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CASE REPORT

Incessant slow bundle branch reentrant ventricular tachycardia in a young patient with left ventricular noncompaction

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KEYWORDS
Left ventricular noncompaction;
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Electrophysiologic study;
Automatic implantable cardioverter-defibrillator

Abstract A 15-year-old girl was admitted to the cardiology outpatient clinic due to mild palpitations and documented incessant slow ventricular tachycardia (VT) with left bundle branch block (LBBB) pattern. The baseline electrocardiogram revealed first-degree atrioventricular block and intraventricular conduction defect. Transthoracic echocardiography showed prominent trabeculae and intertrabecular recesses suggesting left ventricular noncompaction (LVNC), which was confirmed by cardiac magnetic resonance imaging. During electrophysiological study, a sustained bundle branch reentrant VT with LBBB pattern and cycle length of 480 ms, similar to the clinical tachycardia, was easily and reproducibly inducible. As there was considerable risk of need for chronic ventricular pacing following right bundle ablation, no ablation was attempted and a cardioverter-defibrillator was implanted. To the best of our knowledge, no case reports of BBR-VT as the first manifestation of LVNC have been published. Furthermore, this is an extremely rare presentation of BBR-VT, which is usually a highly malignant arrhythmia.

PALAVRAS-CHAVE
Ventrículo esquerdo não-compactado;
Taquicardia ventricular por reentrada de ramo;
Estudo electrofisiológico;

Taquicardia ventricular por reentrada de ramo lenta e incessante em adolescente com ventrículo esquerdo não-compactado

Resumo Uma jovem de quinze anos de idade foi observada em consulta externa de Cardiologia por palpitações ligeiras e documentação de taquicardia ventricular (TV) lenta e incessante com padrão de bloqueio de ramo esquerdo (BRE). O electrocardiograma (ECG) basal revelou bloqueio auriculoventricular (BAV) de primeiro grau e perturbação da condução intraventricular. Um ecocardiograma transtorácico documentou trabeculação proeminente e recessos intertrabeculares, alterações sugestivas de ventrículo esquerdo não-compactado (VENC), diagnóstico confirmado por ressonância magnética cardíaca. No estudo electrofisiológico, uma taquicardia

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Cardioversor-desfibrilhador implantável

ventricular sustentada por reentrada de ramo, com padrão de BRE e ciclo de base de 480 ms, semelhante à taquicardia clínica, foi repetidamente induzida. Considerando o risco elevado de necessidade de *pacing* ventricular crónico em caso de ablação do ramo direito (BAV de primeiro grau e BRE no ECG basal e intervalo HV 100 ms no estudo electrofisiológico), não foi efetuado qualquer procedimento ablativo e um cardioversor-desfibrilhador foi implantado. Até ao momento atual, nenhum caso de TV por reentrada de ramo como primeira manifestação de VENC foi publicado. O caso descrito revela uma apresentação extremamente atípica deste tipo de TV, que habitualmente é rápida e maligna.

Introduction

Bundle branch reentrant ventricular tachycardia (BBR-VT), an uncommon form of macroreentrant tachycardia, generally occurs in the context of dilated cardiomyopathy, previous valve surgery or other cardiac conditions with underlying His-Purkinje system (HPS) disease. This case is a very unusual presentation of BBR-VT in a young patient with isolated left ventricular noncompaction (LVNC). Although our patient presented with HPS disease allowing initiation of this arrhythmia, it is rare for BBR-VT to be the first manifestation of isolated LVNC. Furthermore, BBR-VT is a highly malignant arrhythmia, yet our patient was almost asymptomatic due to the surprisingly long cycle length of the ventricular tachycardia (VT) and despite its unusual incessancy.

Case report

A 15-year-old girl was referred to our arrhythmology department for mild palpitations and documented incessant VT (hemodynamically stable VT lasting hours). She was otherwise healthy, with no relevant medical history and no family history of significant cardiomyopathy or sudden cardiac death (SCD). Her palpitations were not related to effort and were persistent but otherwise extremely well tolerated. She denied precordial pain, dizziness, or presyncope/syncope. A previous electrocardiogram (ECG) had revealed wide-QRS tachycardia (WCT) at 115 beats per minute (bpm).

Investigations

The patient was examined shortly after being referred, and denied any symptoms, including palpitations. The physical exam was unremarkable. The first ECG showed a VT with left bundle branch block (LBBB) pattern and superior axis at 118 bpm. A transthoracic echocardiogram revealed a normal-sized LV and preserved overall systolic function, hypertrabeculation of the LV posterior and lateral walls and intertrabecular recesses communicating with the LV cavity as demonstrated by color Doppler flow, suggestive of LVNC, which was confirmed by cardiac magnetic resonance imaging (Figure 1). Subsequent ECGs alternated between sinus rhythm with intraventricular conduction abnormalities and first-degree atrioventricular (AV) block and slow VT with LBBB pattern and superior axis (Figure 2).

An exercise stress test was stopped at 3:58 because of the sudden induction of a well tolerated yet sustained slow wide QRS tachycardia with right bundle branch (RBB) and left posterior fascicular block patterns. During most of the recovery time, an incomplete RBB block pattern with right axis deviation was seen, as at the beginning of the test.

An electrophysiological study (EPS) was performed. The baseline ECG revealed sinus rhythm, intraventricular conduction defects (QRS 122 ms) and significant PR prolongation (296 ms) (Figure 3). A standard protocol using 6-F diagnostic electrophysiology catheters was followed. Two quadripolar catheters were placed in the high right atrium, His bundle and right ventricle as required, and a decapolar catheter in the coronary sinus. The programmed ventricular stimulation protocol included three drive-cycle lengths (CL) and two ventricular extrastimuli while pacing from the right ventricular apex. A 125 bpm-rate monomorphic sustained VT was reproducibly inducible with a single extrastimulus 360 ms after an eight-beat drive-cycle length of 600 ms. It had a LBBB pattern, superior axis and a clear right bundle deflection preceding each ventricular complex, suggesting the RBB was part of the circuit (Figure 4). A short postpacing

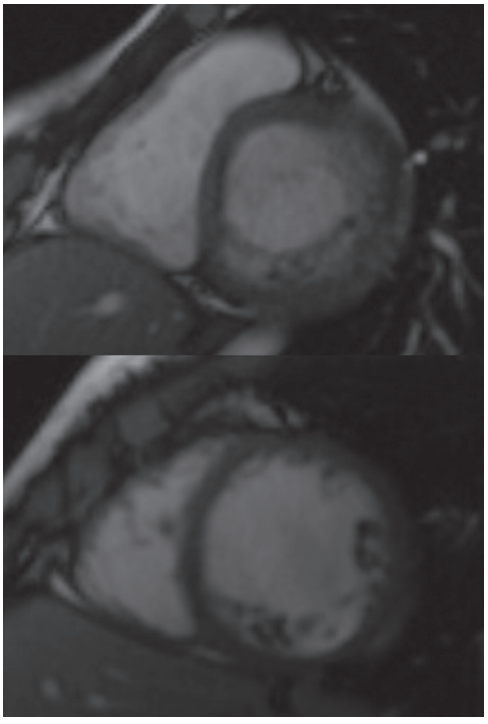

Figure 1 Cardiac magnetic resonance images suggesting left ventricular noncompaction.