Right atrial myxoma: An unusual cause of clinical right heart failure

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ABSTRACT

Myxomas are the most common type of primary cardiac tumour with 86% occurring in the left atrium. Right atrial myxomas is far less common. Patients usually present with progressive dyspnoea, fever, weight loss, right-sided heart failure, or pulmonary embolisation. We report the case of a 55-year-old man who presented with progressive dyspnoea and signs of right heart failure secondary to a large right atrial myxoma, which was diagnosed with a bedside transthoracic echocardiogram. This was successfully removed surgically.

Keywords: Atrial myxomas, dyspnoea, right atrium, right heart failure, outcome, treatment

INTRODUCTION

Primary cardiac tumours are rare with a reported incidence ranging from 0.001% to 0.19%. [1] Myxomas are the most common accounting for over 50% of primary cardiac tumour with 84% occurring in the left atrium, as a mobile mass connected by a stalk to the inter-atrial septum usually close to the fossa ovalis. [1, 2] Right atrial myxomas however are rarer with a reported incidence of 11% of all cardiac myxomas [2] Patients with right atrial myxomas usually present with progressive dyspnoea, fever, weight loss, right-sided heart failure, or pulmonary embolisation. [1, 3] Diagnosis usually requires a high degree of suspicion and confirmation by echocardiography. We report the case of a 55-year-old man who presented with progressive dyspnoea and signs of right heart failure secondary to a large right atrial myxoma, measuring 8cm x 10cm which was confirmed by a bedside transthoracic echocardiogram. This was successfully removed surgically.

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CASE REPORT

A 55-year-old man was seen at the Accident and Emergency Department with a history of progressively worsening shortness of breath on moderate exertion with associated mild oedema of the lower limbs for the last two

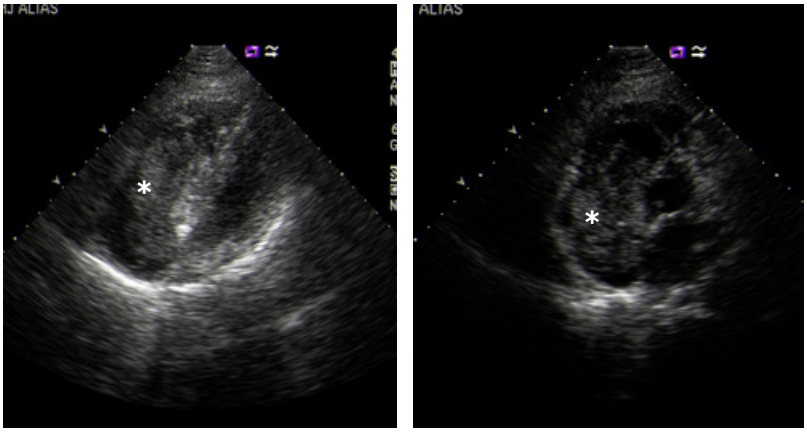

Fig. 1: Transthoracic echocardiography showing right atrial myxoma (asterisk) prolapsing into the right ventricle.

months. There was no history of angina or syncope. He is known to have diabetes mellitus, hypertension, and dyslipidaemia for which he was currently on treatment. He is a current smoker with no family history of any cardiac disorders.

On clinical examination, he was afebrile with a resting pulse rate of 80 beats per minute, blood pressure of 140/90 mmHg and a normal respiratory rate of 14 breaths per minute. The only abnormal clinical findings were a raised jugular venous pulse, a loud grade 2/4 systolic murmur and mild pitting pedal oedema. Breath sounds normal. Abdominal examination showed mild non-tender hepatomegaly but no splenomegaly. The rest of the physical examination was otherwise normal.

He was admitted to the medical ward with a preliminary diagnosis of right heart failure. A chest radiograph revealed normal cardiac size and an electrocardiograph (ECG) revealed normal sinus rhythm. Routine blood investigations were also normal.

An urgent bedside trans-thoracic echocardiogram was performed which showed a large mobile caulifower-liked tumour arising from the base of the inter-atrial septum and prolapsing into the right ventricle and inferior vena cava (IVC). A formal departmental transthoracic echocardiogram confirmed the above findings and a diagnosis of a right atrial myxoma was made. An urgent coronary angiogram showed normal coronary arteries.

He underwent an emergency open-heart surgery on the same day to remove the large right atrial myxoma. Intraoperative transoesophageal echocardiogram was performed prior to a median sternotomy, which showed the large right atrial myxoma (Figure 1a). In the mid-oesophageal atrioventricular short axis and transgastric left ventricular short axis views, the myxoma measured about 6cm x 7cm on echocardiography and was prolapsing into the right ventricle and right ventricular outflow tract causing an impairing right ventricular inflow (Figure 1b). Bicaval cannulation of both the superior and inferior vena cava was achieved carefully so

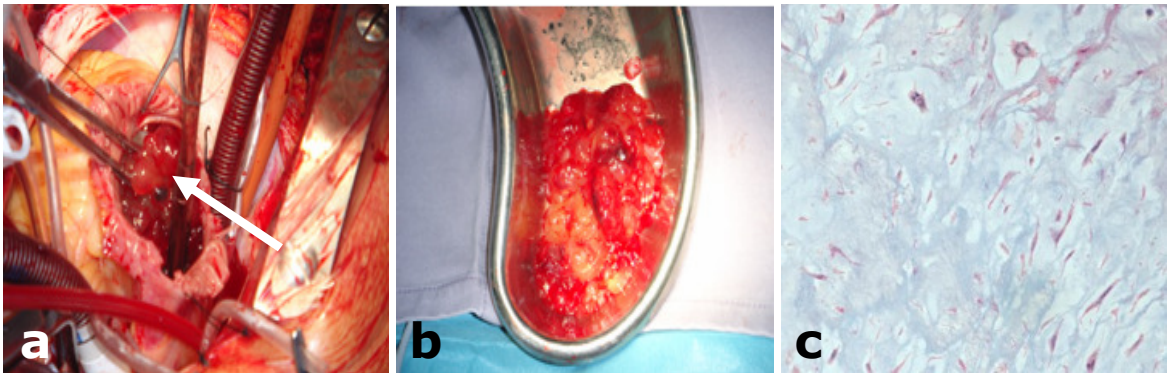

Fig. 2: a) Intraoperative image of atrial myxoma (white arrow), b) specimen after resection and c) the histology of the myxoma.

as not to disturb the tumour mass which is usually very friable and may fragment and embolise into the pulmonary artery. Ascending aortic cannulation was achieved and cardiopulmonary bypass was started on full heparinisation at an activated coagulation time of greater than 400 sec.

Aortic cross clamp was applied and the heart arrested using antegrade cold blood cardioplegia. Right atriotomy was made and the large sessile myxoma (Figure 2), measuring 8 x 10 cm arising from the base of the interatrial septum was excised and the resultant posterior interatrial septum defect was repaired directly by suturing the edges together. After complete de-airing, the atriotomy was closed using 3/0 prolene continuous sutures. Post-operative recovery was uneventful and the patient was well enough for discharge a week after surgery. Histopathology assessment of the myxoma showed abundant extracellular myxoid matrix, composed of proteoglycans, with polygonal, stellate or spindle cells spread out in the matrix (Figure 2).

He was seen in the clinic one week and one month after discharge without symptoms of right heart failure. Repeat echocardiogram showed no residual lesion in the right atrium with good RV and LV functions. He is currently under regular review at the cardiology clinic.

DISCUSSION

Cardiac myxomas account for 50% of all benign cardiac tumours in adults with over 84% found in the left atrium. [1, 2] Right atrial myxomas however are rare and consist of only 11-20% of all cardiac myxomas. [1, 2] They usually arise from the interatrial septum, at the border of the fossa ovalis. [1] Atypical locations and multiple myxomas occur most frequently in cases of familial myxoma but familial history was absent in this patient. [1] Myxoma can present in any age group with a peak incidence between the 3rd and 6th decades of life. [1]

Although cardiac myxomas are histologically benign, they may be lethal because of their strategic position. They can mimic every cardiac disease ranging from valvular heart diseases, cardiomegaly, syncope, cardiac arrhythmias and as in this case symptoms of right heart failure from a right cardiac myxoma. [1, 3] Symptoms can also mimic infec-

tive endocarditis, immunologic and malignant processes. [1]

Signs and symptoms arising from a cardiac myxoma are dependent on the size, mobility, and location of the tumour. [1, 4] Only 7% of patients are asymptomatic at diagnosis. The majority of symptoms consist of exertional dyspnoea (46%), palpitations (44%), syncope (17%) and low grade fever (24%). [4] Blood parameters may show increased erythrocyte sedimentation rate (26%) and hypochromic anaemia (9%). [4] Less common signs include pleural (7%) and pericardial effusion in 7%. [4] Because of the friable nature of the tumour, embolic events are reported to occur in 30-40% of patients. [1]

Trans-thoracic 2D-echocardiography is commonly used to determine the location, size, shape, attachment and mobility of the tumour. [1] Trans-oesophageal 2D-echocardiography will provide better visualisation of the site of tumour attachment, atrial septum, atria and both ventricles. [1] Other more sensitive modalities such as computerised tomography and magnetic resonance imaging have been used to diagnose smaller (0.5 to 1.0 cm) lesions. [1] Coronary arteriography in patients over 40 years of age is generally recommended to assess for concomitant coronary artery disease before undergoing surgery. [1]

Surgical removal of the tumour should be performed as soon as possible. Excessive manipulation of the atrium should be avoided during bicaval cannulation to avoid embolisation of the tumour. The tumour should be handled with care and resected together with a cuff of normal atrial septum from which the tumour arises to avoid recurrence. [4] Repair can be achieved either by direct suturing in small defect to replacement with autologous pericardium if the defect is large. The long-term prognosis is excellent with actuarial 10-yr and 20-yr survival of 96.8 ± 1.8% and 91 ± 4% respectively. [2, 4] Tumour recurrence is rare and was first reported by Gerbode and has since been noted by others. [5] With good surgical clearance of tumour, re-operation was not required in as high as 98.4 ± 1.3% at five years and 96.8 ± 1.8% at 10 years have been reported. [4] Regular follow-up with echocardiography is essential.

In conclusion, our case highlights the importance of doing a routine echocardiography for any patient with any significant cardiac symptoms before doing any stress cardiac evaluation. This is also a clear example of a curable clinical condition if diagnosed at first presentation and that giving an emergency treatment, can save a life.

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