Case report

Left atrial myxoma: an unusual cause of postpartum tachycardia and shortness of breath

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SUMMARY

Primary cardiac tumours are exceedingly rare in clinical practice and a myxoma is certainly not the first suspected diagnosis on a list of differentials in a patient presenting with breathlessness and tachycardia. Naturally, a structured work-up to rule out much more common conditions will take place. Nonetheless, an index of clinical suspicion is required especially when a clear-cut diagnosis is not forthcoming. We describe a case of a postpartum woman who presented with shortness of breath and palpitations, and was eventually diagnosed with a left atrial tumour. She underwent a successful surgical excision of the mass which was found to be a myxoma on histological examination. We review and present an up-to-date literature review on epidemiology, pathophysiology, clinical features, diagnosis and treatment options for cardiac myxomas.

BACKGROUND

Primary cardiac tumours are rare with an incidence ranging from 0.0017% to 0.19% in general population.¹ In adults, nearly three quarter of these tumours are benign; of which more than half are myxomas, followed by lipomas, fibromas, haemangiomas, neurofibromas and lymphangiomas.² The literature on the incidence and characterisation of cardiac tumours during pregnancy and postpartum period is very sparse, mainly limited to small case series and case reports.

CASE PRESENTATION

A 39-year-old woman presented to acute medical unit with shortness of breath, palpitations and tachycardia on exertion, 5 weeks post partum. She had given birth to a full term healthy baby via normal

vaginal delivery. Her medical history included sero-negative rheumatoid arthritis for which she was on no regular treatment. She had been given prophylactic low molecular weight heparin throughout the pregnancy and postpartum period. She was a life-long non-smoker. Her physical examination revealed tachycardia at rest with a heart rate of 110 beats per minute, with normal blood pressure, normal respiratory rate and oxygen saturations. Heart sounds were normal and chest was clear to auscultation. There was no clinical evidence of a deep vein thrombosis.

INVESTIGATIONS

Preliminary investigations showed a mild normocytic anaemia with haemoglobin of 111 g/L; renal function tests, cardiac troponin T and inflammatory markers were all normal. D-dimer was elevated 1.30 µg/L (normal range <0.5 µg/L). Arterial blood gas showed mild hypoxia with a PO₂ 10.5 kPa with a normal Pco₂ and pH. ECG showed sinus tachycardia with a rate of 110/min, with no ischaemic changes and no right ventricular strain. Chest X-ray revealed no abnormality.

DIFFERENTIAL DIAGNOSIS

Due to the symptoms of breathlessness and tachycardia during postpartum period and an elevated D-dimer, she was initially suspected to have pulmonary embolism. Treatment was started with subcutaneous low molecular weight heparin at a therapeutic dose. However, subsequently a ventilation-perfusion scan showed normal perfusion in both lungs, ruling out pulmonary embolism.

As her symptoms persisted, a transthoracic echocardiogram was performed mainly to rule out postpartum cardiomyopathy. This showed a 3.5 cm diameter ovoid mass in the left atrium attached to the inter-atrial septum that appeared to impinge on the mitral valve (figure 1). Colour Doppler showed a mild increase in the velocity of blood flow across the mitral valve possibly due to intermittent obstruction caused by the mobile mass. Estimated pulmonary artery pressure was within normal limits. Transoesophageal echocardiography confirmed this mass to be a left atrial myxoma (figure 2).

TREATMENT

She was urgently transferred to the regional cardiothoracic centre for surgical management. Left atrial tumour was surgically resected in full using a median sternotomy approach under general anaesthesia and

Figure 1 Transthoracic echocardiogram apical four chamber view showing a mass in the left atrium (arrow).

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Figure 2 Transoesophageal thoracic echocardiogram image providing a detailed view of the left atrial mass along with its attachment to the inter-atrial septum (encircled).

cardioplegic cardiac arrest. Histological examination revealed polygonal cells with scanty eosinophilic cytoplasm scattered in a myxomatous stroma confirming the diagnosis of myxoma (figure 3). The patient made a quick and complete recovery without any postoperative complications.

OUTCOME AND FOLLOW-UP

The patient was reviewed in cardiology clinic a few weeks after her surgery for a follow-up; reassuringly, excision of the atrial myxoma led to a complete resolution of her symptoms.

DISCUSSION

Although cardiac myxomas can occur at any age, the incidence is relatively high between the third and sixth decade of life. Pregnancy is not known to influence the incidence and progression of myxomas. Most of these tumours are sporadic, nonetheless familial cases have been reported in the literature which tend to affect younger individuals and are often associated with systemic involvement, for example, pituitary adenomas, adrenal dysplasia, breast fibroadenomas and skin myxomas.³

Over three quarter of cardiac myxomas are found in the left atrium where most of them arise from the inter-atrial septum. The rest of them are found in the right atrium, left ventricle and the right ventricle, in a descending order of frequency.⁴ Multifocal myxomas and bi-atrial myxomas occur very rarely and usually have a familial origin. Carney complex is a rare autosomal dominant disorder comprising pigmented skin lesions, cardiac myxomas and endocrine tumours.⁵ Relatively little is

Figure 3 Microscopic view of the excised tumour shows scattered polygonal cells in an acid-mucopolysaccharide stroma confirming the diagnosis of myxoma (image courtesy of Dr Patankar, Blackpool Victoria Hospital, Blackpool).

known about the rate of myxoma growth and their malignant potential.

Macroscopically, cardiac myxomas are usually pedunculated with a lobulated surface, often covered with thrombi.⁶ Less commonly, villous and papillary myxomas are also seen, which tend to be more friable and as a result, understandably have a higher risk of embolisation.⁷ Most myxomas are 4–5 cm in diameter although, rarely, larger tumours exceeding 10 cm in diameter have been reported too. Myxomas are thought to originate from pluripotent mesenchymal cells from the embryonic foregut.⁸ Histologically, myxomas comprise rows and clusters of polygonal cells embedded in an acid-mucopolysaccharide rich matrix.⁹ Vascular endothelial growth factor secreted by these cells leads to the formation of a rich network of blood vessels particularly at the base of the tumour providing nutrients to support its growth.¹⁰ In addition, foci of calcification, cysts and haemorrhage are occasionally seen within the myxomas.

Clinical features of myxoma mainly depend on their diameter, location and consistency, and can be categorised into those caused by embolisation, intracardiac obstruction and constitutional symptoms. Nearly one third of the patients with myxoma develop embolisation of the tumour, the most dreaded complication is the systemic embolisation of left atrial myxomas leading to cerebral, renal, coronary and visceral infarctions.^{9 11} Less commonly, right sided cardiac myxomas can embolise to the pulmonary circulation resulting in saddle pulmonary embolism, pulmonary infarction and pulmonary hypertension.⁵ Paradoxical embolism in those with a patent foramen ovale has also been reported.

While myxomas rarely develop on the heart valve leaflet surface, they can cause a functional obstruction of the mitral and tricuspid valves ultimately resulting in diminished filling of the left ventricle.⁹ The degree of valvular dysfunction depends on the size and mobility of the tumour and may be influenced by the body position. Intracardiac obstruction by a myxoma may lead to dyspnoea, pulmonary oedema, right sided heart failure, syncope and even, sudden death.

Non-specific constitutional symptoms including fever, lethargy, fatigue, skin rash, anorexia and weight loss in patients with myxoma are considered to be mediated by cytokines released by the tumour cells.¹² Physical examination is usually normal but may reveal a rumbling diastolic murmur due to the tumour or a functional mitral valve obstruction.

Routine laboratory investigations are often unhelpful in making the diagnosis of myxoma. Normocytic anaemia with a moderately elevated erythrocyte sedimentation rate and C-reactive protein are common findings on blood tests.¹³ Occasionally, there may be an associated haemolytic anaemia due to mechanical breakdown of red blood cells by the tumour surface. This may be manifest on blood film with a finding of schistocytes, indicative of a microangiopathic haemolytic anaemia. ECG may be normal, however new onset atrial fibrillation, bundle branch block, and changes in keeping with atrial enlargement and ventricular hypertrophy have also been recorded. Chest X-ray is usually normal but may show an enlarged cardiac shadow with or without pulmonary congestion and oedema.

Transthoracic echocardiography is a widely available non-invasive diagnostic modality and can demonstrate the size, location and mobility of a myxoma in a two-dimensional mode. Transoesophageal echocardiography provides further information regarding tumour characteristics including consistency, attachments and presence of thrombi.¹⁴ Moreover, it can also help to detect coexisting infective vegetations. It can also help to differentiate between left atrial myxoma and intramural thrombus based on their size, origin, morphology, mobility and prolapse.

Atrial thrombi are classically located in the atrial appendage and may be associated with atrial fibrillation, dilated cardiac chambers, systolic dysfunction and prosthetic heart valves. Cardiac myxomas, on the other hand, tend to be larger, often polypoidal, typically originate from fossa ovalis. Cardiac myxomas also tend to enhance with echo contrast agents as opposed to the blood clots which show no enhancement at all.¹⁵ To add to the complexity, myxoma surface is often covered with thrombi making the distinction between the two, even harder. Cross-sectional imaging especially cardiac MRI scan can offer detailed views of providing clues to its composition and anatomical relationship with surrounding mediastinal structures. This is integral to the management of these tumours as the imaging modality helps determine a plan for surgical resection.¹⁶ Nonetheless, if cardiac MRI scan is not available, cardiac CT scan can be used for further evaluation. In case of multiple tumours, positron emission tomography–CT can be utilised to identify the pattern and nature of these lesions. Other differential diagnosis of myxoma mainly includes metastatic deposits in the heart which have over 30 times higher incidence than primary cardiac tumours.¹⁷ Large infective vegetations can also mimic cardiac myxomas.

Coronary angiography is not routinely needed for younger individuals with low probability of ischaemic heart disease and is often reserved for patients aged 40 years and above in order to rule out coexisting coronary artery disease, prior to surgery.¹⁸

To date, surgical resection is the only available effective treatment for cardiac myxomas.⁹ In order to avoid tumour embolisation and other cardiovascular complications, it is imperative that the surgery is carried out promptly after the diagnosis is made. While surgical resection of cardiac tumours can be carried out during pregnancy, it requires careful consideration of intended benefits and perceived risk of fetal complications. Following successful excision of myxoma, the thromboembolic risk during subsequent pregnancy is not increased hence routine anticoagulation is not required.¹⁹ Surgical resection of the tumour is usually performed under general anaesthesia using a median sternotomy approach and cardiopulmonary bypass.²⁰ Postoperative complications include supraventricular arrhythmias and atrioventricular blocks, which can affect up to one quarter of patients with atrial lesions and especially those located close to the atrio-ventricular (AV) node. Surgery is associated with a low perioperative mortality (<3%) and offers excellent long term prognosis.²¹ Minimally invasive surgical techniques involving anterior thoracotomy, mini sternotomy and trans-septal approach have been described in the literature as safe and effective alternatives to the standard median sternotomy.²² In addition, many centres have

reported promising results with the use of port-access endoscopic and robotic myxoma surgery under live transoesophageal echocardiography guidance.²³ These newer techniques offer successful tumour resection with the added benefit of smaller skin incision, quicker recovery and reduction in the length of stay. Recurrence rates are generally low at approximately 2% in sporadic cases. However, patients with familial cardiac myxomas are at a higher risk and may need monitoring for recurrence after surgery.²⁴

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Learning points

- Cardiac myxoma is an extremely rare cause of postpartum breathlessness and tachycardia.
- Transthoracic echocardiogram is a useful initial investigation to determine the size and location of cardiac tumours.
- Tumour embolisation and intracardiac obstruction can lead to serious complications if these lesions left untreated.
- Surgical excision is the only available treatment option and should be carried out promptly in order to avoid complications.

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