CASE REPORT

Congenital Giant Right Atrium in an Elderly Woman


Abstract

Enlargement of right atrium is usually secondary to pulmonary hypertension due to valvular heart diseases or obstructive pulmonary disorders, atrial septal defect, tricuspid atresia or stenosis, pulmonary stenosis, primary pulmonary hypertension, Ebstein’s anomaly. Congenital enlargement of right atrium is rare and it commonly presents in children. Our patient presented with congenital giant right atrium at 65 years of age, other cardiac diseases being excluded. Patient developed tricuspid regurgitation, but pulmonary hypertension was absent till the date. Congenital giant right atrium has rarely been reported from India.

Introduction

Right atrial malformations receive less attention than other cardiac structural abnormalities. This can occur as a congenital malformation or secondary to other diseases. Massive enlargement of right atrium (RA) is rare and often reported in association with Ebstein’s anomaly. Giant right atrium (GRA) has also been reported in association with right ventricular myopathy.1 We here report a case of congenital GRA. As far as we could search, this could be the first report of this anomaly from eastern India.

Case Report

A sixty five year old woman from West Bengal presented with exercise intolerance and swollen legs of two months’ duration. She started having these complaints for last one year but did not visit any physician. She only used to take over-the-counter medications for symptom control. She did not have any past history of heart disease, diabetes or hypertension. She had three pregnancies, the last one 30 years ago; and all were well tolerated. There were no family history of heart

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Fig. 1: Shows raised jugular venous pulse
disease. At presentation to our hospital, she had mild cyanosis, raised pulsatile jugular venous pulse with prominent V wave (Figure 1) and bipedal oedema. Her pulse rate was 120/min; irregular and blood pressure was 80/60 mm of Hg. All the heart sounds were muffled. There were scattered crepitations in both lung fields. Oxygen saturation by pulse oximetry came as 89%. There was tender hepatomegaly but no splenomegaly.

Laboratory investigations revealed haemoglobin of 10.8 gm% with total leucocyte count of 9000/cu mm. Platelet count was 1.9 lac /cu mm. ESR was 25 mm / hour. Blood kidney parameters and liver function tests were normal. Chest X-ray revealed a hugely enlarged cardiac shadow with a prominent right heart border (Figure 2) and cardiothoracic ratio of almost 0.9. Lung fields did not show any pathology. Meanwhile, with oral antibiotics her crepitations disappeared although the other symptoms persisted. Also, after 2 days in hospital, on repeat examination, we found a right sided third heart sound. But the raised JVP persisted despite diuretic use.

Electrocardiography showed atrial fibrillation, right axis deviation, RBBB pattern and low voltage complexes. Echocardiography showed (Figure 3) a hugely dilated RA which takes up most of the echo window in four chamber view, making the other chambers compressed. The RA measured 11.7 cm by 8.7 cm with an estimated volume of 400 ml. The septae were normal and there were normal chamber and great artery relations. As far as could be seen, the left sided chambers and all valves were normal. Left ventricular ejection fraction was 54%. There was significant tricuspid regurgitation (Figure 4) with smoke like opacities in RA. The right ventricular ejection fraction was 45%. Pulmonary artery dimensions were normal and continuous wave Doppler study did not show any pulmonary regurgitation. The tricuspid valve leaflets were all in normal position and right atrial walls were normal. Cardiac catheterisation showed normal pulmonary artery pressures. The patient did not consent to trans-oesophageal echocardiography. Cardiac biopsy could not be done due to technical reasons. The case was finally diagnosed as congenital GRA, presenting with features of right heart failure and atrial fibrillation.

**Discussion**

Congenital malformation of RA or coronary sinus is rare, and cases are classified into four categories: (1) congenital enlargement of right atrium (CERA), (2) single diverticulum, (3) multiple diverticula of the RA, and (4) diverticulum of the coronary sinus (CS). CERA is also called as giant right atrial aneurysm.

The CERA may be huge enough to call as “giant right atrium” (GRA). However, the term GRA is not well defined. Kurz et al in 2004 defined idiopathic enlarged RA as an increased right atrial long axis indexed to body surface area (RALAXi, men > 2.6 cm/ m², women > 2.8 cm/ m²) in absence of other cardiac abnormalities and severe enlargement was defined arbitrarily as RALAXi ≥ 4 cm/m². Our patient had congenital/ idiopathic GRA, the dimensions of RA being remarkably high (11 cm by 8.7 cm).

Usually, the cases of CERA are sporadic. However, a report from Switzerland depicted a family with idiopathic dilatation of right atrium. In India, a case
was reported from Mumbai, describing a 16 year old boy with CERA. Like our case; this boy also had no other cardiac anomaly. Our patient presented at 65 years of age. Such late presentation of this anomaly is rare, though diagnosis of this anomaly has been reported over a wide range of ages starting from in utero to old age. In a case series from Japan, idiopathic bi-atrial enlargement has also been described.

Congenital GRA may remain asymptomatic and found incidentally after a chest X-ray showing enlarged cardiac shadow with prominent right border of heart. Symptoms of GRA include shortness of breath, palpitations or arrhythmia. Usually, there are features of right sided congestion, but sometimes, tachycardia induced left ventricular failure can also occur. Repeated pulmonary embolism from the dilated atrial chamber can cause features of pulmonary hypertension. In our patient though elderly, there was no pulmonary hypertension. As the figure shows, smoke like echo contrast was present in RA of our patient and we started her on warfarin.

Among the patients of RA malformation, CERA and large diverticula of RA frequently come to medical attention because of a gross enlargement of cardiac silhouette on chest X-ray or because of atrial arrhythmias. In contrast, cases of right atrial dilatation with diverticula of coronary sinus usually present with supraventricular tachycardia associated with accessory pathways that transverse the diverticulum to form an atrioventricular connection. Ebstein’s anomaly, pulmonary hypertension or atrial septal defect can also cause dilated RA, but in these cases, other symptoms of the associated disease will be present. Ventricular pathologies like endomyocardial fibrosis can also present with GRA. Although endomyocardial fibrosis is rare in India, we need to be aware of this entity and appropriate tests should be done to rule out this entity.

Echocardiography is commonly used for diagnosis of atrial enlargement, but cardiac MRI is more sensitive and specific. It helps to delineate cardiac structure and function, specially when the images are displayed in cine loops. Flow volumes and velocities of forward and regurgitative flow can be assessed across the cardiac valves. In our patient, cardiac MRI could not be done due to cost factor. Another cost effective diagnostic test is echocardiography with saline contrast. Surgery is the definitive management of congenital GRA. This usually involves reduction atrioplasty.

References