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A case report of atrial flutter and heart failure in patient with Right Ventricular Endomyocardial fibrosis

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Abstract

Endomyocardial fibrosis is a restrictive cardiomyopathy with highest prevalence in Sub-Saharan Africa, usually affecting children and adolescents. The disease usually occurs in a geographical distribution within 15 degrees on either side of equator. Literature shows that most common form is the biventricular endomyocardial fibrosis. This is a report on a 44 years old female patient with recurrent acute heart failure episodes whom diagnosed to have right ventricular endomyocardial fibrosis on Echocardiogram. Her electrocardiogram showed atrial flutter with variable blocks. Currently, she is attending out-patient cardiology clinic and her medications include tablets amiodarone 200mg daiy, warfarin 5mg daily, aldactone 25mg daily, bisoprolol 2.5mg daily and telmisartan 40mg daily. Her International Normalized Ratio (INR) is therapeutic. This case involves the presence of atrial flutter in a patient with right ventricular endomyocardial fibrosis that has not been reported. Also, emphasizes the burden of endomyocardial fibrosis in our local setting, its diagnosis and management.

Keywords: restrictive, cardiomyopathy, warfarin, amiodarone, endocardiectomy, valve replacement.

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Introduction

Endomyocardial fibrosis is a restrictive cardiomyopathy with highest prevalence in Sub-Saharan Africa usually in children and adolescents.[1]In literature, both left and right ventricular endocardial fibrosis have been described with small proportion of patients with right ventricular involvement. Left ventricular endomyocardial fibrosis usually presents with severe pulmonary hypertension and right ventricular type presents mainly with heart failure.[2] Incidence has also been reported to be reducing due to improvement in living standards, decrease in childhood malnutrition, infections, worm infestation and associated eosinophilia.[2]It can be diagnosed by non-invasive test. The main diagnostic test is the transthoracic echocardiogram. This is a report on patient with acute heart failure episodes who has right ventricular endomyocardial fibrosis and atrial flutter. This case emphasizes the burden of endomyocardial fibrosis and highlights the presence of atrial flutter in a patient with right ventricular endomyocardial fibrosis that has not been reported.

Case report

A 44-years old female patient, mother of three children, presented to our emergency department with complaints shortness of breathing and palpitations. The shortness of breathing was of gradual onset, increasing in severity, mainly on exertion for at least a week prior. This was accompanied with non-sustained palpitations and history of swelling of both lower limbs. She had several episodes of similar complaints during the past three years. During these episodes, she was treated with anti-heart failure medications that included tablets frusemide 40mg daily. There was no history of reduced urine frequency, fever, joint pains, and cough or chest pain. No history of sore throat or facial puffiness. No history of tuberculosis, hypertension, diabetes mellitus or asthma was present. She had irregular out-patient follow up with poor adherence to her medications. On examination, the BP was 123/51mmHg, Heart rate 100beats per minute of normal character, Respiratory rate 20breaths per

minute and spo2 93% on room air. Spo2 improved to 98% on 6L of oxygen by face mask. She was fully conscious, afebrile, moderately dyspneic with moderate pitting ankle, pedal and pretibial edema. She had no high arched palate, hyperextensible finger joints, arachnodactyl, finger/toe clubbing, conjuctival paleness, oral thrush, peripheral or central cyanosis. No tremors or skin lesions or thyroid enlargement. No presence of splinter hemorrhages, janeway lesions, osler nodes or conjunctival hemorrhages. There was no significant difference between blood pressure of left and right arms, jugular venous pressure was raised up to angle of mandible, displaced apex with no apical heave, no left parasternal heave or loud P_2. There was right ventricular gallop. There was also tender hepatomegaly, ascites but lungs were clear. These findings were compatible with right sided heart failure. The blood work-up showed; WBC (K/UL): 4.4, Hb level (g/dl): 13.1, Platelets (K/UL): 174, Eosinophil: 4%, Na (mmol/l): 140, K (mmol/l): 3.4, Creatinine (umol/l): 90, ESR = 12mm/1st hour. Total bilirubin (µmol/L): 12, albumin (g/L): 38 and ALT (U/L): 24 were within normal limits. T3 (ng/dl): 78, T4 (ugdl): 5, TSH (Uiu/ml): 1.9 and D-dimer (mcg/mL): 0.03 were within normal limits. Total cholesterol (mmol/L): 3.9, Triglycerides (mmol/L): 1.1 and LDL (mmol/L): 1.8 were within normal limits. Chest radiograph showed cardiomegaly and normal lung parenchyma and aortic knuckle. Electrocardiogram showed inverted T waves in leads V2 to V4 and atrial Flutter with variable blocks with ventricular rate of 60beats per minute. (Figure 1)

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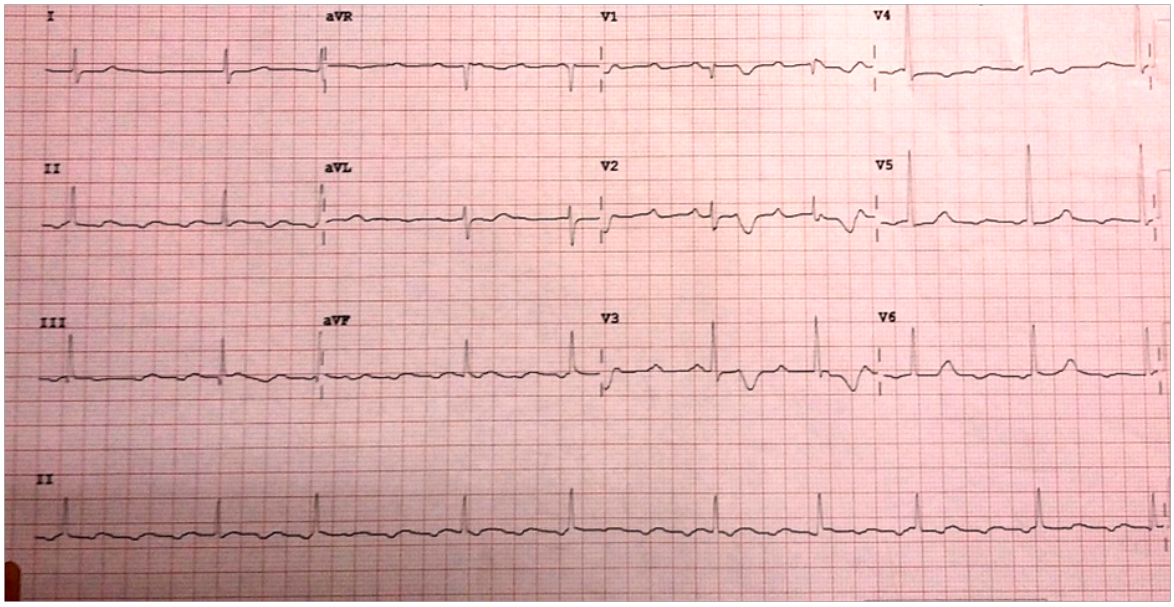
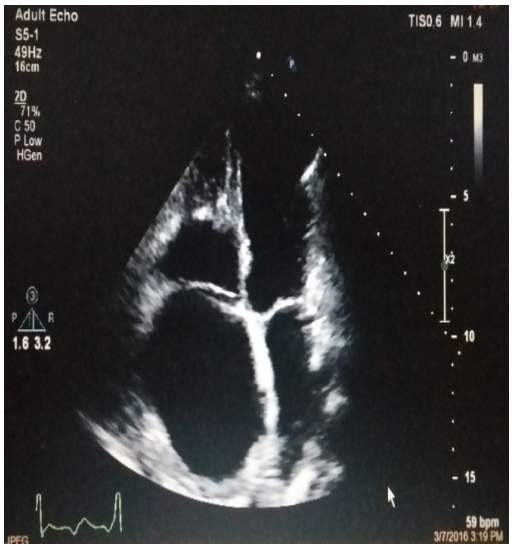


Figure 1

Echocardiogram showed grossly dilated right atrium, dilated right ventricle with right ventricular apical fibrotic obliteration. Mild systolic dysfunction of the right ventricle. Left ventricular systolic function was normal. Mitral and tricuspid valves at same level. Normal mitral inflow. Mild tricuspid regurgitation was present but no pulmonary hypertension.(Figures 2 and 3)



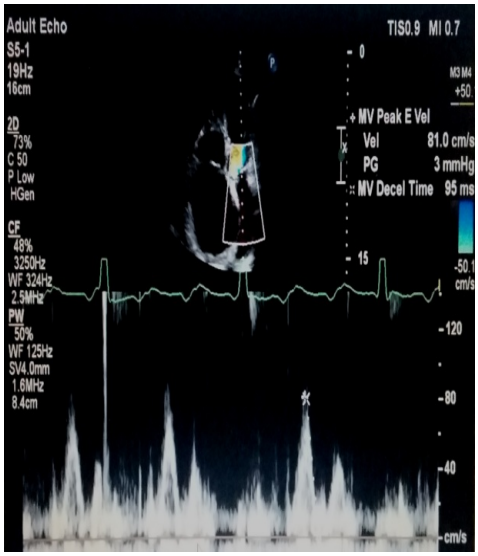


Figure 2

Figure 3

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Cardiac MRI showed hypertrophied right ventricular apex with reduced right ventricular volume. The right atrium was grossly dilated. The delayed phase showed sub-endocardial enhancement in inferior part of interventricular septum and along right ventricle apex. No thrombus was found. No features of non- compaction. The left ventricular systolic function was 63%. These findings were in keeping with right ventricular endomyocardial fibrosis in presence of right sided heart failure.

At the emergency department, she was put in cardiac position and started on oxygen 6L per minute by face mask. Intravenous Frusemide 40mg and tablet bisoprolol 2.5mg was given. She was admitted to the ward where following medications were added. Tablet aldactone 25mg daily, tablet telmisartan 40mg daily and tablet amiodarone 400mg daily. Due to high risk of thrombotic events like stroke, she was started on tablet warfarin 5mg daily. Daily weight was checked with drop from 84kg on admission to 75.6kg at discharge. There was much clinical improvement including reduced difficulty in breathing, reduced lower limbs swelling and improved exercise tolerance. Currently, she is on follow-up in out-patient cardiology clinic. She is well counseled on her fluid intake to maintain at 1.5L per day. Her current medications include tablets amiodarone 200mg daily, warfarin 5mg daily, aldactone 25mg daily, bisoprolol 2.5mg daily and telmisartan 40mg daily. Her INR is therapeutic.

Discussion

Endomyocardial fibrosis has highest prevalence in Sub-Saharan Africa usually in children and adolescents.[1] In literature, both left and right ventricular endocardial fibrosis have been described, involving apex and inflow region with atrioventricular valve regurgitation. It is also known as Davies disease.[3,4] In a study done in Uganda, the disease accounted for 15 per cent of deaths due to congestive cardiac failure.[3] Endomyocardial fibrosis is a very common and neglected cause of restrictive cardiomyopathy in Africa.[5] The disease affecting children and young adults with a

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geographical distribution within 15 degrees on either side of equator.[6] This patient has right ventricular endomyocardial fibrosis.

While, left ventricular endomyocardial fibrosis usually presents with severe pulmonary hypertension, the right ventricular type usually presents with heart failure.[2] This patient also had features of right heart failure with no pulmonary hypertension. In study done in Mozambique, 20 per cent of people studied in a rural survey had echocardiographic evidence of endomyocardial fibrosis with a male preponderance.[7] Also, literature shows that most common form is of biventricular endomyocardial fibrosis (55.5%) with a small proportion with right ventricular involvement.[7] In Cameroon, a pediatric series on endomyocardial fibrosis suggests that the disease can occurs in different age groups.[8] Also, in a case series on endomyocardial fibrosis from India, only two patients had isolated right ventricular EMF.[9] This shows that right ventricular endomyocardial fibrosis is less commonly encountered and therefore, this case is reported.

Echocardiography is main diagnostic tool used for management of disease allowing assessment of severity and extension of disease, atrioventricular valves, and presence of intracardiac thrombi.[1] Structural abnormalities of chronic severe endomyocardial fibrosis can be accurately diagnosed by transthoracic echocardiography, allowing this non-invasive technique to be used as gold standard for diagnosis and surgical management in endemic areas.[10,11] Magnetic resonance imaging is also an important diagnostic tool in endomyocardial fibrosis.[12,13] In this patient, both echocardiography and magnetic resonance imaging studies were used to reach to diagnosis.

Few case reports on right ventricular endomyocardial fibrosis have been reported worldwide. In a case report, it was reported that a female patient with right ventricular endomyocardial fibrosis presented with right-sided heart failure and no eosinophilia.[14] Another, case report from India described isolated right sided endomyocardial fibrosis with massive right atrial enlargement, complete disorganization of tricuspid valve, massive pericardial effusion, normal absolute

eosinophil count and its sporadic occurrence outside 15 degrees of the equatorial belt were interesting features in that case of endomyocardial fibrosis.[15] In this patient, there was also no eosinophilia . Our patient had no pulmonary hypertension though; pulmonary hypertension in right endomyocardial fibrosis may occur and, is related to chronic thromboembolism.[1]

Atrial fibrillation was observed in a small proportion of patients with endomyocardial fibrosis. This was associated with a greater prevalence of dyspnea, peripheral edema, hepatomegaly, lower left ventricular ejection fraction, lower right ventricular systolic pressure, and greater incidence of tricuspid regurgitation.[16] Atrial arrhythmias occur in patients with endomyocardial fibrosis and these can be persistent or intermittent.[17] Our patient has atrial flutter in background of right ventricular endomyocardial fibrosis not previously reported. This can explain why the patient had several episodes of acute decompensated heart failure.

Endomyocardial fibrosis carries a very poor prognosis. In addition to medical management of heart failure, early open heart surgery (endocardiectomy and valve repair/replacement) appears to improve outcomes to some extent; however, surgery is technically challenging and not available in most endemic areas.[18] In our local setting, these surgical approaches are not possible due to technical difficulties hence, patients need to be referred abroad for these procedures.

Hence, this is a rare case of acute heart failure episodes and atrial flutter in a patient with right ventricular endomyocardial fibrosis.

Conclusion

Right ventricular endomyocardial fibrosis is a very important cause of restrictive cardiomyopathy. Atrial flutter can also be found in addition to heart failure as present in this patient. Both can be conservatively managed.

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Legends;

Figure 1: Electrocardiogram showing atrial flutter with variable blocks.

Figure 2: 2D echocardiogram showing grossly dilated right atrium with right ventricular apical obliteration.

Figure 3: 2D echocardiogram showing normal mitral inflow.

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