Cardiac tumours

What are cardiac tumours?

The heart may (rarely) be affected by primary or secondary tumours. Most cardiac tumours arise from or occupy the myocardial or pericardial tissues. Secondaries may occur by local extension or haematogenous spread. Myxomas account for around 50% of all cardiac tumours with the remainder made being a mixture of rare primary and secondary tumours. [1]

Cardiac tumours can present a significant diagnostic challenge causing symptoms and signs that mimic other cardiac diseases. Detection is now much easier with echocardiography and MRI scanning.

How common are cardiac tumours? (Epidemiology)

- Primary cardiac tumours are rare and account for 5-10% of all cardiac tumours. [2]
- Metastases to the heart from other primary cancers are much more common. [3]
- Cardiac myxoma is the most common cardiac tumour, accounting for 24-37% of primary cardiac tumours. [4]
- Only 25% of primary cardiac tumours are malignant and, of these, 75% are sarcomas. [3]

Presentation [5]

Clinical presentation can vary from a commonly known triad of symptoms, including obstructive, embolic, or systemic symptoms, to asymptomatic presentations. Symptoms may include:

- Congestive symptoms, such as orthopnoea, dyspnoea, and frank haemoptysis as a result of florid pulmonary oedema.
- Embolic phenomenon, which can lead to acute pulmonary embolisms, strokes, or other cerebrovascular events.
- Systemic symptoms, including fevers, arthralgia, and rigors.

Cardiac tumours may also present with arrhythmias or heart murmurs that may vary with body position.[6]

Cardiac myxoma[7]

- Cardiac myxomas are benign tumours. Approximately 75-80% of myxomas are located in the left atrium, 10-20% are located in the right atrium, and 5-10% are in both atria or either ventricles.
- They are typically solitary, pedunculated, and arising in the vicinity of the fossa ovalis. They may also be multicentric, sessile or attached to other areas of endocardium.
- They present with symptoms of haemodynamic obstruction, embolisation, or constitutional symptoms such as fever, malaise, tachycardia and tachypnoea.
- They may produce symptoms and signs of ischaemia or infarction in the peripheries, due to embolisation of adherent thrombus.
- Large myxomas may impair intracardiac blood flow, causing dyspnoea, syncope or symptoms and signs of congestive cardiac failure.
- Treatment is by prompt excision.

Rhabdomyoma[8]

- Rhabdomyomas are the most common primary cardiac tumour in infancy and childhood.
- Multiple tumours are associated with tuberous sclerosis in 60-80% of cases.
- Arrhythmias may occur.

- Rhabdomyoma cells lose their mitotic potential after birth and more than 80% disappear during early childhood.
- Surgery is only considered for serious, life-threatening symptoms, mainly due to obstruction of the cavity outflow/inflow or intractable arrhythmias.

Papillary fibroelastoma[9]

- Papillary fibroelastomas can occur at any age but are usually seen in older age groups.
- They are usually small pedunculated tumours connected to the mitral valve.
- They usually occur on the mitral (most commonly) or aortic valves.
- Often they are discovered after embolisation has occurred. They may also be asymptomatic and discovered during echocardiography.

Lipomatous septal hypertrophy

- Lipomatous hypertrophy of the interatrial septum is characterised by fatty deposits in the interatrial septum with sparing of the fossa ovalis. It is benign and most patients remain asymptomatic.[10]
- It may cause arrhythmias, particularly atrial dysrhythmia and heart block.

Teratomas and paragangliomas

- Teratomas are usually located in the pericardial cavity. The main clinical findings either relate to pericardial effusion or to the mass effect of the tumour, if intracardiac.
- Although generally considered benign, tumour recurrence and malignant differentiation have been reported and so surgical resection of teratomas is recommended.[8]
- They often cause external compression of the heart. This leads to symptoms and signs of pericardial tamponade such as hypotension, fatiguability, dyspnoea, elevated jugular venous pressure (JVP) and Kussmaul's sign (JVP rising during inspiration).

- They may also cause a superior vena cava syndrome.
- Paragangliomas may manifest due to systemic symptoms caused by tumour products leading to hypertension, headache, flushing and palpitations. See the separate article on Phaeochromocytoma.

Rare benign tumours

Fibromas, haemangiomas and lipomas

- Fibromas are the second most common cardiac tumours in infancy. They may cause obstruction to blood flow, valvular dysfunction, arrhythmias, syncope or sudden death. Surgery should be offered early because of the tendency for the tumour to grow fast and cause severe haemodynamic complications.[8]
- Haemangiomas are benign and undergo spontaneous regression, but may rarely have an unfavourable prognosis due to high-output cardiac failure, haemorrhage from ruptured vessels, thrombocytopenia, ventricular tachycardia and cardiac tamponade. Complete surgical excision has excellent prognosis when feasible.[8]
- Lipomas of the heart are often located in the left ventricle or right atrium. Signs and symptoms, if present, depend on the size and location of the tumour but may cause dyspnoea, conduction abnormalities or sudden death. Slow growth often delays recognition until the lipoma has already grown to a considerable size.[11]

Purkinje cell tumours[8]

- Purkinje cell tumours (also known as histiocytoid cardiomyopathy, oncocytic cardiomyopathy, focal lipid cardiomyopathy and idiopathic infantile cardiomyopathy) are rare arrhythmogenic disorders and are essentially hamartomatous proliferation of cardiac cells.
- The most common clinical presentation is with arrhythmias such as paroxysmal atrial tachycardia, atrial fibrillation, ventricular fibrillation, ventricular tachycardia and junctional tachycardia. They may lead to sudden unexpected death in infancy.

- They may be familial and may be associated with atrial septal defect, ventricular septal defect, hypoplastic left heart syndrome and anomalies of eye, skin and CNS.
- Electrophysiological mapping and surgical excision, direct vision cryo-ablation or catheter ablation may be indicated in patients with refractory arrhythmias.

Rare malignant tumours

They often affect the right side of the heart. These are usually of the sarcoma group. The most common of these rare tumours is the haemangiosarcoma which usually affects the right atrium. Tumours of the sarcoma group:

- The vast majority of cardiac sarcomas present in a similar way to cardiac myxomas with symptoms of dyspnoea, chest pain and fatigue.[2]
- Tumour embolisation from the left side of the heart may lead to stroke, fits, visceral or limb infarction and tumour metastasis at distant sites.
- Signs associated with these tumours include diminution of the heart sounds, pericardial friction rub, pericardial effusion, crackles in the lung fields, refractory dysrhythmias, heart block and cardiac failure.
- Extensively infiltrating tumours may cause superior vena cava syndrome and dysphonia due to recurrent laryngeal nerve palsy.

Secondary tumours

These are much more common than primary tumours. They usually affect the pericardium and are most often secondaries from lung cancer, breast cancer or lymphoma.

Examination

In cases of suspected cardiac tumour

Most patients with cardiac tumours will have no specific signs unless the tumour is large or produces derangement through secretion of bioactive tumour products.

- Observe the general appearance:
  - Ascertain whether there is any dyspnoea at rest, facial flushing, congestion or engorgement.
  - Note whether there is any finger clubbing.
- Check peripheral pulses, looking for evidence of dysrhythmia or tumour/thrombotic embolisation.
- Look carefully at the hands and feet and the nail beds to detect any previous embolic events.
- Check for evidence of peripheral oedema.
- Observe the JVP closely, looking for evidence of its elevation or paradoxical rise during inspiration (Kussmaul's sign).
- Carefully listen to the heart sounds:
  - Note loudness of heart sounds and any pericardial rub.
  - Myxomas, if large and prolapsing through the atrioventricular valve, can cause an added noise known as 'tumour plop' as the mass impacts against the endocardial wall at the beginning of ventricular systole.
  - Large tumours (particularly myxomas) obstructing the mitral valve may produce the murmur of mitral stenosis.
- Listen to the chest:
  - To check for added sounds or basal crackles as evidence of left ventricular failure.
- Examine the nervous system to detect any evidence of cerebral embolisation.
- Palpate the abdomen, seeking evidence of hepatic congestion.

Differential diagnosis

- Heart valve disease.

- Infective endocarditis.
- Ischaemic heart disease causing angina or other abnormalities.
- Restrictive cardiomyopathy.
- Cardiac tamponade.
- Lung cancer.
- Mesothelioma.
- Mediastinitis - for example, due to oesophageal rupture.
- Other mediastinal tumours or disease processes.
- Pericarditis.
- Primary cardiac dysrhythmia.

Diagnosing cardiac tumours (investigations)[5]

- ESR and other inflammatory markers may be elevated.
- CXR may show an abnormally large cardiac outline or evidence of atrial enlargement.
- ECG may show evidence of dysrhythmia, voltage reduction due to pericardial disease and nonspecific ST-segment and T-wave abnormalities.
- Pericardiocentesis may be used to obtain pericardial fluid for biochemical and cytological analysis.
- Radiological imaging plays a key role in the diagnosis of cardiac tumours. Echocardiography, cardiac CT, and cardiac MRI are key imaging modalities, with echocardiography being favoured given its non-invasive, accessible, and cost-effective nature.
- More advanced imaging techniques such as fluourodeoxyglucose-positron emission tomography (FDG-PET) may be utilised to look for metastatic involvement of cardiac tumours or identify a primary source in the case of cardiac metastasis..
- Cardiac catheterisation may be employed to analyse haemodynamic parameters.

- Endomyocardial biopsy may (rarely) be conducted. Most biopsy samples are obtained during surgical exploration and intervention.
- A specific test for paragangliomas is the metaiodobenzylguanidine (MIBG) scan. The radioactively labelled guanidine analogue is taken up by neuroendocrine cells and demonstrates the tumour presence and position.

Associated diseases

- Carney complex is a rare autosomal dominantly inherited multiple endocrine neoplasia syndrome that includes spotty skin pigmentation, recurrent cardiac myxomas, endocrine hyperactivity, pituitary adenomas, peripheral nerve tumours, and testicular tumours/ovarian lesions.[12]
- Tuberous sclerosis is strongly associated with rhabdomyomas in children. They may be diagnosed prenatally on ultrasound examination.[13] They may be the only presenting feature of tuberous sclerosis in some children who do not have other typical features, or in whom signs of the disease are not detected until the cardiac tumour raises the suspicion.

Management of cardiac tumours

- Medical therapies such as beta-blockers (particularly for paragangliomas), other antihypertensives and anti-arrhythmics may be used as initial stabilising therapy.
- Most intracardiac tumours are excised surgically. This therapy is curative for benign tumours, although they may recur later in life.
- When possible, surgical excision in combination with systemic chemotherapy remains the best treatment for malignant cardiac tumours.[3]
- Secondary cardiac tumours usually carry a poor prognosis and respond poorly to surgical or other curative attempts.
- Pericardiotomy or pericardial fenestration may help to relieve symptoms caused by pericardial disease.

- Cardiac transplantation has been used to treat malignant and some benign cardiac tumours. Long-term outcomes are unclear due to the small number of cases involved.[14]

Complications of cardiac tumours

- Left, right or combined ventricular failure.
- Cardiac valvular dysfunction.
- Heart block of varying degrees.
- Other cardiac dysrhythmias.
- Pericardial effusion and cardiac tamponade.
- Embolic events including stroke.
- Distant tumour metastasis.
- Superior vena cava obstruction.
- Systemic upset due to secretion of bioactive mediators by a tumour.
- Sudden cardiac death due to acute haemodynamic compromise in advanced cases.
- Tumour recurrence after resection.

Prognosis

- Benign tumours usually carry a good prognosis with normal life expectancy post-resection.
- Patients who have had benign tumours resected are usually followed up with regular echocardiography and cardiological supervision.
- Malignant tumours such as sarcomas tend to do very poorly despite intervention.
- Secondary malignancy affecting the heart has a grave outlook, although there is much that can be done to palliate the worst effects of the condition.

Prevention of cardiac tumours

- Patients with diseases known to be associated with cardiac tumours should be screened clinically and usually with echocardiography to detect the presence of intracardiac growths.
- Patients who have had benign tumours resected should usually be followed up with echocardiography and cardiological assessment to detect recurrence.

Further reading

- Careddu L, Oppido G, Petridis FD, et al; Primary cardiac tumours in the paediatric population. Multimed Man Cardiothorac Surg. 2013;2013:mmt013. doi: 10.1093/mmcts/mmt013.
- Tyebally S, Chen D, Bhattacharyya S, et al; Cardiac Tumors: JACC CardioOncology State-of-the-Art Review. JACC CardioOncol. 2020 Jun 16;2(2):293-311. doi: 10.1016/j.jaccao.2020.05.009. eCollection 2020 Jun.
- Poterucha TJ, Kochav J, O'Connor DS, et al; Cardiac Tumors: Clinical Presentation, Diagnosis, and Management. Curr Treat Options Oncol. 2019 Jun 27;20(8):66. doi: 10.1007/s11864-019-0662-1.

Authored by:	Peer Reviewed by: Dr Krishna Vakharia, MRCGP	
Originally Published: 20/11/2023	Next review date: 17/08/2023	Document ID: doc_1094

Patient Access

To find out more visit www.patientaccess.com
or download the app

Download on the App Store

GET IT ON Google Play

Follow us