Atypical cardiac magnetic resonance findings in a case of suspected endomyocardial fibrosis

Eduard Claver1*, José González-Costello1, Río Aguilar2, Guillem Pons-Llado3, and Àngel Cequier1

1Heart Disease Institute, Bellvitge University Hospital, IDIBELL, University of Barcelona, L’Hospitalet de Llobregat, C Feixa Llarga s/n, 08028 Barcelona, Spain; 2Department of Cardiology, La Princesa Hospital, Madrid, Spain; and 3Department of Cardiac Imaging, Clínica Creu Blanca, Barcelona, Spain

* Corresponding author. Tel: +34 932607500; fax: +34 932607807, Email: eclaver@bellvitgehospital.cat

Endomyocardial fibrosis (EMF) is the leading cause of restrictive cardiomyopathy worldwide. Cardiac magnetic resonance (CMR) with assessment of myocardial late gadolinium enhancement (LGE) allows the diagnosis of the disease by means of the characterization of the fibrous tissue deposited on the endocardium. We sought to discuss the CMR findings in a young man from Equatorial Guinea with a history of exertional dyspnoea and ascites. On chest radiography (Panel A), an enlarged cardiac silhouette is observed due to right atrial dilatation. Echocardiography: non-dilated left ventricle with preserved ejection fraction and non-dilated right ventricle (RV) with a prominent mass in the apex obliterating partially the cavity. Eosinophilic blood count was within the normal range. No endomyocardial biopsy of the RV mass was obtained.

CMR, surprisingly, did not corroborate the diagnosis. On cine sequences, a localized apical RV mass is observed (Panel B; see Supplementary data, Video S1), with signal intensity similar to the myocardium. On basal first-pass perfusion sequences (Panel C; see Supplementary data, Video S2), the mass shows an increase in the intensity of the signal, similar to the adjacent myocardium, consistent with vascularized tissue, contrary to what would be expected in the case of fibrotic or thrombotic tissue. Finally, on LGE sequences (Panel D), no hyperenhanced signal indicative of the presence of myocardial contrast is found on the RV apical endocardium; this ruling out the presence of fibrotic tissue.

In summary, despite clinical and echocardiographic signs suggestive of RV EMF, the CMR study did not support this diagnosis. Instead, CMR findings were consistent with RV apical obliterative myocardial hypertrophy.

Supplementary data are available at European Heart Journal – Cardiovascular Imaging online.

Panel A. Chest radiography: enlargement of the cardiac silhouette due to right atrial dilatation.
Panel B. Cardiac magnetic resonance, four-chamber view, cine sequences: localized apical RV mass (asterisk) with signal intensity similar to the myocardium. Severe right atrial enlargement is observed. LV, left ventricle; LA, left atria.
Panel C. Cardiac magnetic resonance, four-chamber view, basal gadolinium first-pass perfusion sequences. The signal intensity of the RV mass (asterisk) is similar to the adjacent myocardium.
Panel D. On late gadolinium sequences, no hyperenhanced signal is observed on the RV apical mass (asterisk).

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