalent are pseudotumors (thrombi, vegetations, abscesses, foreign bodies, aneurysms of native coronary arteries or saphenous venous grafts) (Figs. 2 and 3).[2] The estimated frequency of primary tumors of the heart ranges from 0.0017% to 0.33% (Table 1).[3] Seventy-five percent of these are benign (Figs. 4 and 5). Myxomas account for nearly half of them (Fig. 4).[4,5] Primary malignant cardiac tumors are predominantly sarcomas in nature (Figs. 6–9). Angiosarcomas and undifferentiated sarcomas are the most prevalent.[3,4] Metastases to the heart are far more common (40 to 100 times) (Fig. 10).[4,6]

Extracardiac malignant tumors can spread to the heart by four paths: direct invasion (typically from the tumors of the mediastinum), hematogenous spread, lymphatic spread and intracavitary extension (from the inferior vena cava).[4] Metastastic cardiac tumors originate mainly from lung, breast and renal cancer, melanomas, lymphomas and leukemias (Figs. 11 and 12).[7] Melanoma is the neoplasm with the greatest propensity for cardiac metastases (50% to 70% of patients).[3,4] Although infrequent, since non-invasive diagnostic modalities have become more sensitive, there is a marked increase in the number of patients diagnosed with primary and metastatic cardiac neoplasms. Yet, in some instances (e.g. sarcoma, lymphoma) the final diagnosis of primary/secondary cardiac tumor cannot be made until autopsy.

1.1. Clinical presentation

Malignant tumors of the heart may present with a variety of symptoms and are known to be great mimickers.[4] Clinical presentation mainly depends on the size of the tumor and its anatomical location, rather than on its histological type. The symptoms can be grouped into five major categories:[3–5,8]

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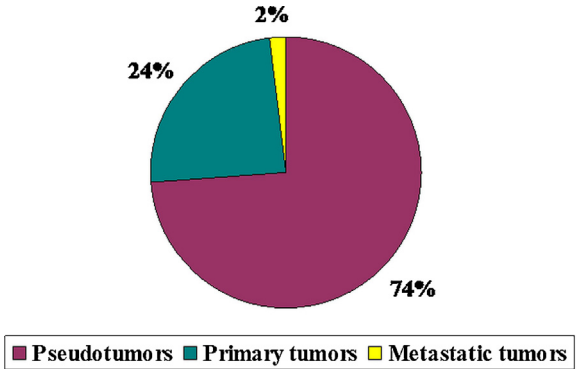

Fig. 1. Frequency of pseudotumors, primary, and secondary tumors of the heart.

Table 1
Incidence of primary and metastatic tumors of the heart.

Primary		Secondary/metastatic
Benign – 75%	Malignant – 25%	
Myxoma 45%	Sarcoma 20%	Lung cancer
Fibroelastoma 15%	Angiosarcoma	Brest cancer Melanoma
Lipoma 10%	Undifferentaited sarcoma	Leukemia/Lymphoma
Hemangioma 3%	Rhabdomyosarcoma	Sarcoma
Fibroma 2%	Leiomyosarcoma	Esophageal cancer
Other <1%	Fibrosarcoma	Ovarian cancer
	Liposarcoma	Kidney cancer
	Primary lymphoma 2%	
	Pericardial tumors 2%	
	Mesothelioma	
	Synovial sarcoma	

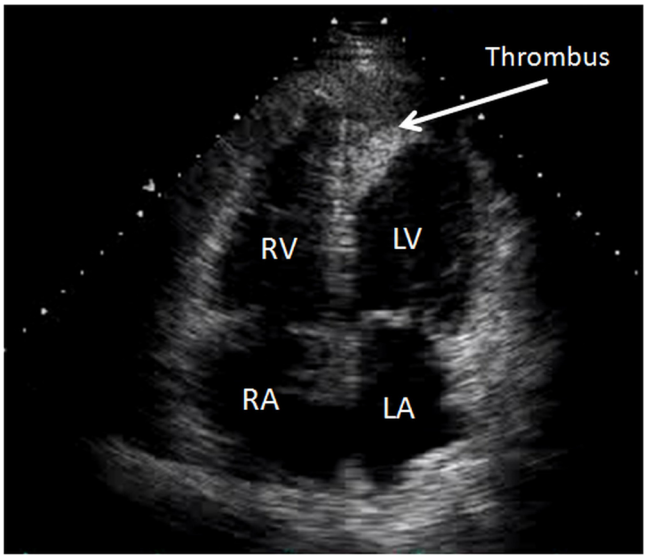

Fig. 2. Thrombus located in the apex of the left ventricle in a 64-year-old patient. Echocardiographic 4-chamber view (RA – right atrium, RV – right ventricle, LA – left atrium, LV – left ventricle).

- Constitutional or systemic symptoms: weight loss, malaise, fatigue, anemia, arthralgia, fever, polycythemia, leukocytosis, thrombocytosis.
- Obstructive cardiac symptoms: congestive heart failure, pulmonary edema, chest pain, dizziness, syncope, sudden cardiac death.
- Other cardiac symptoms: atrial/ventricular tachyarrhythmias, conduction abnormalities, pericardial effusion, cardiac tamponade, dyspnea, orthopnoe, cough, hemoptysis.
- Systemic embolization symptoms: stroke, transient ischemic attack, myocardial infarction, retinal artery embolism, embolism of the arteries of the lower or upper extremities, pulmonary embolism.
- Manifestations due to metastases – cardiac sarcomas most frequently metastasize to lungs, brain, and bones causing respective symptoms.

1.2. Diagnosis

Imaging techniques are used to define tumor size and location, to establish the degree of functional impairment and the relationship to other cardiac structures (the valves, the coronary arteries, the pericardium). Diagnostic modalities include echocardiography, computed tomography (CT), magnetic resonance imaging (MRI), positron emission tomography (PET), and cardiac catheterization with coronary angiography and/or endomyocardial biopsy.[4,5,8] Imaging features suggestive of cardiac malignancy include right atrial location, involvement of >1 cardiac chamber, size >5 cm, hemorrhagic pericardial effusion, broad base attachment and extension to the mediastinum or great vessels.[9]

- Echocardiography has the best spatial and temporal resolution. It can define the size, the shape, the mobility of the tumor, its location and relation to adjacent cardiac structures. It can provide information on the presence and the degree of blood flow obstruction or valve regurgitation. Suboptimal image

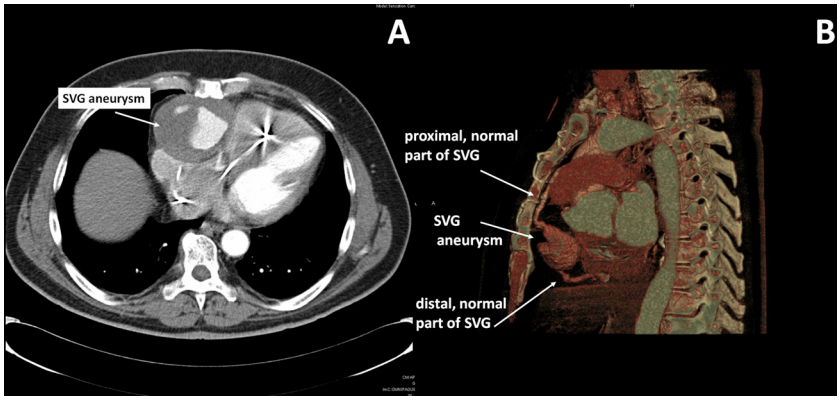

Fig. 3. Pseudoaneurysm of the saphenous vein graft to the right coronary angiography in a 55-year-old patient. (A) CT scan in transverse view. (B) CT 3D reconstruction (SVG – saphenous vein graft).

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