An Unusual Cause of Breathlessness in a 70 Years Old Man

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Abstract :

Ebstein's anomaly is a complex, congenital heart defect characterized by a malformation of the tricuspid valve and right side of the heart. A variety of cardiac abnormalities are associated with Ebstein's anomaly, including atrial septal defect, conduction system abnormalities, patent foramen ovale, pulmonary stenosis or atresia, and ventricular septal defect. The clinical course of a patient with Ebstein's anomaly depends on the severity of the abnormalities present. Surgical repair of Ebstein's anomaly involves repair or replacement of the tricuspid valve and repair of any associated cardiac abnormalities. When patients of all ages are taken together, the predicted mortality is approximately 50% by the fourth or fifth decade. We present here an uncommon presentation of this complex congenital heart defect in a 70 years old man.

Key Words : congenital heart defect, Ebstein's anomaly, echocardiography

Introduction :

Ebstein's anomaly is a rare congenital heart disorder occurring in 1 per 200 000 live births and accounting for 1% of all cases of congenital heart disease. This anomaly was described by Wilhelm Ebstein's in 1866 in a report titled, Concer ning a very rare case of insufficiency of the tricuspid valve caused by a congenital malformation. [1]

Ebstein's anomaly is an abnormality of the tricuspid valve in which the septal leaflets and often the posterior leaflets are displaced into the right ventricle and the anterior leaflet is usually malformed, excessively large, and abnormally attached or adherent to the right ventricular free wall. Thus, a portion of the right ventricle is "atrialized" in that it is located on the atrial side of the tricuspid valve, and the remaining functional right ventricle is small. The tricuspid valve is usually regurgitant but may be stenotic. [2]

Case History

A 70 year old man presented with gradual onset dyspnea since last four months. He was totally asymptomatic before 4 months and living active life. Initially he had breathlessness after heavy work which at present is grade 2.His cardiovascular system examination was unremarkable except irregularly irregular pulse, systolic murmur in tricuspid area and audible third heart sound. He was evaluated for the same. His complete blood count, renal function, liver function, thyroid function tests were normal. Chest X ray was reported as showing cardiomegaly and right side pleural effusion (Fig1). Electrocardiogram showed atrial fibrillation and right bundle branch block (Fig 2).

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Figure 1: Chest X ray showing cardiomegaly and minimal Right sided pleural effusion.

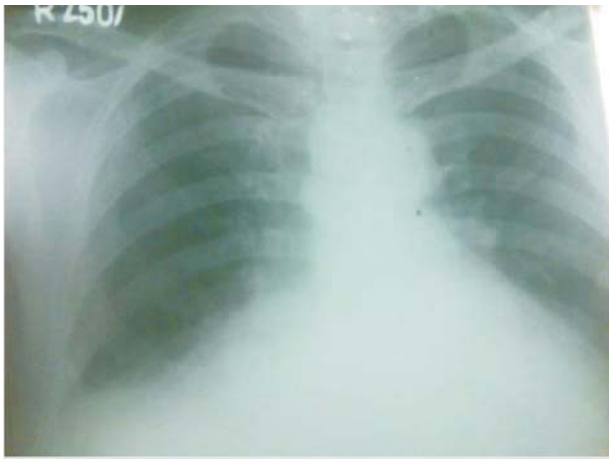
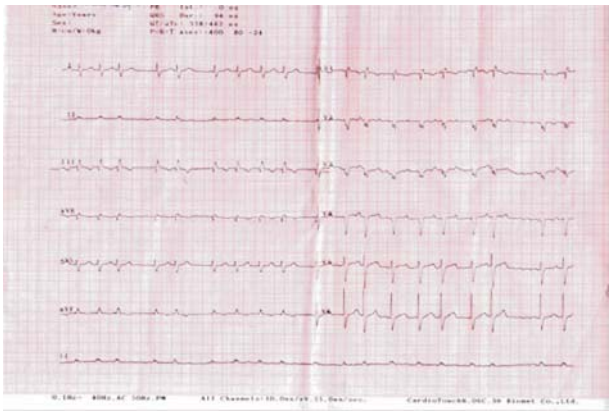


Figure 2: Electrocardiogram showing atrial fibrillation.



Echocardiogram findings were very interesting and uncommon for the age 70. As shown in images 3, 4 & 5 it revealed downward displacement of tricuspid valve into right ventricle with atrialisation of right ventricle and moderate tricuspid regurgitation without significant pulmonary hypertension suggestive of Ebstein's anomaly. (Fig 3, 4 &5)

Figure 3: Echocardiogram in APICAL 4C VIEW which shows atrialised portion of right ventricle and apical attachment of tricuspid valve leaflet.

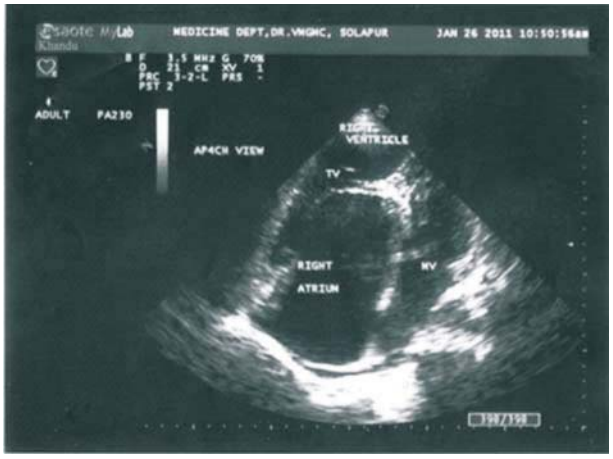


Figure 4: Echocardiogram with CONT WAVE DOPPLER across TV showing low pressure TR.

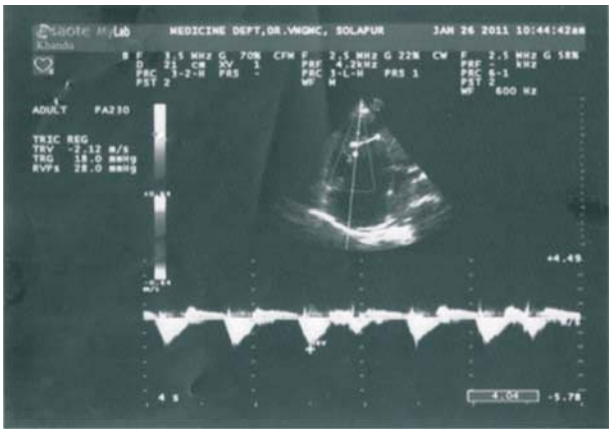
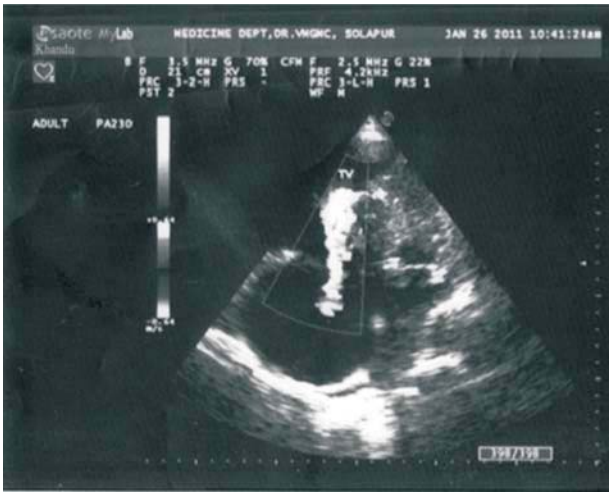


Figure 5: Echocardiogram with COLOUR FLOW across TV.



Discussion

The natural history of patients with Ebstein's anomaly depends on its severity. When the tricuspid valve deformity and dysfunction are extreme, death in utero from hydrops fetalis is the norm. When the tricuspid valve deformity is severe, symptoms usually develop in newborn infants. Patients with moderate tricuspid valve deformity and dysfunction usually develop symptoms during late adolescence or young adult life. Adults with Ebstein's anomaly can occasionally remain asymptomatic throughout their life if the anomaly is mild exceptional survival to the ninth decade has been reported. [3]

The best echocardiographic view for the evaluation of Ebstein's anomaly is the four-chamber view. Of principal importance is the accurate recording of the level of insertion of the septal leaflet of the tricuspid valve relative to the anulus. Apical displacement of this insertion site is optimally assessed in this view and is the key to diagnosis. [4]

Treatment of Ebstein's anomaly is complex and dictated mainly by the severity of the disease itself and the effect of accompanying congenital structural and electrical abnormalities. Treatment options include medical therapy, radiofrequency ablation, and surgical therapy. Antibiotic prophylaxis for infective endocarditis, medical therapy for heart failure - Angiotensin-converting enzyme (ACE) inhibitors, diuretics, and digoxin; Arrhythmia treatment- Medical treatments such as anti-arrhythmic drugs or radiofrequency ablation of the accessory pathways.[5]

References

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