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CASE REPORT

Two potentially fatal surprises in the preoperative assessment of an asymptomatic young adult☆

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KEYWORDS
Isolated left ventricular non-compaction;
Preoperative care;
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Abstract Isolated left ventricular non-compaction is a rare disease classified as a primary genetic cardiomyopathy and is characterized by heart failure, systemic embolism and ventricular arrhythmias. The diagnosis is established by Doppler echocardiography. We report the case of an asymptomatic young adult, with no history of heart disease, who underwent preoperative assessment for low-risk orthopedic surgery. The electrocardiogram showed left bundle branch block, which prompted further investigation with Doppler echocardiography, cardiac computed tomography angiography and cardiac magnetic resonance imaging. A diagnosis of isolated left ventricular non-compaction and pulmonary embolism was made. Some aspects of preoperative assessment in low-risk surgical patients are discussed.

PALAVRAS-CHAVE
Miocárdio ventricular isolado não compactado;
Cuidados pré-operatórios;
Embolia pulmonar

Duas surpresas potencialmente fatais em avaliação pré-operatória de adulto jovem assintomático

Resumo A cardiomiopatia não compactada isolada do ventrículo é uma doença rara, classificada como uma cardiomiopatia genética primária. A doença é caracterizada por insuficiência cardíaca, embolia sistémica e arritmias ventriculares. O diagnóstico é estabelecido pelo ecodopplercardiograma. Relata-se o caso de adulto jovem assintomático, sem história prévia de cardiopatia, que realizou avaliação pré-operatória para cirurgia ortopédica de baixo risco. Eletrocardiograma apresentou bloqueio do ramo esquerdo, que suscitou investigação complementar com ecodopplercardiograma, angiotomografia do tórax e ressonância cardíaca.

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Concluiu-se pelo diagnóstico de cardiopatia não compactada isolada do ventrículo e embolia pulmonar. Discutem-se aspetos da avaliação pré-operatória em pacientes de baixo risco cirúrgico.

Introduction

Isolated left ventricular non-compaction (LVNC) was described for the first time by Chin et al. in 1990.[1] It is a rare disease classified as a primary genetic cardiomyopathy by the American Heart Association.[2] LVNC is attributed to intrauterine arrest of compaction of the meshwork of the fetal myocardium.[3]

The prevalence of LVNC in the general population is unknown. Existing information is mainly derived from patients undergoing Doppler echocardiography. A Swiss review identified 34 cases, corresponding to 0.014% of all echocardiograms performed over a period of 15 years.[4] The prevalence of LVNC in patients with heart failure (HF) has been estimated as 3–4%.[5] It appears to affect predominantly males, as shown in the four main series on LVNC patients.[1,4,6,7]

A family history of LVNC is described in 12–50% of case reports.[8] Autosomal dominant inheritance is more common than X-linked or autosomal recessive inheritance.[9] There is growing recognition of a considerable overlap in the genetic loci involved in the pathogenesis of the main cardiomyopathies. Phenotypic expression of different cardiomyopathies may be found in the same patient, including LVNC and hypertrophic cardiomyopathy,[10] although LVNC is more commonly associated with congenital heart disease and Wolff-Parkinson-White syndrome.[11]

Preoperative assessment provides an opportunity for the physician to identify silent conditions and to optimize treatment of known cardiovascular disease. In recent years, in an attempt to avoid wasting resources on unnecessary medical exams, medical societies have established protocols for requests for preoperative exams. An editorial by the Cleveland Clinic,[12] which is in agreement with the II Preoperative Guideline of the Brazilian Society of Cardiology (SBC),[13] questions the value of the electrocardiogram (ECG) in preoperative assessment of asymptomatic individuals scheduled for low-risk surgery.

Case report

A 35-year-old man with partial rupture of the left Achilles tendon caused by a sports accident was referred by the surgeon for preoperative assessment. He had no cardiovascular symptoms or history of heart disease, was not taking prescription medication and did not smoke. Physical examination revealed blood pressure of 135/78 mmHg and heart rate of 89 bpm and no relevant alterations. He had a walking boot on his left leg. The chest X-ray was normal, and complete blood panel, blood glucose, plasma creatinine, urea and international normalized ratio were within reference values. The ECG showed complete left bundle branch block (LBBB). Tissue Doppler imaging (TDI) revealed left ventricular (LV) ejection fraction (LVEF) of 61%, left atrial volume index of 22 ml/m^2, and LV mass index of 94.9 g/m^2. Alterations in LV relaxation (E′=6 cm/s) and increased filling pressures (E/E′=16) were observed. TDI showed hypertrabeculation of the mid anterolateral wall and apical region (Figure 1). As the differential diagnosis of LVNC includes other forms of cardiomyopathy, the patient was referred for computed tomography coronary angiography, which showed signs of bilateral pulmonary embolism (Figure 2). D-dimers were 2310 ng/ml. Doppler echocardiography of the venous system of the left leg demonstrated hypoechogenic material in the popliteal and posterior tibial veins, partially obstructing the lumen. Cardiac magnetic resonance imaging (Figure 3) showed LV hypertrophy and marked mid-apical subendocardial trabeculation (estimated at 22% of total myocardial mass), myocardial late enhancement indicative of mid anteroseptal and basal mid inferoseptal myocardial fibrosis, and moderate global LV dysfunction, with LVEF estimated at 38%, confirming the diagnosis of LVNC.

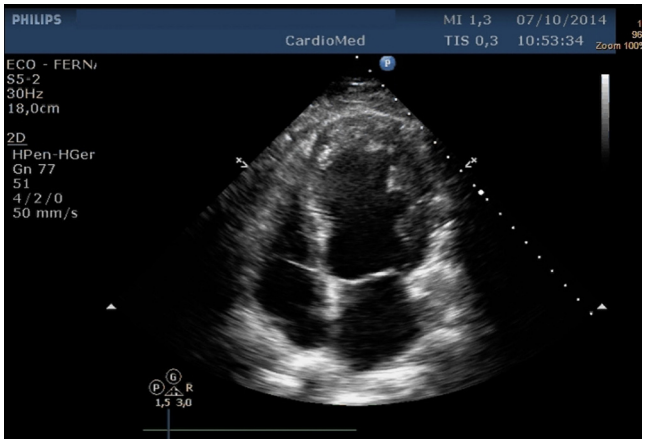

Figure 1 Echocardiogram, apical 4-chamber view, showing global left ventricular systolic dysfunction located mainly in the mid anterolateral and apical walls. Non-compaction is also more prominent in the subendocardial myocardium than in the subepicardial region.

Outpatient Holter 24-hour electrocardiographic monitoring showed an episode of paroxysmal supraventricular tachycardia and ventricular extrasystoles (Figure 4), but the patient remained asymptomatic.