

CASE REPORT

Isolated Marked Right Atrial Enlargement: What Could Be the Cause?

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A 65-year-old asymptomatic male was referred for unexplained mild cardiomegaly. Electrocardiogram was unremarkable. Echocardiography revealed significant enlargement of right atrium (RA) (Figure 1a). Left atrium (LA) was normal in size (anteroposterior dimension 32 mm in parasternal long axis view) and was not compressed. Both ventricles were normal in dimensions and showed normal contractility. Tricuspid valve was normal. There was no evidence of Ebstein's anomaly. Tricuspid inflow early diastolic velocity (E wave) was 70 cm/sec with a deceleration time of 160 msec. The ratio of E and the late diastolic tricuspid inflow velocity (A wave) (E/A ratio) was 1.2. There was mild systolic mitral and tricuspid regurgitation (TR) (Figure 1a). TR gradient was 15.8 mm Hg. In addition, there was mild presystolic TR (before QRS of simultaneously recorded electrocardiogram) (Figure 1c) also. Inferior vena cava size was 15 mm in expiration and 10 mm in inspiration. Hepatic vein flow Doppler revealed diastolic flow velocity nearly equal to systolic flow velocity. Tissue Doppler imaging of tricuspid annulus was normal (Figure 1d). Other valves were normal with normal flow. Mitral E and A velocities were 100 cm/s and 60 cm/s respectively, with E-wave deceleration time 170 msec. There was no systemic illness. There was no endomyocardial pathology, thrombi or apical obliteration. Patient could not afford cardiac magnetic resonance imaging (MRI) and did not agree for endomyocardial biopsy.

Thus, our patient presented with isolated marked RA enlargement with only mild TR and normal pulmonary

arterial systolic pressure. Differential diagnoses include right-sided restrictive heart disease, idiopathic isolated RA cardiomyopathy, localized constriction, persistent or intermittent atrial fibrillation (AF), and secondary causes of RA enlargement such as Ebstein's anomaly or primary tricuspid valve disease. Our patient did not have any echocardiographic evidence of Ebstein's anomaly. Lack of significant TR and right ventricular (RV) enlargement ruled out the possibility of primary tricuspid valve disease or atrial septal defect. Persistent or intermittent AF can cause atrial enlargement, but our patient was in sinus rhythm and had no previous history of intermittent palpitations. Moreover, AF is unlikely to produce isolated significant enlargement of RA without causing LA enlargement.

Normal dimensions and normal systolic function of RV with enlargement of RA and mild TR suggested the possibility of right-sided restrictive cardiomyopathy. Presystolic TR, <50% respiratory variation in inferior vena cava size and diastolic flow preponderance in hepatic veins supported this possibility by indicating raised RV end-diastolic pressure. In addition, though tricuspid inflow Doppler findings were not classical, relatively taller E velocity with a deceleration time of 160 msec would be consistent with pseudonormal filling (esp. in view of RA enlargement and hepatic vein flow abnormalities) (1,2). However, tricuspid annular early diastolic velocity was absolutely normal (>15 cm/s) which strongly argued against the possibility of right-sided restrictive cardiomyopathy. Furthermore, isolated or dominant RV idiopathic restrictive cardiomyopathy is rare (3-5), whereas the absence of any systemic illness, endomyocardial thickening or apical obliteration excluded common secondary causes of the same. Cardiac MRI and endomyocardial biopsy are more sensitive than transthoracic echocardiography in excluding these secondary causes and could have helped, but the

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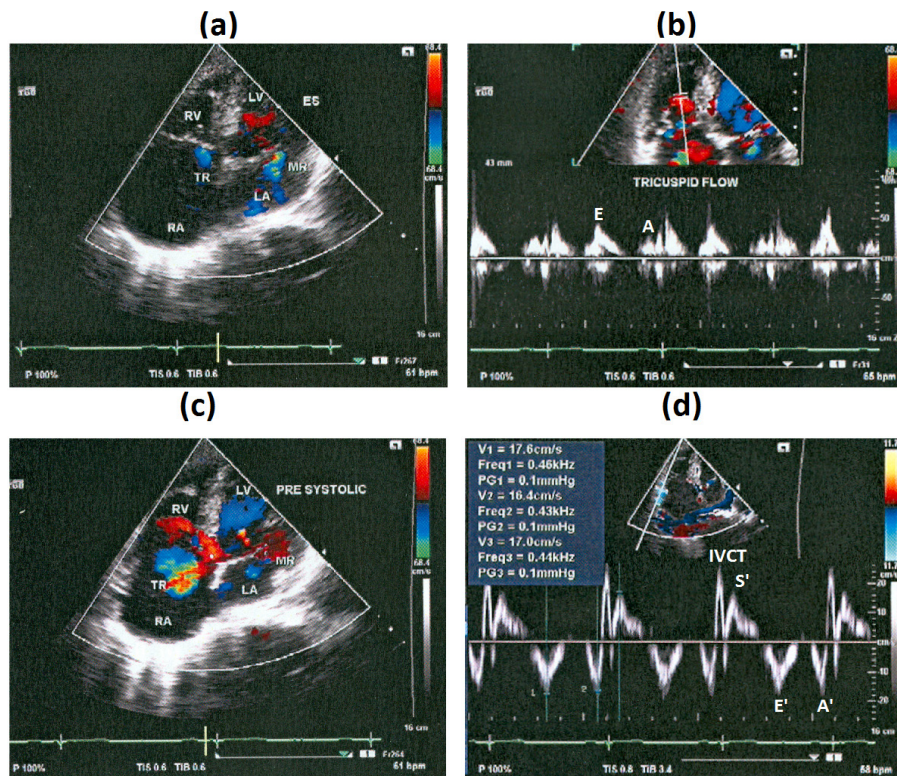


Figure 1.

- (a) Apical four chamber view during systole showing grossly dilated right atrium (RA) with mildly enlarged left atrium (LA) and normal size of left ventricle (LV) and right ventricle (RV), MR- mitral regurgitation, TR- tricuspid regurgitation.
- (b) Pulsed Doppler evaluation of tricuspid flow. E- Early diastolic flow, A- Flow during atrial contraction.
- (c) Apical four chamber view in presystole (before QRS of simultaneously recorded (ECG) showing mild tricuspid regurgitation (TR). LA- Left atrium, LV- Left ventricle, RA- Right atrium, RV- Right ventricle.
- (d) Tissue Doppler imaging of lateral tricuspid annulus. IVCT- Isovolumic contraction, S'- Systolic velocity, E'- Early relaxation velocity, A'- Velocity during atrial contraction.

patient was not willing for the same. Absence of any pericardial thickening, lack of any respiratory variation in mitral or tricuspid inflow velocities and relatively longer deceleration time of tricuspid inflow E wave argued against the possibility of right-sided constrictive physiology.

Finally, idiopathic RA cardiomyopathy is another possibility of isolated RA enlargement (6). However, in these conditions, LA is usually compressed (7,8), which was not so in our case. In addition, it would also be difficult to explain presystolic TR in isolated idiopathic RA enlargement.

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