Different focal delayed gadolinium-enhancement patterns using cardiac magnetic resonance in a case of diffuse giant cell myocarditis

Arshid Azarine1,* , Romain Guillemain2, and Patrick Bruneval3

1Department of Cardio-Vascular Radiology, Hôpital Européen Georges Pompidou, 20-21 rue Leblanc, Paris 75015, France; 2Thoracic Transplant Unit, Hôpital Européen Georges Pompidou, 20-21 rue Leblanc, Paris 75015, France; and 3Department of Pathology, Hôpital Européen Georges Pompidou, 20-21 rue Leblanc, Paris 75015, France

* Corresponding author. Fax: +33 156 092 311, Email: aazarine@free.fr

A 17-year-old man was admitted for new onset of fatigue with dyspnoea. He did not present fever or a recent history of flu-like symptoms. The results of the physical examination and ECG were unremarkable except for a sinus tachycardia at 116 b.p.m. Echocardiography demonstrated severe global hypokinesia of both ventricles with left ventricular ejection fraction (LVEF) of 10%. Laboratory tests revealed a troponin I level of 0.02 ng/mL (normal <0.015 ng/L), C-reactive protein of 0.015 ng/L, creatine kinase of 54 UI/L (normal <15 UI/L). Cardiac magnetic resonance (CMR) confirmed severe global dysfunction of both ventricles with LVEF of 12% and RVEF of 10%. Gadolinium delayed enhancement (DE) imaging demonstrated three different types of focal linear DE: mid wall and sub-endocardial ‘right-sided’ DE of the septum (black arrow), sub-epicardial DE of the lateral wall (white arrow) and sub-endocardial ‘ischaemic-like’ DE of the lateral–basal wall (Panel A, short-axis view, Panel B, four-chamber view, LV: left ventricle). The patient condition worsened rapidly and he underwent emergency heart transplantation. Pathology of the explanted heart revealed a diffuse giant cell myocarditis (GCM) occurring predominantly in the delayed enhanced areas as demonstrated by CMR (haematoxylin–eosin-stained specimen, original × 40; Panel C, septal specimen; Panel D, lateral LV wall specimen). Higher magnification of the most infiltrated areas demonstrated foci of dense lymphocytic infiltrates with numerous giant cells without evidence of ischaemic myocardial injury, particularly, in the endocardial area of the lateral basal wall (haematoxylin–eosin stain, original × 400, Panel E, midwall area of the septum; Panel F, endocardial area of the lateral basal wall). A 2 year follow-up by routine endomyocardial biopsy has shown no recurrence of GCM or rejection.

On these DE images, the signal of the diffusely infiltrated myocardium by GCM was null, the different patterns of focal hyper-enhanced areas being relevant for the most infiltrated areas when compared with histology.

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