Case Report

Absent right bundle branch block: Is it a clue of pre-excitation in Ebstein's anomaly?

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1. Introduction

Ebstein's anomaly is a congenital heart disease contributing to nearly 1% of all congenital heart lesions.1 The electrocardiogram (ECG) is abnormal in most patients with Ebstein's anomaly. It may show tall and broad P waves as a result of right atrial enlargement, as well as complete or incomplete right bundle-branch block.2 Electrophysiological evaluation and radiofrequency ablation of symptomatic accessory pathway(s) should be performed when feasible in patients with Ebstein's anomaly who have tachyarrhythmia.

A 14-year-old male with a history of symptomatic tachycardia was referred for ablation. Sinus rhythm electrocardiogram was not showing any pre-excitation. Tachycardia episode was showing antidromic tachycardia with left bundle branch block morphology. Echocardiographic examination was showing Ebstein's anomaly with septal tricuspid leaflet displaced 24 mm apically. The right bundle branch block (RBBB) was concealed during sinus rhythm. The RBBB was revealed with ablation of right posterior atriofascicular accessory pathway.

2. Case report

A 14-year-old boy presented with repeated episodes of paroxysmal palpitations that occurred for the past 3 months. Palpitations were sudden in onset, not associated with dyspnea, angina, or giddiness. Clinical examination revealed...
evidence of tricuspid regurgitation. Echocardiographic examination revealed Ebstein’s anomaly of tricuspid valve (24 mm apical displacement of septal leaflet) with moderate tricuspid valve regurgitation (Fig. 1). The sinus rhythm surface ECG was not showing any pre-excitation (Fig. 2). The tachycardia episodes were usually broad QRS complex. The tachycardia ECG showed a LBBB-like pattern with superior axis and was terminated with injection verapamil.

The patient underwent electrophysiologic (EP) study to identify the mechanism of tachycardia and for radiofrequency ablation of accessory pathway. During incremental stimulation from the right atrium, there was gradual increase in pre-excitation and tachycardia was induced. The surface ECG during tachycardia suggested a regular pre-excited tachycardia using a right free wall accessory pathway. The intra-cardiac electrogams revealed an antidromic tachycardia, with the earliest retrograde activation in the AV node His bundle region (Fig. 3). Detailed mapping revealed an atriofascicular pathway located in the right posterolateral region. Mapping during sinus rhythm showed the shortest AV time to be 75 msec at the 8 o’clock position in the 45° left anterior oblique view. A discrete pathway potential was achieved at this site, and the A/V ratio was 1:10. Successful ablation was achieved at this site. During radiofrequency energy application, the 12 lead ECG changed to a complete RBBB (Figs. 4 and 5). Note the splintered QRS (best seen in lead V2), a classic finding in Ebstein’s anomaly of the tricuspid valve. No tachycardia was inducible after ablation. One month later, the patient remains free of tachycardia and the ECG shows a similar RBBB pattern.

3. Discussion

Catheter ablation has a lower success rate in patients with the anomaly than in those with structurally normal hearts, and the risk of recurrence is increased. Bizarre morphologies of the terminal QRS pattern result from infra-His conduction disturbance and abnormal activation of the atrialised right ventricle. The atrioventricular node may be compressed and the central fibrous body abnormally formed. The right bundle branch may be abnormal or show marked fibrosis (or both).

In most patients with atriofascicular pathways, there is no pre-excitation during sinus rhythm because of slow conduction in the pathway. However, with incremental stimulation of the right atrium and consequent physiologic delay in the AV node, atriofascicular pathway conduction becomes manifest. As highlighted in our patient, one can miss pre-excitation through a decremental conduction through atriofascicular accessory pathway during sinus rhythm. Despite the normal PR interval, HV interval, and QRS duration, there is pre-excitation as suggested by the local ventricular activation in the His bundle catheter. In sinus rhythm, antegrade conduction takes place over both the atriofascicular pathway and the AV node; because of the slow conduction in the atriofascicular pathway, the impulse courses further down the His bundle, activating the interventricular septum with a normal HV interval. This is suggested by the initial sharp “r” wave in V1. The subsequent conduction along the normal His Purkinje system would be expected to manifest as an R’ is abolished by the local pre-excitation by the atriofascicular pathway. The proof of this phenomenon is that after ablation in the right posterolateral region (far away from the right bundle branch), RBBB with splintering of QRS occurs. This morphology persists at 3-month follow-up. Thus, in essence, the clue of pre-excitation in a patient with Ebstein’s anomaly of the tricuspid valve is the absence of the expected RBBB during sinus rhythm.
Fig. 3 – Antidromic tachycardia, with the earliest retrograde activation in the AV node His bundle region.

Fig. 4 – During radiofrequency energy application, the 12 lead ECG changed to a complete right bundle branch block.
Con
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icts of interest

The authors have none to declare.

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Fig. 5 – Post-EP study and RF ablation 12 lead sinus rhythm ECG classical RBBB pattern with pre-excitation.