Right atrial myxoma at Muhimbili National Hospital: a case report

ETM Nyawawa, EV Ussiri, H Mwakyoma, IJ Mosha, T Waane, E Lugazia, R Luchemba, BJ Nyangasa, P Chillo, B Wandwi and U Mpoki

Abstract

Background: Primary cardiac neoplasms are rare most of them are benign. Cardiac myxomas are the commonest benign lesion and the initial presentation may be accompanied with pulmonary, cerebral or systemic complications. Complete resection of the tumour is associated with good prognosis but recurrence is a frequent post operative complication. We report a first rare case from our centre of right atrial myxoma occurring in a young lady of 20 years old, a diagnosis and technique of resection is also explained.

Case presentation: We report a rare case of a young girl who had presented to us with history of shortness of breath and easy fatigability, she was also found to have features of both upper and lower cava hypertension and was in NYHA class IV. The 2-D echocardiography revealed a right atrial tumor encroaching the tricuspid valve, chest radiography showed gross cardiomegally and right lower lung collapse. A clinical diagnosis of right atrial tumour was reached. The patient was scheduled to undergo open heart surgery and tumour resection on 23rd September 2009 and was prepared accordingly. A classical median sternotomy followed by major vessel cannulation in which the cava were cannulated distally. Patient was cooled to 22 centigrade the tumour was found filling the whole of right atrium cavity, friable with a broad stalk on the right atrial appendage extending and infiltrating the crista terminalis. It was excised and tissue was taken for histopathology. The right atrium was reconstructed and closed with adequate size of atrium. The histopathology revealed a typical right atrial myxoma. Postoperatively the patient developed massive right sided pleural effusion that was managed by tube thoracostomy. Eventually the patient recovered and was discharged to be followed at outpatient clinic.

Conclusion: Right atrial myxomas are rare lesion occurring in 5-10% of all cases of cardiac myxoma. The peak incidence is between 3rd to 6th decades of life. Our case was unusual as it occurred in the 2nd decade and in the right atrium with multicentric origin though still confined in the right atrium. She also presented with pulmonary complication of right sided lung collapse. Early diagnosis and resection is followed by good prognosis and recurrence rate is low. Resection was done and the postoperative period was uneventful.

Keywords: cardiac myxoma, atrial tumour, cardiac tumour, multicentric, subendocardium, mesechymal tissue

Background

Case presentation

A 20 years old lady from Tanga region had made several visits to the regional Hospital for the past six years presenting with easy fatigability and shortness of breath. At the time she was referred to our centre her symptoms had progressed to a state of being bed ridden. Physical examination was found to have splenomegally and tender hepatomegally, the jugular venous pressure was raised with features of superior vena cava obstruction. The chest radiography showed gross cardiomegally with consolidation of the right lower zone. The electrocardiogram showed a low voltage with multiform ventricular premature complexes, ectopic atria tachycardia and the heart rate was 102 beats per minute. The Echocardiography showed gross dilatation of the right atrium, moderate left ventricular dysfunction, asymmetry of the intraventricular septum, compressed tricuspid leaflets, dilated superior and inferior vena cava, presence of a soft tissue mass filling the large part of the right atrium, the fraction of shortening was 17% and left ventricular ejection fraction of 30%. The abdominal ultrasonography showed moderately enlarged liver with preserved echo texture, hepatic vessels were dilated, there was minimal ascites and rests of the organs were normal. Ascitic tap for cytology showed few lymphocytes and histiocytes. She had hyperbilirubinemia direct 17uml/l (0-8.8), total 22uml/l (3.4-20.5), raised globulin level 49g/l (26-44), Aspartate transaminase was raised to 80u/l while the rest of liver parameters were normal. The CBC showed haemoglobin of 11.8g/dl, hematocrit 35.3%, mean corpuscular volume 80fL, platelet count of 294K/ul, leucocytosis 14.6K/ul, neutrophilia 78.8%, monocytosis 6.98% and lymphopenia 11.2%. She was negative for Hepatitis B surface antigen and Human immunodeficiency virus. A clinical diagnosis of heart failure NYHA class IV due right atria myxomatous lesion was reached. She was scheduled to undergo open heart surgery and tumour excision on the 23th July 2009. Preoperative preparation including consent, blood grouping and cross-matching, fresh frozen plasma and platelet concentrate were done.

Surgical technique

Having anaesthetized with arterial and central line in place; routine positioning and skin preparation were done. The mediastinum was opened through classical median sternotomy, mobilization of major vessels done, patient heparinized followed by major vessels cannulation, a needle for cardioplegia was also inserted cardiopulmonary bypass started on and patient cooled to 27 Centigrade; aortic cross-clamp applied and cardioplegia administered heart was arrested in diastole. The tumour was approached through trans-right atriotomy with finding of a gross degenerative tumour filling almost the entire right atrium, infiltrating the crista terminalis and the tumour had broad stalk on the right atrial appendage and the large part of the right atrial appendage was infiltrated with tumour.

The tumour was excised with large part of the right atrial appendage including part of the crista terminalis and taken for histopathology examination. Reconstruction and closure of the right atrium done and the patient was gradually weaned from cardiopulmonary by-pass. The heart attained good sinus rhythm, contractility and good hemodynamic function. The total operating time was 3.5 hours, duration of aortic cross-clamp 35 minutes and duration of cardiopulmonary bypass 77 minutes.

Postoperative management

Post-operatively the patient was shifted to the intensive care unit kept on inotropic support with Dobutamine 12ug/kg/min at flow of 22ml/hr, midazolam 1mg/hr, morphine 1-2mg prn within 5min, intravenous fluids; Ringer's lactate alternate Dextrose saline at 95ml/hr to keep CVP above 8-12cmH_2o. Ventilator setting; mode IPPV,VT=600ml, FIO2=60%. F=14, PEEP=5, PAW

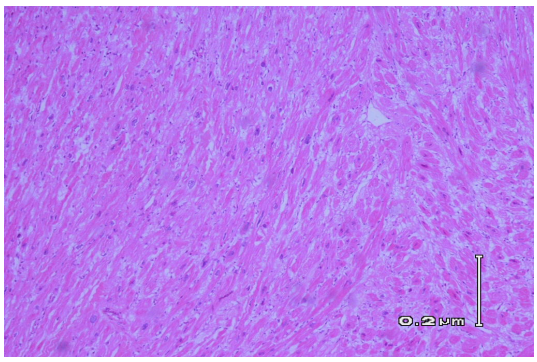
Correspondence to: Evarist T.M Nyawawa E-mail:magendoetn@yahoo.com

38cmH20 intention to change the mode to SIMV on show of effort to breathe. She also received i/v ceftriazone 1g once a day for five days, i/v ranitidine 50mg 8 hourly for five days, i/v lasix 40mg bid for five days.

While in the ICU the patient had good hemodynamic functions and was extubated 21.7 hours post operatively. She started lung physiotherapy and resumed anti- failure heart drugs. A control chest radiography on the 3rd day showed fluid level and lung collapse on the right lower and mid zones that was the site with previous consolidation. She had a chest tube insertion on the same day. Noted on the 8th day was gross hepatosplenomegally with massive ascites necessitating tapping; it was also noted that she started desaturating to 75-90%. The prothrombin and activated partial prothrombin time were prolonged while the international normalized ratio remained within normal range. She was also found to have hypoprotenemia and was kept on globulin supplement. Patient stabilized and was discharged from the ICU on the 8th day postoperative day and discharged from the hospital 25h day

The tissue was fixed in 10% neutral buffered formalin, paraffin embedded and slides were routinely stained by Haematoxylin and Eosin. Microscopic evaluation of the slides revealed a tumour characterized by extensive fibrosis, recent haemorrhage and old haemorrhage (hemosiderin) associated lymphocytic inflammatory cell infiltrates. Vascular thrombi were also encountered. Calcifications evidenced Gamma bodies (Black staining of elastic fibers) and cholesterol clefts were frequently seen. These histomorphological features were consistent with the diagnosis of Myxoma. (Fig 1, 2, 3, 4, 5)

A



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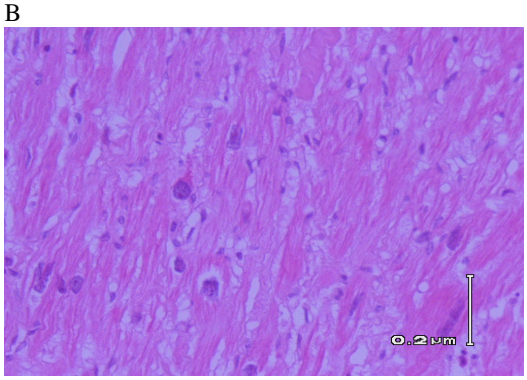
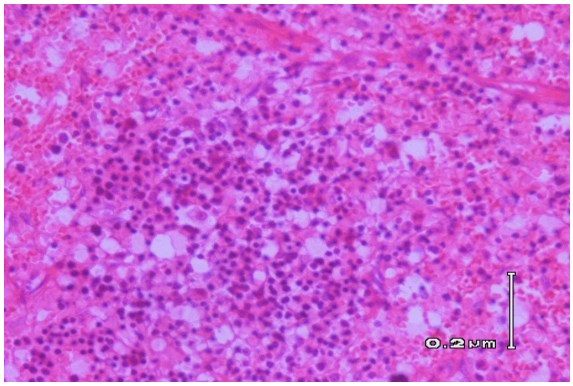


Figure 1: Fibrosis and gamma bodies (×40)

A



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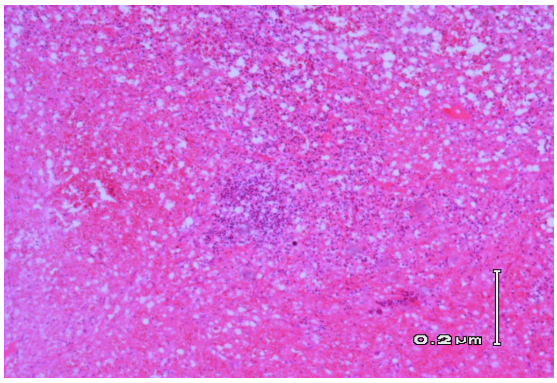
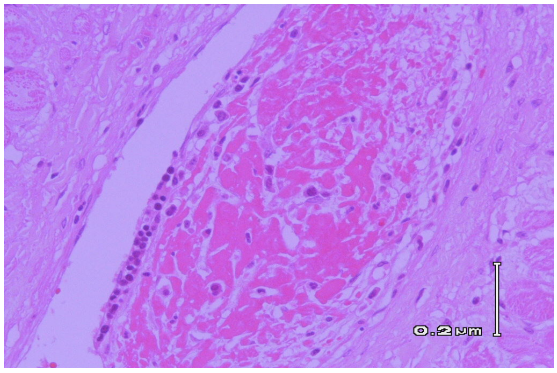


Figure 2: Lymphocytic infiltrate (A) and Haemorrhage (A&B) in a portion of cardiac myxoma(×40)

A



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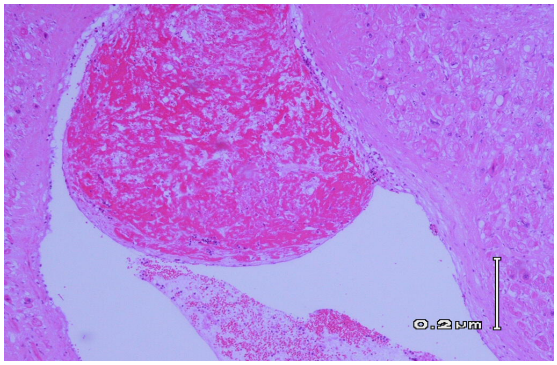

Figure 3: A thrombus and Inflammatory cell (A) and another thrombus without inflammatory cells (B) in a blood vessels in cardiac myxoma (×40)

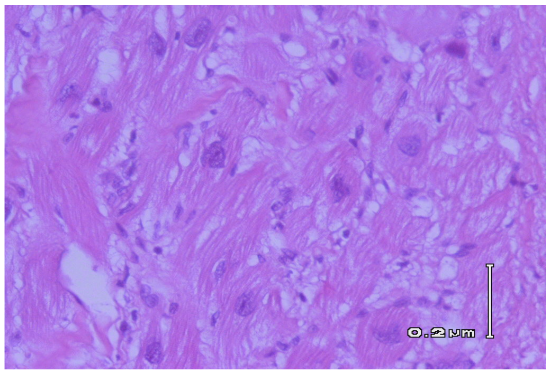

Figure 4: Calcification of elastic fibers (gamma bodies) in an atrial cardiac myxoma(×40)

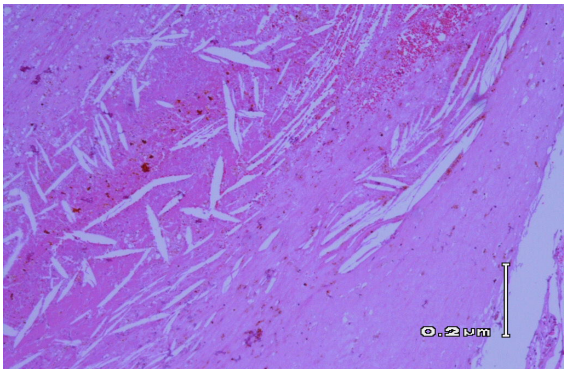

Figure 5:Cholesterol clefts and haemosiderin pigments in a cardiac myxoma(×40)

Discussion

Cardiac tumours can be primary or secondary. Primary tumors of the heart are not uncommon. The incidences range between 0.17% and 0.19% in unselected autopsy series.[1-6] Approximately 75% of primary cardiac tumours are benign, cardiac myxoma accounts for 50% of all benign cardiac tumors. Malignant primary cardiac tumours accounts for 25% and of these 75% are sarcomas[7, 8]

The first resection of a left atrial myxoma was done by Crafoord[9] using extracorporeal circulation in 1954. Primary tumors of the heart are very rare, and the majority of them are benign, with myxomas located in the left atrium being the most common form. Cardiac myxomas usually originate from the endocardium of the atrial septum, and 95% of them are located in the atria.

A left atrial myxoma is exceptionally rare in the elderly. An autosomal dominant inherited disorder, Carney complex, is associated with atrial myxoma, spotty skin pigmentation, and endocrinopathy. If patients with cardiac myxoma show signs and symptoms of this disorder, their family members should be examined. Cardiac myxoma comprises 50%[7,8] of all benign cardiac tumuors in adults but only 15% of such tumours in children. Occurrence in infancy is rare. A vast majority of myxomas occur sporadically and tend to be more common in women than men[2,10] The peak incidence is between 3rd and 6th decades of life and 94% of tumours are solitary[11] Our case occurred in a young girl of 20 years old and it was multicentric in origin.

The Deoxyribonucleic Acid (DNA) genotype of sporadic myxoma is normal in 80% of patients.[12] About 5% of patients show a familial pattern of tumour development based on autosomal dominant inheritance.[13,14] These patients and 20% of those with sporadic myxoma have abnormal DNA genotype chromosomal pattern.[12] In contrast to the typical sporadic myxoma profile of middle aged, frequently female patients with a single left atrial myxoma, familial myxoma patients are more likely to be younger equally likely to be male or female and more often 22% have multicentric tumours originating from either the atrium or the ventricle.[15-20] Although familial myxoma has a higher recurrence rate after surgical resection 21-67%.[16,21,22] Approximately 20% of familial patients have associated conditions such as adrenocortical nodular hyperplasia, sertoli tumour of testes, pituitary tumours, multiple myxoid breast fibroadenoma, cutaneous myomas and facial or labial pigmented spots (Carney's complex).[11,21] Our case despite that it originated from the right atrium the large part had a stalk and there was diffuse multicentric origin of the tumour involving the large part of the right atrial appendage and infiltrating about three quarter of the crista terminalis. Despite that the tumour occurred in a young girl there was no family history of such illness and she had no marks of Carney's complex. Myxomas are benign mesenchymal tumours and are usually pedicled[2] with diameters of 5-6 cm. They occur in any chamber of the heart but have special predilection for the left atrium from which approximately 75% originate.[23] The next more frequent site is the right atrium where 10-20% are found. The remaining 6-8% are equally distributed between the left and right ventricles.[7] Both biatrial and multicentrics tumour are common in familial diseases. Biatrial tumours probably arise from bidirectional growth of tumour originating with the atrial septum. Atrial myxomas generally arise from the interatrial septum at the border of the fossa ovalis but can originate anywhere within the atrium including the appendage. Our patient had a tumour originating in the right atrial appendage and infiltrating the crista terminalis. In addition isolated reports confirm that myxomas arise from the cardiac valves, pulmonary artery, veins and vena cava.[24,25] Right atrial myxomas are more likely to have broad-based attachment than left atrial tumours. They are also more likely to be calcified[16,17] and thus visible on chest radiography. Ventricular myxoma occurs more often in women and children and may be multicentric.[7,26] Right ventricular tumours typically arise from the free wall and left ventricular tumours tend to originate in the proximity of the posterior papillary muscle. Grossly, about 2/3 of myxomas are round or oval with a smooth or slightly lobulated surface.[10] Most are polypoid, relatively compact, penduculated, mobile, and not likely to fragment spontaneously.[2,7] Mobility depends on the length of the stalk, the extent of attachment to the heart, and the amount of collagen in the tumour.[2] Most tumours are penduculated with a short broad base, and although sessile forms occur, they are unusual.[7,27] Less

common villous or papillary myxomas are gelatinous and fragile and prone to fragmentation and embolization occurring in about one third of the tumours.[9, 28] Myxomas are white, yellowish or brownish in colour frequently covered with thrombus[7] Focal areas of hemorrhage, cyst formation or necrosis may be seen on cut section. Most myxoma tumours appear to grow rapidly but growth rates vary and occasionally tumour growth arrest spontaneously.[2] The tumour in our patient was yellowish with areas of necrosis and friability, area of hemorrhage and thrombus. Histologically myxomas are composed of polygonal shaped cells and capillary channels within an acid mucopolysaccharide matrix.[2] The cells appear singular or in small cluster throughout the matrix and mitoses are rare[29] The matrix also contains a smattering of smooth muscle cells, reticulocytes, collagen, elastic fibres and few blood cells. Cyst, areas of hemorrhage and foci of extramedullary hematopoises are present throughout the matrix.[21,28,30] Ten percent of tumours have microscopic deposits of calcium and metastatic bone deposits as well as glandular like structures.[21,28] The base of the tumour contains a large artery and veins that connect with the subendocardium but do not extend deeply beyond the subendocardium in most cases.[21] Myxomas tend to grow into the overlying cardiac cavity rather than into the surrounding myocardium. The tumour surface is covered by a monolayer of polygonal cells with interspersed primitive blood vessels. Myxomas arise from the endocardium and are considered derivative of the subendocardium multipotential mesechymal cells[31,32] although origin from endocardial nervous tissue also has been suggested[33]. The multipotential mesenchymal cells are thought to be embryonic cells left behind during septation of the heart and capable of differentiating into endothelial cells, smooth muscle cells, angioblasts, fibroblasts, myoblasts and cartilage. This account for the occasional presence of hematopoetic tissue and bone in these tumours. There is no evidence that these tumours are of thrombotic origin.[34]

May develop after cardiac trauma including repair of atrial septal defects and transseptal puncture of the percutaneous dilatation of the mitral valve. The classic triad of myxoma clinical presentation is intracardiac obstruction, with congestive heart failure 67%, signs of embolization 29%, systemic or constitutional symptoms of fever 19% and weight loss or fatique 17% and immunological manifestation of myalgia, weakness and arthragia 5% with almost all patients presenting with one or more of these symptoms[10]. Cardiac rhythm disturbances and infection occur less frequently. Our patient had right atrial hypertension with a prominent 'a' wave in the jugular venous pulse and congestion in the neck veins typical features of superior vena cava syndrome. She had also lower body manifestation of venous hypertension with hepatomegally, ascites and oedema. Cardiomegally is frequently seen in chest radiography and the ECG shows a low voltage, chamber enlargement, bundle branch block[35], and axis deviation fewer may have atrial fibrillation in 20%[20] The 2-D Echocardiography has 100% sensitivity in detecting myxomatous tumour.[36] The TEE can detect tumour as small as 1-3mm in diameter.[37, 38,39] Both CT and MRI can detect lesions that are even small[2] 0.5-1 cm and provide information regarding the composition of the tumour[35, 38, 40,41] But echocardiography is adequate diagnostic facility.

In our case the diagnosis was made using a 2-D echocardiography. Surgical management: surgical resection is the only effective therapeutic option for patients with cardiac myxoma and should not be delayed due to anticipated complications. A median sternotomy with ascending aorta and bicaval canulation is employed. Manipulation of the heart before initiation of the CPB is minimized. For left atrial myxoma the vena cavae are cannulated through the right atrial wall with the inferior vena cava cannula placed close and laterally to the IVC-right atrial junction. Cava snares are always used to allow opening of the right atrium if necessary. The body temperature is allowed to drift down but not to induce systemic hypothermia unless reduction in flows is required. Generally approaches depend on the exact location of the tumour. Right atrial myxoma poses a special venous cannulation problems and intraopertive echocardiography may be of benefit in allowing safe cannulation. Both venae cavae may be cannulated directly when low or high lying tumour pedicles precludes safe transatrial cannulation. Cannulation of the jugular or femoral vein can provide venous drainage of the upper or lower body. Generally the SVC is cannulated distally away from the right atrium to allow adequate tumour resection, but occasionally the femoral vein cannula drainage has been used when a low-high lying right atrial tumour enchroaching on the IVC-orifice. Resection is done and inspection for multicentricity is necessary. In our case we had distal bicaval cannulation and patient was cooled to 22-26 Celcius and there was no multicentricity of the tumour. The interatrial septum, tricuspid valve and right ventricle were free from tumour.

Conclusion

Cardiac myxomas are benign mesenchymal tumours and are usually polypoid myxomatous or pedicled. They are frequently located in the left atrial atrium in 90% of cases. The diagnosis of these tumours is very important as they are frequently diagnosed when the tumour have pulmonary, cerebral or systemic complication. Right atrial myxoma are even rare and presents both diagnostic and management challenges. Early surgical resection is associated with good prognosis and low recurrence rate. Our patient had presented with features of both lower and upper hypertensive venous obstruction with pulmonary complication. She underwent a successful right atrial myxomatous resection and the postoperative period was essentially uneventiful.

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