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An unusual WPW syndrome: What is the preexcitation variant?

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ABSTRACT

A 15-year-old female with WPW syndrome and normal heart underwent an electrophysiology study for paroxysmal palpitations and syncope. Intravenous adenosine produced an unexpected response of QRS changes and advanced AV block. During isoproteronol infusion, short-lasting and poorly tolerated wide QRS tachycardia was inducible, but pacing maneuvers were not feasible during tachycardia to determine its definitive mechanism. However, various electrophysiologic phenomena including adenosine response, junctional beats pattern, and multisite atrial pacing were helpful to overcome the diagnosis challenges. Finally, careful evaluation of tachycardia features and the comprehensive electrophysiology study were crucial to establish presence of unusual preexcitation variants, and thus to guide successful catheter ablation of the arrhythmic substrate.

Case presentation

A 15-year-old female with WPW and normal heart was referred to our center because of paroxysmal palpitations and syncope. Electrophysiology study (EPS) was performed, and multipolar diagnostic catheters were positioned in the coronary sinus (CS) and para-Hisian region. Her 12-lead ECG showed sinus rhythm with minimal preexcitation and occasional junctional beats (Fig. 1A). Baseline AH and HV intervals were 70 and 25 ms, respectively. Retrograde conduction was concentric and decremental. Incremental distal CS pacing showed progressive prolongation of the stimulus-delta interval with no change in QRS morphology, except for few beats with a wider QRS pattern just after the Wenckebach AV block. During atrial pacing, intravenous adenosine induced an advanced AV block (AVB) episode (Fig. 1B). Based on these observations, what is the preexcitation variant?

Discussion

The differential diagnosis of this preexcitation pattern may include: 1) a right-sided accessory pathway (AP) with enhanced nodal AV conduction. 2) a fasciculoventricular pathway (FVP). 3) a nodoventricular pathway (NVP). 4) an AV

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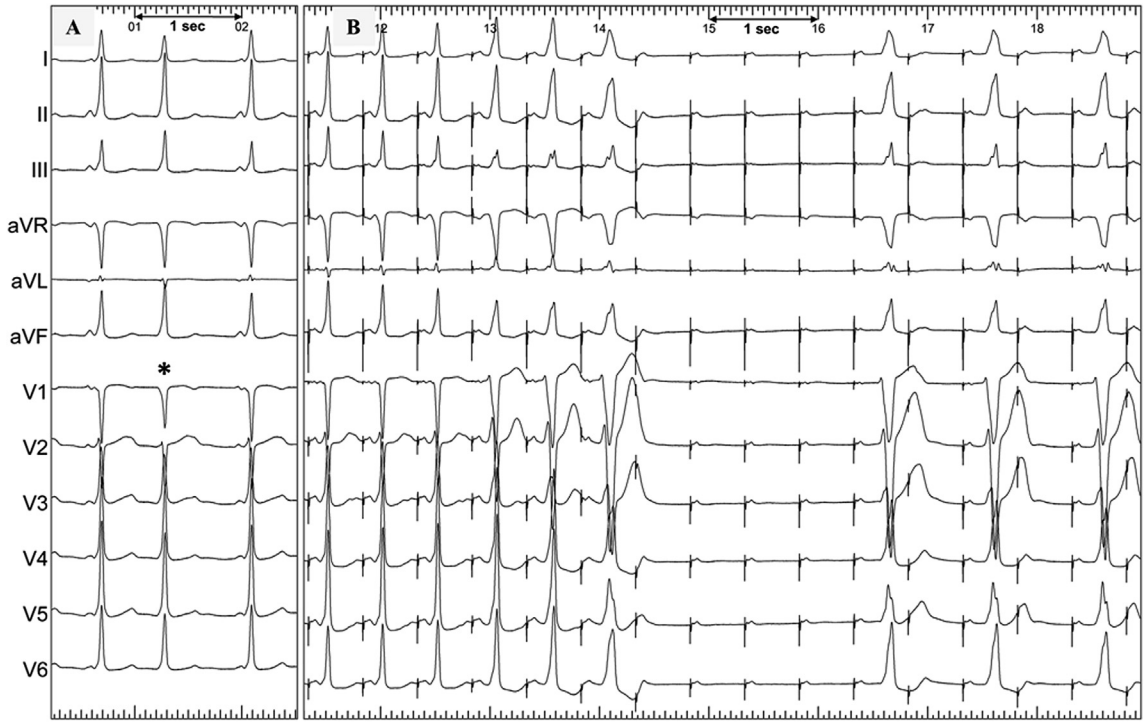

Fig. 1 – 12-lead ECG during sinus rhythm showing an occasional junctional beat (the asterisk, panel A), and during atrial pacing showing adenosine-induced AVB (panel B). See text for discussion.

Mahaim-like AP. 5) a combination of more than one of these AP types.

The preexcitation pattern (Fig. 1A) shows QS morphology in V1 and R/S transition between V2–V3, suggesting a right-sided AP. However, the latter AP typically presents with more evident preexcitation during sinus rhythm (P on Delta phenomenon) and a shorter HV interval. An exception might be the coexistence of accelerated or enhanced nodal AV conduction, which was not present in this case (a normal AH interval of 70 m).

Interestingly, occasional junctional beats replicated the preexcitation morphology (the asterisk in Fig. 1A) indicating the presence of an infra-atrial AP (typically an FVP) [1]. However, preexcited junctional beats could be observed in NVP if they originate proximally in the AV node [2]. Furthermore, this phenomenon does not exclude the coexistence of an AV-AP.

During atrial pacing, intravenous adenosine induced an advanced AVB followed by a phase of 2:1 AVB (Fig. 1B). This indicates the presence of AP(s) with nodal-like properties (Mahaim physiology). Noteworthy, P-delta prolongation and a wider QRS morphology were observed before the advanced AVB and also during the phase of 2:1 AVB. This change in QRS morphology/preexcitation degree is not compatible with a lone FVP which typically shows minimal and fixed preexcitation even with variable nodal conduction times, unless aberrant conduction is present. The wider QRS morphology was unlikely to be caused by a coexistent, bradycardia-dependent, left bundle branch block (LBBB) since it was not only observed during bradycardia (2:1 AVB) but also at a similar pacing cycle length preceding the advanced AVB (Fig. 1B). On the other hand, an NVP may demonstrate increased preexcitation and negative HV intervals since it bypasses only a portion of the AV node. Moreover, conduction delay over the left bundle, or preexcitation over an atrio/nodofascicular AP (inserting into the right bundle), usually present a typical LBBB pattern with rapid QRS onset and not the observed slow delta waves [3].

Accordingly, these electrocardiographic features of a minimal and right-sided preexcitation pattern, preexcited junctional beats, adenosine-responsiveness, and variable preexcitation degrees are best explained by the presence of an NVP or the coexistence of an FVP and a right-sided Mahaim-like AV-AP.

During isoproterenol infusion, short-lasting episodes of a wide QRS tachycardia were inducible by atrial overdrive pacing (independently on AH prolongation), or occasionally after premature ventricular beats (Fig. 2A). Unfortunately, no diagnostic pacing maneuvers were applicable during tachycardia since it often terminated spontaneously (~10 s), and it was also associated with significant hemodynamic instability due to its high rate (240–250 bpm). QRS morphology during tachycardia was similar to the wider QRS morphology observed during the adenosine-test or rapid atrial pacing. Intracardiac recordings showed *constant* 1:1 AV rapport, concentric atrial activation (identical to that during RV pacing), and the AV and VA intervals were 168 and 80 ms, respectively (Fig. 2B). No His potentials preceding the QRS were recorded, being apparently buried within the local ventricular electrograms (i.e., a negative HV interval). Interestingly, spontaneous termination of tachycardia showed V–V