Sawtooth cardiomyopathy: A rare cause of heart failure

Miocardiopatia em dentes de serra: uma causa rara de insuficiência cardíaca

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Sawtooth cardiomyopathy is a rare form of left ventricular dysplasia of which only three cases have been reported in the literature. Its strange phenotype – projections of compacted myocardium – has led some authors to consider it a variant of left ventricular (LV) non-compaction.

We present the case of a male patient, now aged 21 years, referred to our department at one month of life due to congestive heart failure. There was no relevant family history of cardiovascular disease. The initial echocardiogram revealed dilatation of the left chambers and severe systolic dysfunction. Etiological investigation excluded structural heart disease and infectious, metabolic, genetic or neuromuscular causes.

Following the initial episode, the patient remained under regular surveillance in the pediatric cardiology clinic.

At age 16 years, the echocardiogram showed numerous echodense myocardial projections with a sawtooth morphology in the LV inferior and lateral walls and posterior septum, protruding into the chamber (Figure 1A and B).

For further clarification of the morphological and functional features, cardiac magnetic resonance imaging was performed, which revealed numerous projections of apparently compacted myocardium originating in the LV inferior and lateral walls and on the left surface of the interventricular septum (Figure 1C–E). Global systolic function was mildly impaired, with areas of hypokinesia in a non-coronary distribution pattern in the interventricular septum and the LV apex. No late enhancement was seen after gadolinium administration.
Figure 1  Two-dimensional transthoracic echocardiography in modified 4-chamber view showing echodense myocardial projections with sawtooth morphology (A and B); cardiac magnetic resonance imaging in 2-chamber long-axis view showing projections of apparently compacted myocardium originating from the left ventricular inferior wall (C); cardiac magnetic resonance imaging in axial 4-chamber view showing projections of apparently compacted myocardium originating from the left ventricular inferior wall and the left side of the interventricular septum (D and E).

Although the phenotype was similar to that of LV non-compaction, the case did not meet the imaging criteria for the latter entity.

Conflicts of interest

The authors have no conflicts of interest to declare.