

# Cardiac Tumours and Malignancy Diseases

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Cardiovascular disease is commonly found in cancer patients. Due to a combination of an aging population and cardiotoxic cancer treatment, there is an expanding group of patients with cancer with co-morbid cardiovascular disease. In order to find a balance between the risk of undertreating the malignancy and that of damaging the cardiovascular system, it is vital to stratify cardiac risk at the time of cancer diagnosis.

The co-existence of heart disease and cancer in a patient often complicates treatment, because therapy for one disease may negatively affect the outcome of the other.

Extensive literature is available for the diagnosis and treatment of both heart disease and cancer [1–10]. Surprisingly, little data exists on the management of patients who are diagnosed with both illnesses.

Cardiovascular disease and cancer are frequently found in the same patient, due to both the high prevalence of both diseases, and because some forms of cardiovascular disease are caused by either cancer or its treatment.

In addition, guidelines for the treatment of cardiovascular disease are often based on studies which exclude patients who have cancer. Therefore, generally accepted strategies for the diagnosis and therapy of cardiovascular disease may not always apply to patients with cancer.

Cardiac tumours are a heterogeneous group of rare diseases. Furthermore, there is no clear classification or division of these pathologies. The Centre for Rare Cardiovascular Diseases (CRCD) has proposed a classification of cardiac tumours and cardiovascular diseases in malignancy.

A tumour is an overgrown tissue of unknown (undiagnosed) origin that may be detected clinically. A cardiac tumour is an additional structure within the heart chambers, myocardium or endocardium

A form of cardiac tumour known as a malignant rhabdomyoma was the first classified cardiac tumour, described by von Recklinghausen in 1862 [1]. Subsequent reports, mostly on isolated cases diagnosed in the autopsy, were collected and analysed by Mahaima in 1945 [2], followed by Prichard and Bigelow [3,4]. Initially, cardiac tumours were treated solely as a pathological curiosity. Along

with detailing the progress of diagnostic and treatment possibilities of congenital heart diseases, there was also a real chance to save patients suffering from tumours of the heart. The first successful surgical removal of cardiac tumour in an adult patient was performed in 1954 [5].

Cardiac tumours are very rare and histologically heterogeneous. These leads to a lack of established diagnostic schemes that would allow one to distinguish cardiac tumours from metastatic tumours, intracardiac thrombi, vegetations on the valves in infective endocarditis or in the process of connective tissue diseases, or thrombotic changes of the valves in the process of metastatic disease (also known as endocarditis marantica).

Cardiac tumours may be symptomatic or found incidentally during a physical evaluation for a seemingly unrelated problem or physical finding. In symptomatic patients, a mass can virtually always be detected by echocardiography, magnetic resonance imaging (MRI), and/or computed tomography (CT). Because symptoms may mimic other cardiac conditions, the clinical challenge is to consider the possibility of a cardiac tumour so that the appropriate diagnostic tests can be conducted.

There are very few studies assessing the results of cardiac surgery in individual centres. There is also lack of set of standards in the chemotherapeutic treatment of malignant cardiac tumours, although its routine use is sometimes discussed.

The prevalence of cardiac tumours is 0.021% (1 person in 5000). These data are based on a meta-analysis of pathological studies from the early twentieth century. It is likely that in more recent times, the incidence of cardiac tumours and primary tumours of the heart is greater.

Primary malignant cardiac tumours are rare, regardless of the age group. Based on the results of a large number of dissections, Straus and Merliss reported an incidence rate of 0.0017% [6].

Tumours of the heart occur in children slightly more frequently than in adults. Simcha reported that the incidence of cardiac tumours in children is 0.08% [7]. He based his conclusion on an over 20-year follow-up of the patients hospitalised in his facility. Nadas and Ellison provide the data that evaluated the prevalence among 11 000 children undergoing post-mortem examination at of 0.027% [8].

The pathophysiology of cardiac tumours is heterogeneous and depends on the type of tumour.

Heart tumours are divided into two groups:

- Primary heart tumours – deriving from the heart
- Secondary heart tumours – most commonly metastatic malignancies of other organs.

Primary heart tumours may be benign (about 75%) or malignant (25%).

In some cases a genetic background has been proven to be the cause of cardiac tumours. For example, 10% of cases of atrial myxomas are familial, as a part of the Carney complex (a mutation in the *PRKAR1A* gene on chromosome 17, encoding a regulatory subunit of protein kinase)

Taking into account the likelihood of cardiac tumours, the appropriate diagnostic procedures should be performed in all patients with unexplained cardiac murmurs, congestive heart failure, and/or arrhythmias which are accompanied by fever, anemia and weight loss of unknown cause. In such cases, the basic laboratory tests should include tumour markers).

Cardiologic diagnostic procedures should be widened, with particular emphasis on imaging methods.

An ECG may not indicate any change, and some common ECG features found in patients with cardiac tumours are usually non-specific. It has been reported that tumours of the heart are associated with shortening of the P-R interval, right or left bundle branch block (RBBB or LBBB), or complete heart block [8]. Recurrent tachycardia may also occur [9].

A standard chest radiography may not reveal any changes. An uncharacteristically enlarged heart contour following a tumour commonly appears in connection with heart failure. Asymmetrical enlargement of the heart may occur when the tumour is located within the anterolateral wall of left ventricle. Calcifications are rare, usually appearing in cases of rhabdomyomas and lipomas. Pericardial effusion is often the only radiological indicator of a cardiac tumour, as described in the case of rhabdomyoma associated heart failure [10,11].

Modern echocardiography often provides sufficient data about the presence of cardiac tumours, including location, size, mobility and communication with other anatomical structures of the heart. Echocardiography visualises both the myocardium and the cardiac chambers, and can usually identify the presence of a mass and its mobility. In addition, echocardiography may provide information about any circulatory obstruction, as well as the likelihood that the tumour could be a source of emboli. Fetal echocardiography allows for the early diagnosis of cardiac embryonic tumours [12]. While transthoracic echocardiography is simpler and can usually identify a tumour, transesophageal echocardiography (TEE) may be more informative. The superior diagnostic utility of TEE is due to the proximity of the esophagus to the heart, the lack of intervening lung and bone, and the ability to use high-frequency imaging transducers that afford superior spatial resolution [13].

Computed tomography and magnetic resonance are usually performed in cases where clarification is needed as to the degree of the tumour's infiltration into other structures of mediastinum [14]. Although both cardiac MRI [15,16] and ultrafast CT [14,17] provide noninvasive, high resolution images of the heart, MRI is gener-

ally preferred. In addition to furnishing detailed anatomic images, the T1- and T2-weighted sequences reflect the chemical microenvironment within a tumour, thereby offering clues as to the type of tumour that is present [18]. However, CT scanning is still useful when MRI is not immediately available or is contraindicated. An excellent pictorial review of many cardiac tumours and a comparison of MRI and CT scanning have been published by Hoey and colleagues [19].

Positron emission tomography (PET) has been useful in identifying cardiac involvement in patients with metastatic tumours, atrial myxomas, or lipomatous septal hypertrophy [20,21].

Coronary angiography is performed to assess the vascularity of the tumour and its infiltration of the coronary arteries [22,23]. This is of a particular importance in making the decision about surgery and the choice of technique.

Cardiac catheterization provides additional information about the hemodynamic consequences of the presence of the tumour of the heart [24].

With regards to transvenous biopsy, limited data are available on the risks and benefits involved in performing the procedure on suspected cardiac tumours. Due to the embolisation risk of myxomas, transvenous biopsy is not generally warranted if the appearance of the tumour is typical on noninvasive imaging. A biopsy is considered reasonable for other cardiac tumours if the potential benefits are deemed sufficient to outweigh potential risks.

## References

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