CASE REPORT

A PROBABLE CASE OF RIGHT ATRIAL MYXOMA PRESENTING WITH FEATURES OF RESTRICTIVE CARDIOMYOPATHY

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ABSTRACT

Background: Cardiac tumors are very rare and most frequently benign. Myxomas are the most common primary cardiac tumors accounting for about 50% of cases. The clinical presentation of atrial myxomas depends on size, anatomical location and their effects on the surrounding structures rather than the histological type. Restrictive cardiomyopathy can present with features of heart failure with typical massive ascites out of proportion to peripheral oedema known as egg on stick appearance. We present a case of a probable right atrial myxoma in a 20year old lady mimicking restrictive cardiomyopathy. Case report: Mrs. HD, a 20-year old Fulani lady was referred to us with 3 years history of progressive abdominal swelling, dyspnea on exertion and easy fatiguability. No orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitation or syncope. Relevant examination findings include a chronically ill looking young lady, with egg on stick appearance and mild pitting pedal edema. She had tachycardia, blood pressure 90/70mmHg & elevated Jugular venous pressure. The liver was palpable about 4cm below the right costal margin and there was massive ascites demonstrable by fluid thrill. Trans- thoracic echocardiography revealed huge, well-defined right atrial mass measuring 64 x 35 x 56.8 mm attached to the superior wall of the atrium on apical four chamber view. Conclusion: Atrial mass should be considered in the differential diagnosis of a patient presenting with features of restrictive cardiomyopathy. Echocardiography and histology are required for definitive diagnosis.

Key words: Myxoma, Restrictive cardiomyopathy, Atrium, Endomyocardial fibrosis

INTRODUCTION

Cardiac tumors are very rare and most frequently benign. Myxomas are the most common primary cardiac tumors accounting for about 50% of cases.About 75-80% occur in the left atrium and 15-20% in the right atrium.[1-4] Myxomas are commoner in women with a peak incidence in the 3rd and 6th decades of life.[2]The clinical presentation of atrial myxomas depends on size, anatomical location and their effects on the surrounding structures rather than the histological type. They can also present with constitutional features such as weight loss, fever, anemia, elevated erythrocyte sedimentation rate and C-reactive protein.

Endomyocardial fibrosis (EMF) was first described in Uganda by Davies Jack NP in1948. It is a rare form of restrictive cardiomyopathy that is endemic in rural populace of Africa, Asia and South America. The pathological hallmark of EMF is deposition of fibrous tissue in the endocardium sparing the ventricular outflow tracts. Features of chronic heart failure may be evident but marked ascites

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out of proportion to peripheral oedema (egg on stick appearance) is a typical feature of both right and left ventricular EMF.[5] We present a case of a probable right atrial myxoma in a 20year old lady mimicking restrictive cardiomyopathy. Informed consent was obtained from the patient for publication of this case.

CASE REPORT

Mrs. HD, a 20-year old Fulani lady was referred to us with three years history of progressive abdominal swelling, dyspnea on exertion and easy fatiguability. She had history of weight loss and low-grade fever but no cough or drenching night sweats. No orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitation or syncope. She was seen at a peripheral hospital and transfused two pints of blood. Other past medical history are not contributory.

Examination revealed a chronically ill looking young lady who was not pale or jaundiced, with egg on stick appearance and mild pitting pedal edema. Weight, height, body mass index and abdominal girth were 36.5kg, 160cm, 14.3kg/$m^2$and 85.5cm respectively. She had a regular small volume pulse of 116 beats per minute and a blood pressure 90/70mmHg. Jugular venous pressure was elevated up to the angle of the jaw. The apex beat was normal. There was a grade 3/6 pan systolic murmur and a soft mid diastolic murmur at the tricuspid area. Abdomen was uniformly distended with visible anterior abdominal veins draining upward. Liver was palpable about 4cm below the right costal margin, tender, with a smooth outline and span of 14cm. There was massive ascites demonstrable by fluid thrill. Other systems were essentially normal.

Chest x-ray showed mild cardiomegaly, ECG shows sinus tachycardia and low QRS voltages. Trans- thoracic echocardiography revealed huge, well-defined right atrial mass measuring 64 x 35 x 56.8 mm attached to the superior wall of the atrium on apical four chamber view. The mass protrudes through the tricuspid valve into the right ventricle during diastole. Normal tricuspid valve, atrium to ventricle peak gradient (3 mmHg) with trivial TR. The mass compresses the left atrium (LA), with LA size of 38.98 x 12.80 mm, and LA area of 7.0cm^2.The right and left ventricles as well as mitral valve were normal, no pericardial effusion.

Pack cell volume 32%, serum electrolytesurea and creatinine are normal. Abdominal ultrasound scan revealed enlarged liver with regular outline and uniform echotexture, but without mass lesion. The hepatic vein and inferior vena cava were dilated. The gall bladder, pancreas and spleen are within normal limit. There was severe ascites. Liver function test was normal.She was placed on frusemide 40mg twice daily and carvedilol 3.125mg twice daily. Patient and family were counselled on the need for surgery but she is yet to have it due to financial constraint.

Figure 1: Massive ascites with distended superficial abdominal veins

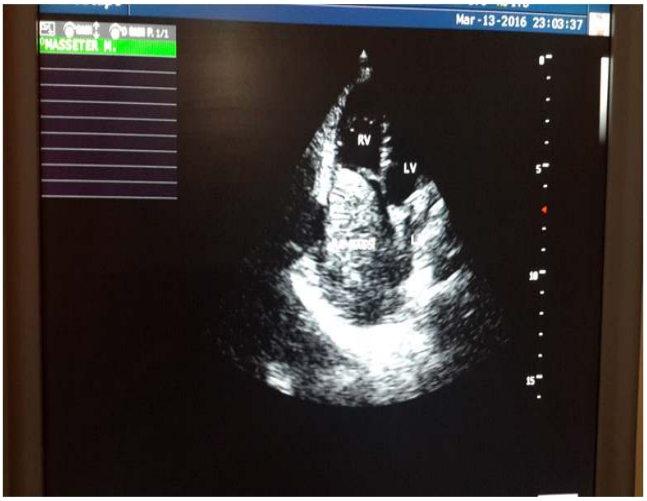

Figure 2: Modified apical four chamber view showing a right atrial mass.

DISCUSSION

Cardiac myxomasare the commonest primary cardiac tumours and arises fromthe primitive mesenchymal cells of the heart. They present in varying sizes up to 15cm. They are commoner in women and in the 3rd to 6th decades of life.[2]Presentation is usually with one or more of the obstructive features, embolic events and / or constitutional symptoms.[3-6]They can be asymptomatic in up to10% of the cases usually diagnosed at autopsy[7]. Our patient is a female but younger than the reported median age, with onset of symptoms in the 2nd decade. She presented with constitutional symptoms but no embolic events. Her presentation with massive ascites (egg on stick appearance) may be misdiagnosed as endomyocardial fibrosis or constrictive pericarditis, especially in the tropics where EMF and tuberculosis areendemic. Paucity of diagnostic facilities in this region may also delay diagnosis.

Right sided myxomas are rare but typically attach on a broad base and are more likely to be calcified than left side lesions,[8-10]similar to these findings, our patient presents with right sided probable myxoma with broad base attachment but no calcifications demonstrated. Right atrial myxoma has been reported as treatable cause of Budd Chiari syndrome, characterized by obstruction of the hepatic veins or supra hepatic part of the inferior vena cava.[11, 12] It is seen in patients with hypercoagulability, tumors or abscesses, andmay present with egg on stick appearance. Budd-Chiari is unlikely in our patient who presented with elevated jugular venous pressure, and dilated hepatic veins and inferior vena cava with no filling defect on abdominal ultrasound.

About 10% of myxomas are familial. Familial type present at younger age and have higher tendency for recurrence after resection. They are also more likely to have myxomas outside the left atrium.[7]Carney syndrome is characterizedby familial recurrent myxomas, pigmented skin lesions and endocrine neoplasm.[8] The most frequent extra cardiac involvements are pigmented skin lesions, cutaneous myxoma, adrenal cortical disease, myxoid mammary fibroadenoma, pituitary adenoma, melanotic shwannomas, thyroid disease and testiculartumors.[8] The patient we presented had no skin lesions and we have not established familial affectation.

Echocardiography is the non-invasive investigation of choice for establishing the diagnosis of atrial myxoma, however it is gradually being replaced by cardiac magnetic resonance imaging as the gold standard for diagnosis of atrial myxoma.Surgical resection is the recommended treatment with low risk of recurrence.[1-6] Cardiac computed tomography is the2nd line modality employed for diagnosis. In most African countries' diagnosis can be made correctly but the effective treatment remains elusive for most patients. This is because cardiac surgery facilities are limited in the region. The index patient was diagnosed to have right atrial based on echocardiography but awaiting surgical excision due to economic reasons.

CONCLUSION

Atrial mass should be considered in the differential diagnosis of a patient presenting with features of restrictive cardiomyopathy. Echocardiography and histology are required for definitive diagnosis. For myxoma which is the commonest primary cardiac tumour, surgery is the treatment of choice.

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