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Intracardiac masses and anaesthesia – a case report

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Introduction

Cardiac disease can present with various pathologies, most of which pose an interesting anaesthetic challenge. The commonest cardiac diseases in developing countries are rheumatic heart disease, cardiomyopathies, and hypertensive heart disease.[1,2] However, occasionally anaesthetists are faced with rare cardiac disorders such as intracardiac masses.

Primary intracardiac masses can be grouped into tumours, thrombi or vegetations.[3] The tumours can be classified as primary or secondary, with either benign or malignant variations.[4] Table I shows the classification of the various intracardiac masses. This review will include only the masses that occur more frequently.

Malignant cardiac tumours are uncommon; they are reported as 25% of all cardiac tumours.[5] Primary cardiac malignancies are even rarer and mainly present as sarcomas. They are aggressive, spread rapidly and have a poor prognosis. These include angiosarcomas, which are vascular tumours found in the adult population, and rhabdomyosarcomas (resembling striated muscle) are seen in the paediatric population. Chemotherapy and radiotherapy have a minimal effect on them.[2,6] Secondary intracardiac masses are commonly due to metastases from lung, breast, liver, oesophagus lymphoma, and even melanoma. Metastatic lesions are 40 times more common than primary tumours. The incidence of cardiac metastases, found during autopsy, was between 2–18%.[7,8] The mediastinal tumours spread to the heart via direct invasion, thus affecting the pericardium. The lymphatics allow metastatic material to invade the epicardium and myocardium. Haematogenous spread results in lesions in the endocardium.[8]

The remaining 75% of all cardiac tumours tend to be benign. The commonest benign cardiac tumour is the myxoma.[5,9-11]

Table I: Intracardiac masses

Intracardiac tumours	
Primary	**Secondary**
Benign • Myxomas • Vegetations • Thrombi • Lipomas • Papillary fibroelastomas • Teratomas • Haemangiomas • Hamartomas	
Malignant Sarcomas • Angiosarcoma • Rhabdomyosarcoma	**Metastases from** • Lung cancer ◦ Pleural mesothelioma ◦ Adenocarcinoma ◦ Squamous cell carcinoma • Breast cancer • Melanoma • Squamous cell carcinoma of the oesophagus • Carcinoid disease • Hepatocellular carcinoma • Renal cell carcinoma • Uterine leiomyosarcoma • Lymphoma

Myxomas

These slow-growing, soft, gelatinous (myxoid) endocardial growths tend to resemble organised thrombi.[4] They develop as a pedunculated mass forming along the fossa ovalis and then extend into atria. Their average diameter is 5–6 cm, but tumours with diameters of almost 15 cm have been reported.[10] Seventy-five per cent of atrial myxomas are found in the left atrium, 20% are in the right atrium, and some become bi-atrial when there is an intracardiac shunt.[11-13] The ventricles are affected in only 3–5% of cases.[14] Myxomas occur between the ages of 30–60 years and show a higher incidence in females than males, by more than 75%.[4]

The patients are generally asymptomatic until the valve orifice is obliterated and impedance of blood flow develops. The clinical symptoms thus depend on the mobility, size and position of the tumours. The classic triad of features includes ***embolism, intracardiac obstruction***, and ***constitutional symptoms***.[11] The intracardiac obstruction may mimic mitral or tricuspid stenosis. These patients, therefore, may present with dyspnoea on exertion (especially due to mitral valve obstruction caused by left atrial myxomas), orthopnoea, oedema, cyanosis, hypotension, and syncope from intermittent obstruction of blood flow, so may also be associated with sudden death. The systemic and pulmonary embolic phenomenon, caused by thrombi or tumour

fragments, often present as pulmonary hypertension, retinal or coronary artery thromboses. The constitutional symptoms include fever, arthralgia, fatigue, myalgia, anaemia and weight loss. These manifestations are due to the release of inflammatory mediators from the tumour.[10,11,15] Myxomas, as with any other intracardiac masses, may be associated with haemolytic anaemia and thrombocytopenia.[11,14]

On clinical examination, in addition to various systolic and diastolic murmurs, a characteristic "*tumour plop*" occurs after S_2. Wide splitting of S_1 from delayed closure of valves is evident on auscultation. A loud P_2 from pulmonary hypertension may also be heard. The right-sided tumours can manifest similarly to cyanotic congenital cardiac lesions. Right atrial enlargement with tricuspid regurgitation, arrhythmias, atrial fibrillation and tricuspid obstruction can also occur. A pericardial friction rub is also linked to right-sided masses. All these features depend on the tumour size, number, shape, location and mobility.[11,16]

The advancement of imaging techniques has resulted in the diagnosis of these masses being made more frequently.[11] However, these patients are often treated for pneumonia and ischaemic heart disease prior to the actual diagnosis being established.

The investigations that are required for a diagnosis include chest radiographs which may show calcification within the atrial myxoma (but only in a third of patients) with prominent pulmonary vasculature and congestion. Electrocardiography (ECG) for detection of arrhythmias and ventricular hypertrophy from the chronic intermittent obstruction. Echocardiography (TTE and TOE) is the cornerstone of the diagnosis, as well as for intraoperative and postoperative evaluations. It allows for confirmation of tumour size, position, shape, presence of intracardiac shunts as well as tumour movement. Three-dimensional TOE has also become indispensable. Computed tomography (CT) and magnetic resonance imaging (MRI) are required for more invasive tumours, atypical tumours, or inconclusive echocardiography. Angiography has lost popularity due to the increased risk of embolisation from the catheters, but is utilised when there is concern regarding coronary patency or ischaemic heart disease.[7,11]

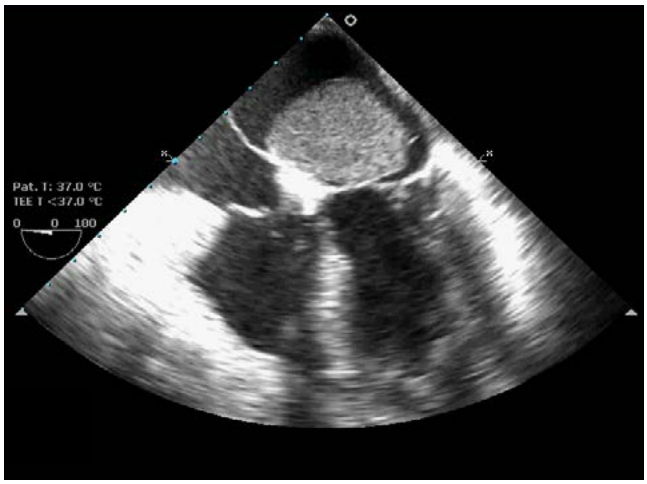

Figure 1: Trans-oesophageal image of left atrial myxoma.
Courtesy Charlotte Maxeke Johannesburg Academic Hospital

The treatment options are surgical resection, chemotherapy, radiotherapy, or a heart transplant. Surgical correction in the benign tumours is curative. In malignant tumours, surgical resection is mainly for palliation.[16]

Surgical resection has a low early mortality rate of 0.5–2% and a late mortality rate of 6.1–7%. Despite the potential of tumour recurrence being 3–13%, patients have an excellent post-surgery survival rate.[17-19] Tumour resection is performed under cardiopulmonary bypass and is easier if the tumour is pedunculated. The risk of embolisation is significantly increased by fragmentation, so ideally it must be removed as one whole mass.[20] When there is more than one mass present or if there is rapid tumour recurrence, the possibility of Carney syndrome should be considered.[21]

Carney syndrome is a rare autosomal dominant familial cluster of cardiac and mucocutaneous myxomas, endocrine hyperfunction and cutaneous hyperpigmentation. It is due to heterogenous mutations of a specific tumour suppressor gene.[9] It is characterised by atypical locations and high rate of tumour recurrence. It presents early in life and peaks by the age of 30. It is also associated with multiple endocrine neoplasia, thus these patients pose a significant anaesthetic challenge.[21]

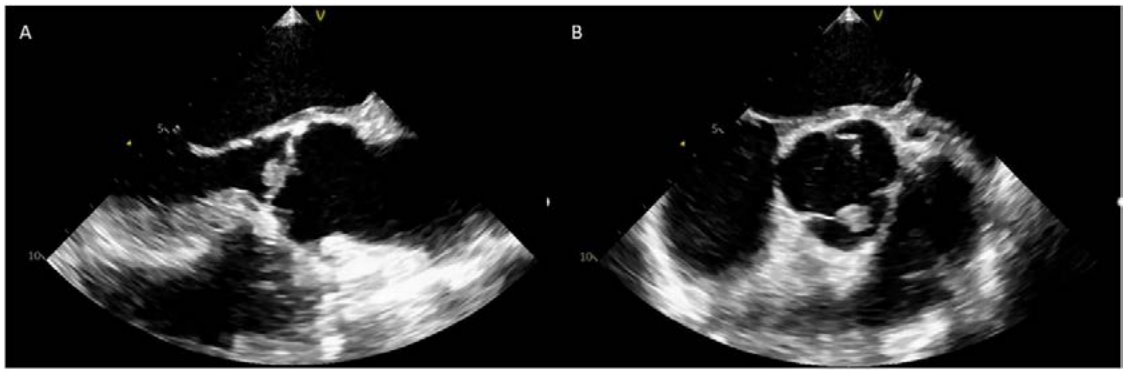

Figure 2: Trans-oesophageal echocardiographic (TOE) image of papillary fibroelastoma on the aortic valve in long axis (A) and short axis (B)
Courtesy Chahin et al.[22]

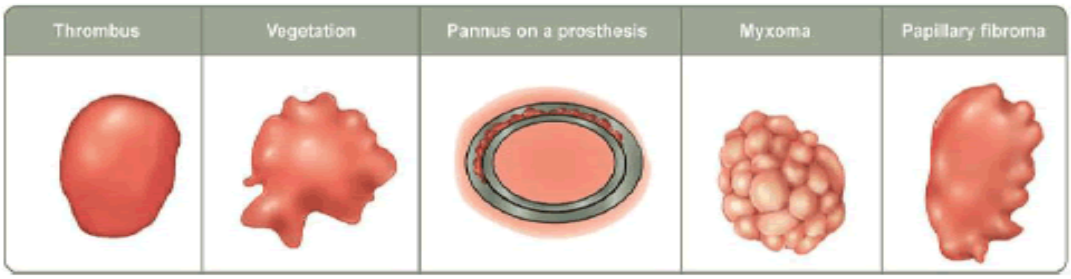

Figure 3: Diagrammatic representation of common cardiac masses
Courtesy of Adam[23]

Papillary fibroelastomas

These rare, benign, slow-growing tumours, also known as *endocardial papillomas,* are the second most common primary cardiac tumour. They affect the cardiac valves of patients around 60 years of age, with the highest prevalence in octogenarians. They also have a higher rate in males than females. Precise risk factors have yet to be determined. These papillary fibroelastomas have a characteristic flower-like appearance because of the short stalks with projections emanating from them. They are small 1–2 cm diameter, white, gelatinous avascular lesions that can affect any of the cardiac valves. Although they develop on the aortic and mitral valves, they rarely cause valve incompetence. The patients can present with angina pectoris, transient ischaemic attacks or sudden death, if the tumours extend from the aortic valve to the coronary ostia. They can be completely resected and have an excellent prognosis, unless there is a large associated thrombus.[9,10,24]

Thrombi

Thrombi and vegetations are the non-tumourous intracardiac masses that will be covered in this article. The distinguishing feature between thrombi and cardiac tumours is the ventricular wall motion abnormalities that contribute to the components of Virchow's triad. These haematogenous collections develop in either the atria or the ventricles. Thrombi found in the right atrium are suggestive of a deep vein thrombotic origin, a previous right-heart myocardial infarction or due to therapeutic foreign materials. Left atrial thrombi are the most common and are associated with atrial fibrillation, atrial flutter, atrial dilatation, ischaemic heart disease or prosthetic valves. They are usually firmly attached to the posterior wall of the atrium or in the atrial appendage. Thrombi have an irregular, lobulated structure with no pedicle, unlike cardiac tumours.[3,11,24]

Ventricular thrombi are unlikely in the presence of normal ventricular function. Myocardial ischaemia leads to ventricular dysfunction. This ventricular hypokinesia, or even akinesia, may also be associated with aneurysms or cardiomyopathies. Inadequate anticoagulation in patients with mechanical prosthetic valves contributes to thromboembolism.[24]

Pulmonary thrombi may occur secondary to intracardiac masses. Fragments of a mass in the right atrium can traverse through the heart and deposit in the pulmonary vessels. The course may be insidious and result in pulmonary hypertension. Severe symptoms and sudden death are associated with massive pulmonary emboli.[25]

Vegetations

Endocarditis predisposes patients to the development of vegetations on either prosthetic or native valves. The causes may be infectious or non-infectious. Infective endocarditis is a serious life-threatening bacterial or fungal infection that presents with valvular or perivalvular vegetations, abscesses and septic emboli. It requires immediate medical intervention or surgery in severe cases. Endocardial vegetations are differentiated from thrombi by constitutional features like a fever, myalgia, eye and skin manifestations, which are part of the Modified Duke diagnostic criteria.[27] Autoimmune disorders, malignancy, pregnancy and severe burns are risk factors for non-infectious vegetations. The lesions are small, sterile and composed of platelets and fibrin.[26,27]

Unlike thrombi, vegetations are associated with valve incompetence. The vegetations develop on the edge of the leaflet and prevent normal coaptation. Differential diagnoses for vegetations include acute rheumatic fever, systemic lupus erythematosus and primary cardiac tumours.[26,27]

Carcinoid heart disease

Carcinoid tumours can develop in the gastrointestinal, bronchopulmonary, and genitourinary systems. They are benign (sometimes become malignant) neuroendocrine tumours that release vasoactive hormones such as serotonin, bradykinin, histamine, kallikrein and prostaglandins. Carcinoid syndrome refers to a complex of clinical features that include flushing, hypotension, diarrhoea, bronchospasm, dyspnoea and wheezing from the surge of these vasoactive substances.[28,29]

Within approximately 18 months after its onset, the capacity of the liver to degrade these substances is overwhelmed, at which point it starts to affect the heart and carcinoid heart disease develops. The excess serotonin causes deposition of carcinoid plaques composed of myofibroblasts, collagen and myxoid matrix along the tricuspid and pulmonary leaflets. This leads to fibrosis and thickening of the tricuspid and pulmonary valves. It is typically characterised by tricuspid regurgitation, mixed pulmonary stenosis and regurgitation with associated right heart failure and myocardial infarction. The left heart remains mostly unaffected because of inactivation of serotonin by the lungs. However, the bronchopulmonary carcinoid tumours may

occasionally metastasise directly to the heart and eventually affect the left side.[28,29]

Carcinoid heart disease thus affects the valves, the myocardium, the cardiac conduction system, or even the coronary vessels. In addition to the previously mentioned investigations, positron emission tomography with radionuclide tracers allows for confirmation of carcinoid cardiac metastases.[30]

The management involves somatostatin analogues (Octreotide) which improves symptoms but not survival. Carcinoid heart disease has considerable mortality, with a life expectancy of 2–5 years and a median survival of 11 months in patients with severe carcinoid heart disease. The definitive treatment, in addition to medical therapy, is valvular repair or replacement.[4,29,30]

Anaesthetic considerations in a patient for resection of an intracardiac mass

The resection of intracardiac masses requires a multidisciplinary approach. There are various issues that the entire team should remain cognisant of.

These considerations include current symptoms of heart failure, thromboembolism and haemodynamic stability. A large mobile mass in the right or left atrium can cause an acute obstruction of blood flow and lead to severe haemodynamic instability with acute decompensation, at any stage of positioning, anaesthesia or surgery. The drug therapy, especially in the context of carcinoid syndrome or heart failure is also an important preoperative consideration. Heart failure can be precipitated by reductions in myocardial perfusion pressure resulting in myocardial depression and arrhythmias. Arrhythmias are poorly tolerated, especially during intermittent obstruction of the valve orifice and worsened by the development of valvular regurgitation or stenosis. In the face of severe pulmonary hypertension, a pulmonary thrombo-endarterectomy may be the recommended surgical option, which requires deep hypothermic arrest.[4]

Tumour manipulation (during placement of invasive lines or surgery) can result in fragmentation and therefore emboli, with strokes, myocardial ischaemia or renal dysfunction. Hence intraoperative TOE is essential for evaluating the risk of embolisation during cannulation, assessing the valvular involvement, planning for surgical approach and for checking for iatrogenic injuries which may lead to intracardiac shunts.[4]

Postoperatively the expected complications include atrial fibrillation, atrioventricular blocks, low cardiac output syndrome, coagulopathies with intracerebral bleeds and acute cholangitis.[17]

Case report

A 22-year-old female who presented to Charlotte Maxeke Johannesburg Academic Hospital, with a history of progressively worsening dyspnoea, pedal oedema and bilateral leg pain over three to four months. She had also suffered a miscarriage six days prior and had a raised body mass index. There were no other identifiable cardiovascular risk factors. She was admitted by the vascular surgeons, who embarked on comprehensive investigations to identify the cause of her limb ischaemia, with the aim of booking her for aorto-iliac embolectomy and possible amputation.

She subsequently developed dyspnoea at rest and severe respiratory distress, during her investigations. She was admitted to the critical cardiac care unit and placed on supplemental oxygen. Her haematological investigations were all within normal limits. She was also screened for disorders of coagulation, infective endocarditis, antiphospholipid syndrome and autoimmune disorders, which were all negative.

Chest radiograph showed bilateral diffuse opacities. ECG revealed a sinus tachycardia with right axis deviation. TTE revealed large mobile masses within both atria, with raised pulmonary pressures of 110 mmHg, due to suspected pulmonary emboli as well as a possible patent foramen ovale. CT of the pulmonary vessels detected bilateral pulmonary emboli along her left and right main pulmonary arteries. CT aortogram showed bilateral common iliac thrombi. Although she had critical limb ischaemia, it was decided that she needed to undergo an emergency pulmonary endarterectomy and excision of the bi-atrial masses (suspected to be myxomas) prior to the aorto-iliac bypass.

Her preoperative condition indicated an extremely guarded prognosis. She was in respiratory distress, diaphoretic and showing signs of congestive cardiac failure, NYHA IV. Her vitals were: BP 109/60 PR 120/min, with inotropic support, RR34 breaths/min, room air saturation 88%, PaO_2 61 mmHg and Hb 11.4. She had absent lower limb pulses, from her femoral arteries distally, a loud palpable P_2 with a parasternal heave and a pansystolic murmur. She also had reduced breath sounds bilaterally.

Upon arrival in theatre, invasive lines were inserted prior to induction of anaesthesia. This was done with the patient in a semi-fowlers position due to her severe distress. The induction consisted of a gentle titration of high-dose Fentanyl, which she did not tolerate very well. She suffered haemodynamic collapse which required chest compressions and significant inotropic support. Spontaneous circulation was re-established, and the surgeons then proceeded to quickly perform a sternotomy and "crash" onto bypass. They had opted against inserting peripheral femoral cannulae due to the presence of aorto-iliac disease and the lack of pulses.

Intraoperatively, a pulmonary endarterectomy was performed and she was found to have large clots in the right and left pulmonary arteries. Thereafter her right atrium was opened and a large myxomatous mass was excised from the right atrium and another one from the left atrium. TOE confirmed a patent foramen ovale. Due to her tumultuous intraoperative course and postoperative requirement of ECMO, her aorto-iliac bypass was postponed. She eventually had bilateral above-knee amputations. She underwent several other surgeries thereafter. She was eventually discharged from ICU six months later.

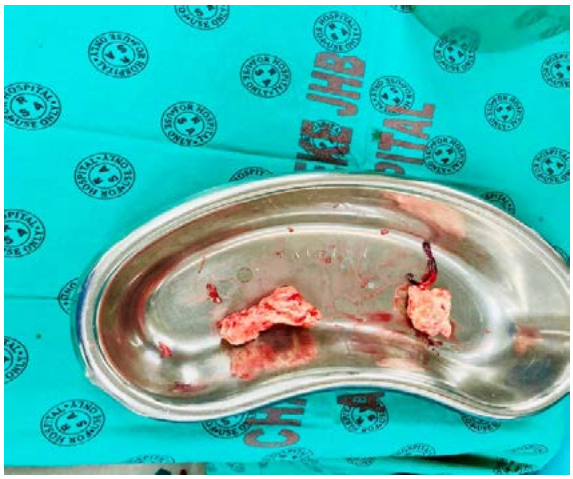

Figure 4: Image of the right atrial myxoma (right) and left atrial myxoma (left) removed from the patient
Courtesy Charlotte Maxeke Johannesburg Academic Hospital

Figure 5: Image of pulmonary thrombus fragments removed from left and right main pulmonary arteries
Courtesy Charlotte Maxeke Johannesburg Academic Hospital

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