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CASE REPORT

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Transient high-degree right bundle branch block masking the type 1 Brugada ECG pattern associated with possible transient early repolarization syndrome

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Abstract

The Brugada syndrome (BrS) was the last electrocardiographic syndrome described in the 20th century. The initial description included right bundle branch block (RBBB), persistent ST-segment elevation in the right precordial leads, absence of structural heart disease, and propensity to unexplained syncope and/or sudden death mainly during nocturnal rest. Currently, we know that the first three components are not constant or true since RBBB is present in only 28% of cases, the ST-segment elevation is dynamic, at times absent, and there are discrete structural changes in the right ventricular outflow tract. Additionally, the presence of RBBB can hide the typical type 1 Brugada ECG pattern. We present a very unusual case of spontaneous transient RBBB that revealed a hidden type 1 Brugada ECG pattern that could be seen in the beat with normal ventricular conduction.

KEYWORDS

Brugada syndrome, masked Brugada pattern, transient right bundle branch block

1 | CASE REPORT

A 32-year-old Asian male (Thai) presented three recurrent syncopal episodes of unknown origin in the previous week. The events were accompanied by urinary incontinence in supine position during nocturnal rest, convulsions and “agonal-type” respirations during the last event.

Family background: A cousin and an uncle had died suddenly at night, labeled as Lai Tai.

Physical examination: unremarkable.

The admission ECG showed complete right bundle branch block (RBBB) (Figure 1).

Normal transthoracic echocardiography.

A Holter recording showed non-RBBB beats with the typical type 1 Brugada ECG pattern (Figure 2). The patient has nine points according to the recently validated Shanghai scoring system for Brugada syndrome (BrS) diagnosis (Kawada et al., 2018) (>3.5 points: probable and/or definite BrS).

2 | DISCUSSION

We consider the type 1 Brugada ECG pattern was hidden or "masked" when there was concomitant high-degree RBBB and J-point elevation <2 mm. According to Wada et al., this occurs in approximately 8% of cases with persistent RBBB associated with the BrS. In these cases, Chiale's maneuver eliminates the block with pacing; the right ventricular apex is paced at an atrioventricular (AV) interval that results fused ventricular activation, nullifying the effect of the RBBB-induced delay, that would obscure the Brugada pattern (Crinion & Baranchuk, 2019). This results in the appearance of the Brugada type 1 (diagnostic) or type 2 (nondiagnostic) ECG pattern in about 3% of cases. In the Wada et al study, there was no prognostic difference between the BrS masked by RBBB and nonmasked BrS (Wada et al., 2015). In a large series, the presence of complete RBBB was registered in only 28% of cases (Maury et al., 2013). The ST-segment elevation in the right precordial leads is dynamic or at times absent

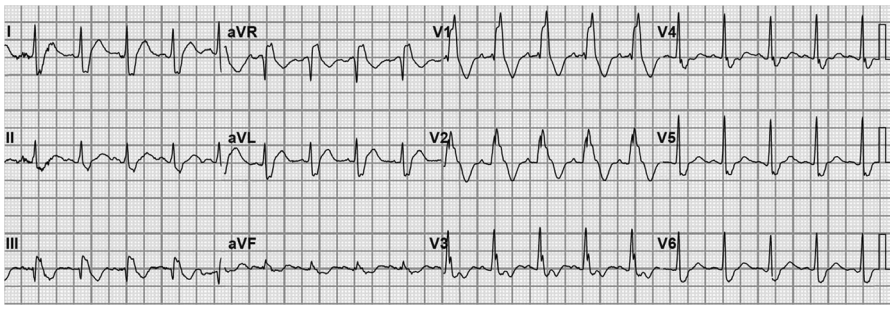

FIGURE 1 The 12-lead ECG. Sinus rhythm, heart rate 75 bpm, P duration 110 ms, bimodal shape in II, PR interval 160 ms, QRS duration 175 ms, QRS axis + 130°, triphasic QRS pattern in right precordial leads, rsR' in V1, prolonged R-wave peak time in V1-2, J-wave insinuation in V2-3, broad final S wave in lateral leads (I, aVL, V5-V6), wide final R wave in aVR, and ventricular repolarization (ST/T) with opposite direction to the terminal deflection of the QRS complex. Conclusion: CRBBB

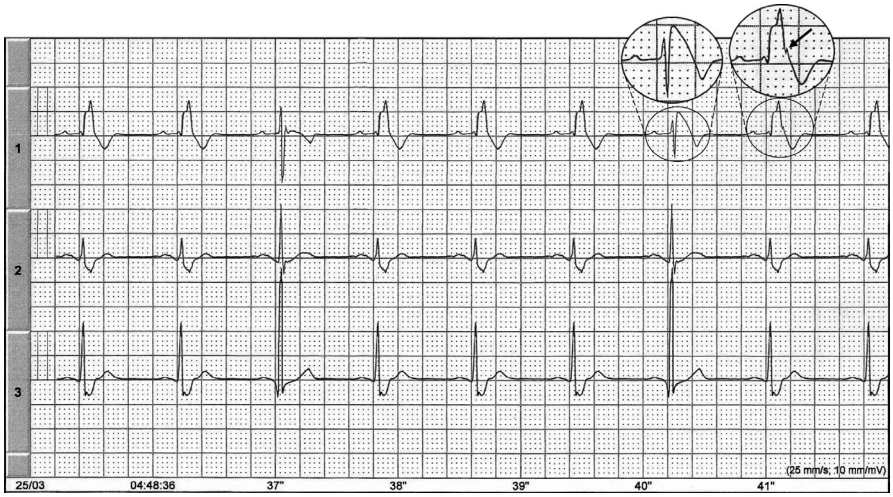

FIGURE 2 Holter monitoring during early morning. The Holter monitoring shows complete right bundle branch block (RBBB) in the first and second beats. The third beat shows a sudden narrow QRS complex without changes in the heart rate. The fourth to sixth beats show reappearance of complete RBBB. The seventh beat shows typical type 1 Brugada ECG pattern (coved type) when complete RBBB disappeared. The eighth beat again shows complete RBBB with a notched J-wave insinuation (arrow). Conclusion: Transient spontaneous CRBBB that masks the type 1 Brugada ECG pattern. Additionally, the eighth beat shows a notched variant of the J wave. Note: There were no changes in heart rate or PR interval in the non-RBBB beats. The probable explanation proposed in the literature is supernormal conduction. The phenomenon is much more common than previously thought, particularly in the presence of certain clinical conditions. On the cellular level, this occurs if the ventricular rate is just beyond the effective refractory period of the septal myocardium, while at the same time, the phase 4 voltage allows for supernormal conduction without the aid of the bundle branches or Purkinje system

as the Brugada brothers described in their first description (Brugada & Brugada, 1991, 1992).

The type 1 Brugada ECG pattern can be masked by the presence of RBBB and exposed by resolution of the block using a pharmacological approach with intravenous ajmaline (Rolf, Haverkamp, & Eckardt, 2005), by pacing according to the "Chiale maneuver" (Aizawa et al., 2013; Chiale, Garro, Fernandez, & Elizari, 2012; Perez-Riera et al., 2017), by placing the right precordial ECG leads at an upper position (Aizawa et al., 2013) or spontaneously (Tomita et al., 2012). The present case is to our knowledge the first case in the literature, where Holter monitoring revealed a masked or hidden Brugada ECG. The beat with a normal QRS duration was not associated with shorter RR and PR intervals.

Zhang et al performed noninvasive ECG imaging to compare intraventricular conduction in patients with BrS and RBBB (Zhang et al., 2015). In RBBB patients, activation of the entire right ventricle was delayed, while in contrast, delayed activation was confined to the right ventricular outflow tract of BrS patients. They also described differences in repolarization, but provided no mechanistic explanation for the fact that RBBB may mask the Brugada ECG pattern.

Although we have no definite explanation for the fact that RBBB masked the Brugada ECG pattern in this patient case, we speculate that the subjacent mechanism could be supernormal excitability and conduction. This electrophysiologic phenomenon is much more common than previously thought. The presence of a rather long relative refractory period of specialized fibers would facilitate the existence of a well-developed supernormal phase and eventually supernormal conduction. In fact, supernormal excitability implies that conduction of the cardiac impulse may improve during such a period (Elizari, Schmidberg, Atienza, Paredes, & Chiale, 2014), particularly when the

phenomenon is systematically searched for in patients with advanced AV block or intermittent conduction in the His-Purkinje system or in the case of accessory pathways. One of the main unresolved issues is to elucidate the exact mechanism of this almost forgotten electrophysiologic curiousness. Another theoretical mechanism could be the so-called AV-gap phenomenon, which occurs when the effective refractory period of a distal site is longer than the functional refractory period of a proximal site, and when closely coupled stimuli are delayed enough at the proximal site to allow distal site recovery. Two variants of gap phenomenon have been described. In type 1 gap, the distal site of block is distal to the His bundle and the proximal site in the AV node. In type 2 gap, both the proximal and the distal sites of conduction block are within the ventricular specialized conduction system (Wu, Denes, Dhingra, & Rosen, 1974).

CONFLICT OF INTEREST

None.

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