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CASE REPORT

Left bundle branch block, atrioventricular block, torsade de pointes and long QT syndrome: Is this too much for a rare cardiomyopathy?☆

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Abstract Left ventricular noncompaction (LVNC) is now recognized as a distinct form of cardiomyopathy with a clinical presentation and natural history of its own. Common manifestations of LVNC include heart failure, ventricular arrhythmias and embolic events, but serious atrioventricular conduction disturbances are rarely reported in the literature. The authors describe the case of a 40-year-old woman who went to the emergency department due to syncope. The ECG revealed left bundle branch block (LBBB) and 2:1 atrioventricular block (AVB) and the patient was admitted for pacemaker implantation. During hospitalization she developed torsade de pointes and complete AVB with increased QTc. The echocardiogram showed images compatible with LVNC. This case provides additional evidence that LVNC may be complicated by 2:1 (or complete) AVB, intraventricular conduction disturbances (LBBB) and repolarization abnormalities (long QT). This combination of electrocardiographic changes has rarely been reported in the literature. We describe a series of affected patients, focusing on electrocardiographic characteristics.

PALAVRAS-CHAVE
Ventrículo esquerdo não compactado;
Bloqueio auriculoventricular;
Torsade de pointes

Bloqueio completo de ramo esquerdo, bloqueio auriculoventricular, *torsade de pointes* e QT longo: será demasiado para uma rara miocardiopatia?

Resumo O ventrículo esquerdo não compactado (VENC) tem sido reconhecido como uma forma distinta de miocardiopatia com uma apresentação clínica e história natural próprias. As manifestações comuns de VENC são insuficiência cardíaca, arritmias ventriculares e eventos embólicos. A presença de perturbações da condução auriculoventricular de maior gravidade encontra-se raramente descrita na literatura. Os autores descrevem o caso clínico de uma

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paciente com 40 anos de idade, que recorreu ao Serviço de Urgência por síncope. O ECG revelou bloqueio completo de ramo esquerdo do feixe de His e bloqueio auriculoventricular (BAV) 2:1, tendo sido internada para implantação de *pacemaker*. Durante o internamento, desenvolve fenómenos de *torsade de pointes* e de BAV completo com aumento do intervalo QTc. O ecocardiograma revelou imagens compatíveis com VENC. Com este caso, os autores fornecem evidência adicional de que a presença de VENC pode ser complicada por BAV 2:1 (ou completo), perturbações da condução intraventricular (BCRE) e da repolarização (QT longo), alterações eletrocardiográficas concomitantes pouco descritas na literatura. Descreve-se uma série de pacientes afetados, com foco nas características eletrocardiográficas.

Introduction

Left ventricular noncompaction (LVNC) is a rare cardiomyopathy originally described in children[1,2] but whose phenotypic characteristics are increasingly documented in adults.[3–7] The most common symptoms of LVNC are heart failure (HF), ventricular arrhythmias and embolic events, but atrioventricular conduction disturbances are rarely reported in the literature.[8–10]

Case report

A 40-year-old woman went to the emergency department due to syncope during a meal, without prodrome and with spontaneous recovery, associated with progressive fatigue for around a month. She reported no chest pain or dyspnea. Personal history included complete left bundle branch block (LBBB) detected on a routine electrocardiogram (ECG) as part of a medical check-up at work three months before. There was no family history of sudden death and she was not taking any medication.

Physical examination showed the patient to be eupneic with no signs of respiratory difficulty. Cardiac auscultation revealed rhythmic sounds and no murmurs but bradycardia (40 bpm); pulmonary auscultation detected no abnormalities. Blood pressure was 100/75 mmHg. The rest of the physical examination was normal.

Laboratory tests, including blood and biochemical analysis, showed no relevant alterations and biomarkers of myocardial necrosis were normal.

A 12-lead ECG revealed second-degree 2:1 atrioventricular block (AVB) with ventricular response of ±45 bpm and LBBB, and the patient was accordingly admitted for pacemaker implantation (Figure 1).

During hospitalization she developed cardiac arrest due to torsade de pointes (TdP) (Figure 2), reverted by cardiac massage. The ECG revealed 2:1 AVB (with a period of complete AVB), heart rate of ±45 bpm and increased QTc interval (Figure 3).

Echocardiography performed in the context of monitoring in the cardiac care unit showed a normal-size left ventricle with mildly impaired global systolic function (ejection fraction ±45%) and images compatible with LVNC at the apex (pronounced trabeculations and ratio of trabeculated to non-trabeculated layers >2, with blood flow on color Doppler) (Figures 4 and 5). Coronary angiography revealed no coronary disease.

A dual-chamber cardioverter-defibrillator was accordingly implanted for secondary prevention of sudden death.

The patient was discharged clinically stable and is currently well with no signs of HF. She is pacing dependent but no shocks have been detected. Screening of first-degree relatives showed normal ECG but echocardiography in her children showed images compatible with inferior and lateral LVNC (Figure 6), although high-resolution ECG showed no abnormalities. The results of the patient's genetic study are awaited.

Discussion

Patients with LVNC usually have impaired left ventricular systolic function and generally present with symptoms of HF.[1,2,4] Other associated cardiac alterations include endocardial thrombi, leading to systemic embolism, and arrhythmias. The high incidence of arrhythmias in LVNC is not explained by a known morphological abnormality, but it has been suggested that myocardial ischemia may play an important role in the development of systolic dysfunction and arrhythmias in these patients, as is seen in ischemic heart disease.[11]A variety of rhythm disturbances have been reported in LVNC, of which ventricular arrhythmias are the most common; they may be refractory to medical treatment and can be fatal.[1–4,11–15]

The literature shows that 88–94% of LVNC patients have alterations on the baseline ECG.[2,3,16] In a series by Oechslin et al.,[4] 94% of the 34 adults studied presented abnormal ECGs and 56% had intraventricular conduction defects, most of them identified in patients who had died. In a series by Steffel et al.[16] (n=78), only 13% of the patients had a normal ECG, while two (3%) had complete AVB block, 19% had LBBB, and 50% had prolonged QTc.

Wolff–Parkinson–White syndrome (WPW) and associated tachycardias have also been described in association with LVNC.[1,2,14,17] Ichida et al.[2] reported WPW syndrome in four of 27 patients with LVNC, and explained this association as a failed regression of developmental embryologic atrioventricular anatomical and electrical continuity during