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CASE REPORT

Isolated left ventricular noncompaction causing refractory heart failure☆

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KEYWORDS
Cardiomiopathy;
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Abstract Left ventricular noncompaction is a rare congenital anomaly characterized by excessive left ventricular trabeculation, deep intertrabecular recesses and a thin compacted layer due to the arrest of compaction of myocardial fibers during embryonic development. We report the case of a young patient with isolated left ventricular noncompaction, leading to refractory heart failure that required extracorporeal membrane oxygenation followed by emergency heart transplantation.

PALAVRAS-CHAVE
Cardiomiopatia;
Disfunção ventricular;
Transplante cardíaco

Miocárdio não compactado isolado evoluindo para insuficiência cardíaca refratária

Resumo A não compactação do miocárdio é uma anomalia congênita rara, definida por excessiva trabeculação do ventrículo esquerdo, profundos recessos intertrabeculares e uma camada compactada fina, devido à interrupção do processo de compactação das fibras miocárdicas durante a fase embriogênica. Relatamos um caso de uma paciente jovem com miocárdio não compactado isolado evoluindo para insuficiência cardíaca refratária, com necessidade de uso de oxigenação por membrana extracorpórea seguido de transplante cardíaco de emergência.

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Introduction

Left ventricular noncompaction (LVNC) is a rare congenital anomaly characterized by excessive left ventricular (LV) trabeculation, deep intertrabecular recesses and a thin compacted layer due to the arrest of compaction of myocardial fibers during embryonic development. It can also be acquired through cardiac remodeling, mainly in young athletes or in pregnancy and sickle cell anemia.[1] LVNC is usually associated with other congenital cardiac malformations.[1,2] The cause of isolated LVNC is unknown and no factor has yet been identified to explain the arrest of myocardial compaction.[1,3]

LVNC is associated with a wide range of clinical manifestations; while some patients may be asymptomatic, symptoms can appear at any age, including heart failure (HF), thromboembolic phenomena and cardiac arrhythmias; the disease has a poor prognosis. LVNC is found in 0.81 per 100 000 infants/year and 0.12 per 100 000 children/year, and has a prevalence of 0.014% in adults.[1,3] There are no specific histological findings, other than fibrosis.[4]

We describe the case of a patient with isolated LVNC who developed refractory HF and required emergency heart transplantation.

Case report

TSA, a 14-year-old white girl, born and resident in the municipality of Barueri, São Paulo, Brazil and a handball player, whose father had died six months previously due to idiopathic dilated cardiomyopathy (no anatomopathological study was performed), sought emergency medical assistance at the Hospital Dante Pazzanese de Cardiologia due to worsening dyspnea over the previous 15 days. On admission she reported dyspnea at rest. Physical examination showed her to be in reasonable general condition, conscious and oriented. Pulmonary auscultation revealed rales in the lower third of both lungs and cardiac auscultation showed a gallop rhythm (B3), with no murmurs; other observations were lower limb edema (3+/4+), blood pressure 100/70 mmHg, and liver palpable 7 cm below the right costal margin. The 12-lead electrocardiogram (Figure 1A) showed sinus rhythm, narrow QRS complex, low-voltage QRS in the frontal plane, left atrial overload, signs of right atrial overload, diffuse ventricular repolarization abnormalities and right axis deviation (−170°) but no criteria for left posteroinferior hemiblock. The chest X-ray revealed a significantly enlarged cardiac silhouette (Figure 1B), while echocardiography (Figure 2A) showed left atrium 38 mm, considerably enlarged right atrium, LV dilatation (60 mm×68 mm), LV ejection fraction 25%, superior vena cava 14 mm, tricuspid regurgitation with annular dilatation (30 mm), and moderate right ventricular systolic dysfunction. A rounded hyperdense mass measuring 17 mm×14 mm was observed in the left atrium, indicating a thrombus, a small pericardial effusion, and moderate pulmonary hypertension (40 mmHg). Cardiac magnetic resonance imaging (MRI) (Figure 2B) revealed severe biventricular dysfunction with thickened myocardium, mainly in the LV anterior wall and apex, with a ratio of noncompacted to compacted layers of 5.5, but no late enhancement suggestive of myocardial fibrosis. LV size was 67 mm×71 mm, LV ejection fraction 8%, and right ventricular ejection fraction 2%. No coronary angiography, Holter ECG or electrophysiological studies were performed. Laboratory tests showed normal cardiac enzymes, NT-proBNP of 6850 pg/ml, and negative serology.

Dobutamine 5 μg/kg/min was begun, together with systemic anticoagulation with full-dose enoxaparin. Pulmonary vascular resistance was calculated at 1.7 Woods units using a Swan-Ganz catheter.

The patient developed low cardiac output, atrial fibrillation with high ventricular response (heart rate 160 bpm), worsening level of consciousness, hypotension and seizures, requiring orotracheal intubation and synchronized electrical cardioversion. Extracorporeal membrane oxygenation (ECMO) was begun. The patient developed multiple organ failure (acute renal failure requiring dialysis and liver failure with changes in transaminases and coagulation parameters, thrombocytopenia and elevated bilirubin). Blood cultures were negative. The patient was put on the priority list for heart transplantation.

After 72 h of ECMO, she underwent heart transplantation, with no complications during the procedure. Acute renal failure persisted, requiring hemodialysis for three days, followed by improvement in the patient's coagulopathy and organ dysfunction.

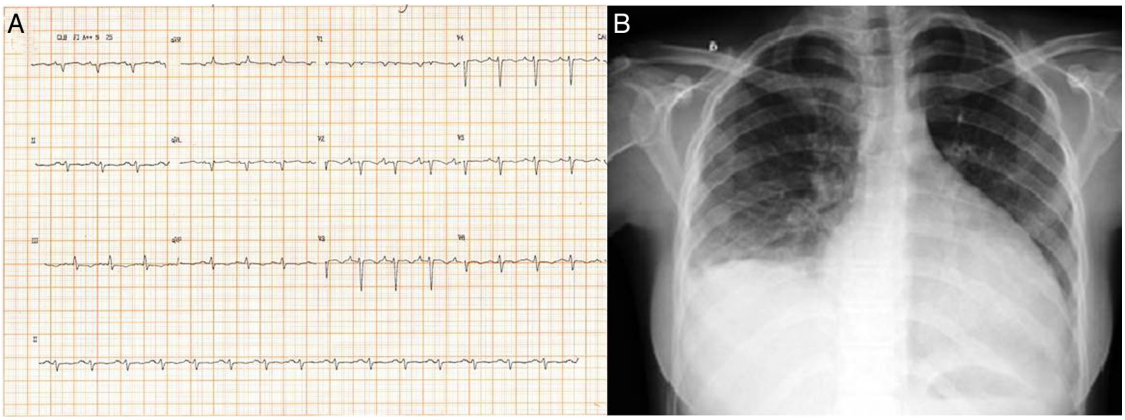

Figure 1 12-lead electrocardiogram (A) and chest X-ray in anteroposterior view (B).