Young male with long-term impaired ejection fraction – endocardial fibroelastosis: The importance of cardiac magnetic resonance

Varón joven con fracción de eyección deteriorada a largo plazo - fibroelastosis endocárdica: la importancia de la resonancia magnética cardíaca

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A 32-year-old male was referred to the cardiology outpatient clinic of our institution for follow-up of dilated cardiomyopathy. At the age of 3 months, he had presented significant dyspnea and an enlarged cardiac silhouette had been discovered at the chest X-ray. Echocardiography had showed the left atrium (LA) and left ventricle (LV) dilation with severely depressed LV ejection fraction (LVEF). A diagnosis of dilated cardiomyopathy was made at that time. He had been treated transiently with loop diuretics and permanently with angiotensin-converting enzyme inhibitors and beta-blockers. He was stabilized and evolved in New York Heart Association functional Class II/IV reaching the adulthood.

His last transthoracic echocardiography showed markedly LA and LV dilation, moderately reduced (43%) LVEF and trabeculated areas at the LV apical segments. Coronary artery disease was ruled out by a normal coronary angiography.

With the suspicion of non-compaction cardiomyopathy, a cardiac magnetic resonance (CMR) was requested. The cine steady-state free precession images demonstrated enlarged left cavities with moderately depressed LVEF, normal interventricular wall thickness, and right and left apical ventricular hypertrabeculation, although not fulfilling non-compaction criteria (Fig. 1).

At late gadolinium images, a diffused subendocardial late gadolinium hyperenhancement involving the basal and midsegments of the LV in a circumferential pattern was present (Fig. 2). At that time, etiological diagnosis of endocardial fibroelastosis (EFE) was performed.

EFE is a rare condition, whose primary form manifests most frequently during the 1st month of life. It consists of a thickening of the endocardium by collagen and elastic fibers¹. Its secondary form is mostly related to hypereosinophilia and lymphoproliferative disorders². Primary EFE is mostly manifested with dilation and systolic impairment of the LV. However, there are primary EFE cases with features of other conditions such as restrictive cardiomyopathy³. A great number of cases have poor prognosis and require heart transplantation during childhood, some of them misdiagnosed mostly as dilated cardiomyopathy⁴.

There are not unified diagnostic criteria for this disease, what renders the diagnosis difficult during the adult age. A high grade of suspicion, comprehensive clinical history, and histological evidence of fibrosis either by biopsy or at the CMR are mandatory.

In our particular case, clinical history with onset at 3 months of age and diffused EFE evidenced at CMR enabled this diagnosis.

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Figure 1. Cardiac magnetic resonance: cine steady-state free precession. Left atrium and left ventricle (LV) dilation with RV and LV trabeculation. (A) Four chamber view. (B) Midventricular short-axis view.

Figure 2. Cardiac magnetic resonance: late gadolinium enhancement sequence. Diffused subendocardial hyperenhancement. (A) Midventricular short-axis view. (B) Four chamber view.

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Authors do not have any conflicts of interest.

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Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

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