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CASE REPORT

Unusual Electrical Presentation of Ebstein's Anomaly – A Case Report

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Abstract:

Ebstein's anomaly is a rare congenital heart disease. A 24 yrs male presented with shortness of breath for 6 weeks and palpitation for same duration. On examination, he found to have Pan-systolic murmur of 3/6 grading present over the left lower parasternal area with no radiation and prominent on inspiration. ECG shows wide QRS complex, Left axis deviation suggestive of LBBB. Chest X-Ray P/A view shows Cardiomegaly. Echocardiography shows septal leaflet of Tricuspid valve 37 mm apically displaced with large RA due to atrialization of RV, small RV and elongated anterior tricuspid leaflet with sail like appearance and partially restricted with RV free wall, moderate TR. So our case is Ebstein's Anomaly (Type C) with LBBB which is a rare variety as the common presentations are typical RBBB with the presence of qR pattern in V1 also suggests an enlarged atrialised RV.

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Introduction:

Ebstein's anomaly occurs in approximately one per 200,000 live births and accounts for < 1% of cases of congenital heart disease.[1,2] This anomaly was first described by Wilhelm Ebstein in 1866 in a report titled 'Concerning a very rare case of insufficiency of the tricuspid valve caused by a congenital malformation'.[3] It is a congenital malformation characterized by malformed and displaced tricuspid valve leaflets that are partly attached to the tricuspid valve annulus and partly attached to the right ventricular endocardium. A wide range of tricuspid valve abnormalities are seen in Ebstein's anomaly, ranging from minimal displacement of the septal and inferior leaflets to severe displacement with tethering. Management of Ebstein's anomaly requires understanding of the anatomy and physiology along with clinical features. Although the diagnosis is largely based on echocardiographic features, ECG is an important tool in the diagnosis of Ebstein's anomaly. Although ECG features may not be specific to the diagnosis, most patients with Ebstein's anomaly have an abnormal ECG.[2] Characteristic ECG features in Ebstein's anomaly include tall P waves (> 2.5 mm) in leads II and V1 due to right atrial enlargement.[4] In some cases, giant 'Himalayan' P waves have been described, defined as height > 5 mm.[5] The QRS complex may be prolonged in Ebstein's anomaly, due to prolonged activation of the RV. Incomplete or complete RBBB is seen in 44% of patients,[6] as seen in all three ECG cases. This happens due to a delay in conduction through the specialised conduction tissues.[7] Triphasic or tetraphasic QRS complexes can be observed. The QRS complex may be fragmented due to abnormal conduction through the atrialised RV, as was observed in all three cases. The finding of a giant P wave together with an atypical RBBB in V1 is suggestive of Ebstein's anomaly.[6] The amplitude of the R and R' in V1 is smaller than those in V5 and V6, contrary to a typical RBBB. Similarly, the presence of qR pattern in V1 also suggests an enlarged atrialised RV.[8]

Case report:

Mr. X, 24 years old normo tensive, non-diabetic male presented to us with shortness of breath for 1.5 month and palpitation for same duration. Shortness of breath is insidious in onset, gradually progressive, usually felt during moderate to severe exertion (NYHA-II). No history of paroxysmal nocturnal dyspnea&orthopnea. Palpitation is episodic, occurs during exertion and occasionally at rest, relieved by rest and subsides spontaneously. On examination patient is cooperative, Pulse:76 beats/min, BP 120/90 mmHg, JVP not raised, respiratory rate: 16 breaths/min, no visible cardiac impulse over precordium, apex beat is located in left 5th intercostal space which is normal in

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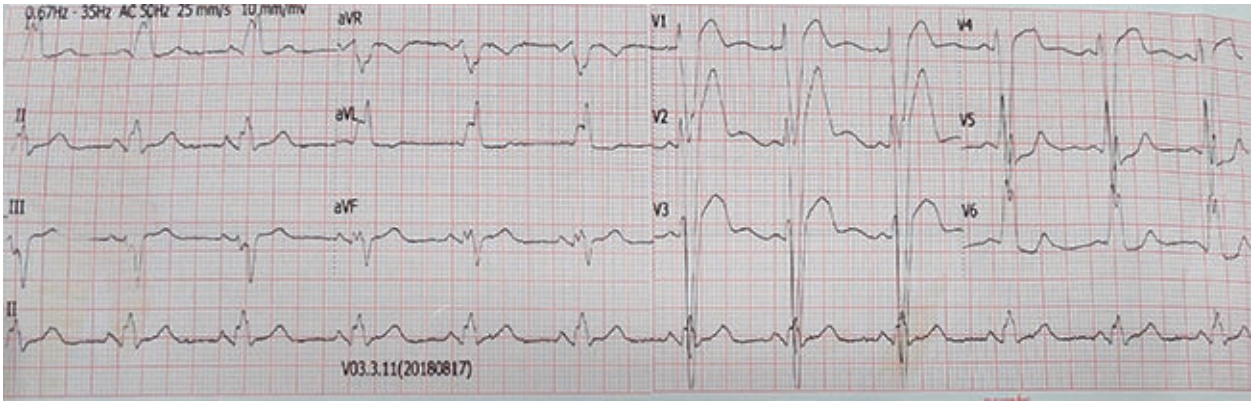

Figure 1: *ECG showing sinus rhythm and LBBB with left axis deviation*

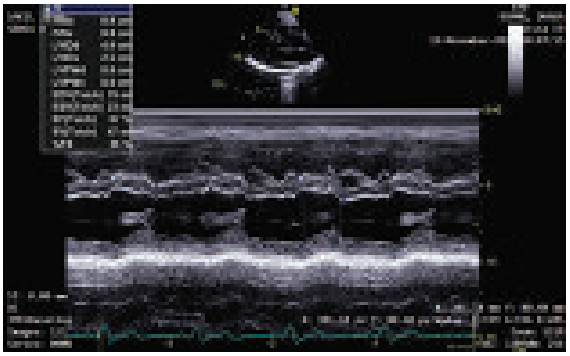

Figure 2: *TTE 2D & M Mode; PLAX view showingparadoxical IVS*

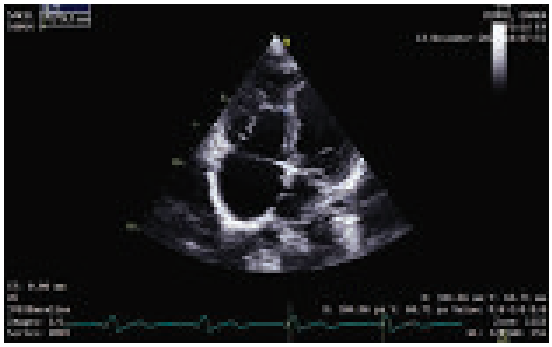

Figure 3: *TTE 2D; A4C RV Focus View Showing septal leaflet of tricuspid valve 37 mm apically displaced.*

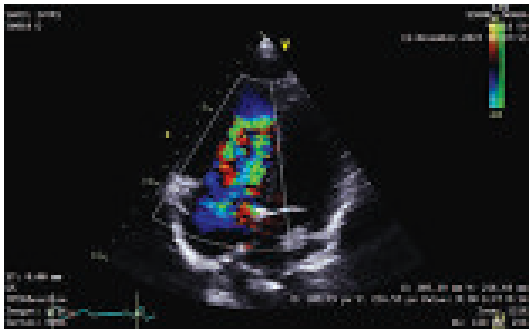

Figure 4: *TTE 2D guided Color Doppler; A4C view showing moderate TR*

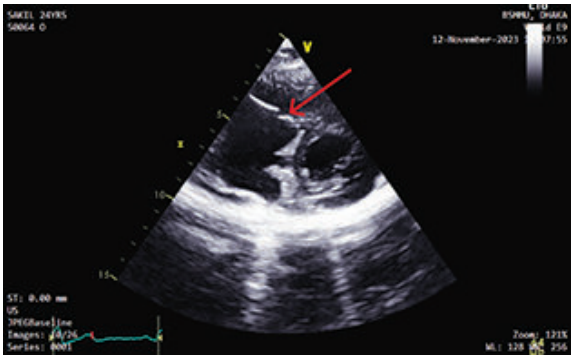

Figure 5: *TTE 2D, PSAX view at papillary muscle level showing origin of septal leaflet of tricuspid valve.*

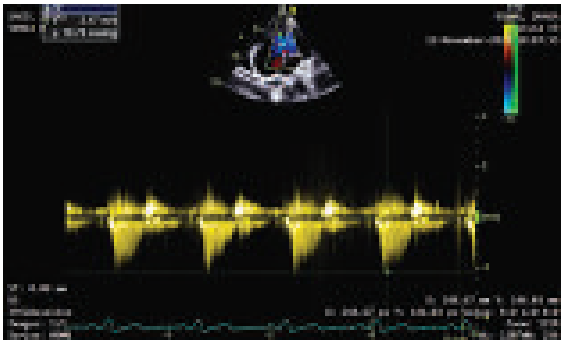

Figure 6: *TTE 2D & color guided Spectral Doppler showing TR gradient 19 mmHg (PASP= 27 mm of Hg)*

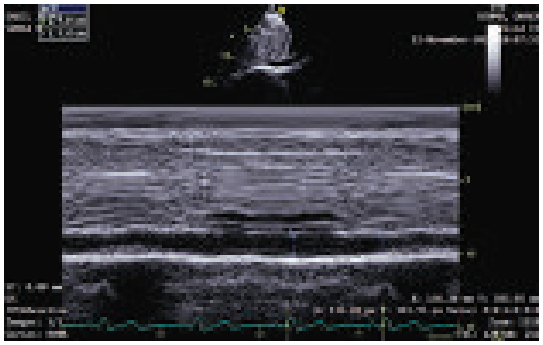

Figure 7: *TTE 2D & M Mode showing normal size IVC and <50% inspiratory collapse.*

character. 1st& 2nd heart sound normal. A Pan-systolic murmur of 3/6 grading present over the left lower parasternal area with no radiation and prominent on inspiration. Lung bases are clear. With all these complaints he was admitted in this hospital. Then he underwent some investigations. CBC shows Hb: 12 gm/dl, ESR: 16 mm/1sthour,RBC:4.94 x 10^12/L, Platelate count: 250 x 10^9/L, WBC: 8.5 x 10^9/L, N: 60%, L: 30%,M: 8%, E :2%. CKMB: 24 U/L, HS TROPONIN I: 6.1 PG/ML, RBS: 6.8 mmol/l, S. Creatinine: .8 mg/dl, SGPT: 21 u/l, ECG shows heart rate: 75 beats/min, sinus regular rhythm, wide QRS complex, Left axis deviation suggestive of LBBB. Chest X-Ray P/A view shows Cardiomegaly. Echocardiography shows septal leaflet of Tricuspid valve 37 mm apically displaced with large RA due to atrialization of RV , small RV and elongated anterior tricuspid leaflet with sail like appearance and partially restricted with RV free wall, moderate TR. So our case is Ebstein's Anomaly (Type C) with LBBB which is a rare variety. No such case has been reported in our country yet.

Discussion:

Pranil Bhalchandra Gangurde et al. showed a case which was diagnosed as Ebstein Anomaly with septal tricuspid leaflet displaced 24 mm apically with tachycardia episode showing antidromic tachycardia with LBBB morphology.[9] But in our Ebstein's Anomaly case septal tricuspid leaflet displaced 37 mm apically and LBBB is seen at rest. Amit M et al. described a case of ebstein anomaly with pre-excitation and after ablation of this case RBBB was noted. RBBB is the most common ECG finding of Ebstein Anomaly.[10] But in our case we found LBBB which is rare. Elizabeth D. Sherwin et al. showed a case of Ebstein's Anomaly where paroxysmal tachycardia with LBBB pattern and left axis deviation was seen but intravenous procainamide restored sinus rhythm with a narrow QRS duration (80 ms), normal axis, and neither right bundle branch block in the anterior precordial leads nor ventricular preexcitation.[11] Our case is unique from this mentioned case as LBBB was found in resting ECG. A case series of 3 Ebstein's anomaly were reported by Mayank Dalakoti et al. with three different electrical presentations in ECG as follows sinus rhythm with RBBB, sinus rhythm with QRS fragmentations and prominent delta wave consistent with WPW.[12] But in our case LBBB with left axis deviation was found which is quite unsual for Ebstein's anomaly.

Conclusion:

This case was Ebstein's anomaly type C with LBBB which is a rare variety. Ebstein's anomaly is mostly associated with electrical disturbance like RBBB, WPW syndrome. But Ebstein's anomaly can also be associated with LBBB.

Declaration of patient concent:

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal.

Conflicts of Interest:

There are no conflicts of interest.

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