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VIABILITY

Cardiomyopathy in Becker muscular dystrophy—does regional fibrosis mimic infarction?

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We present a case of a 39-year-old man with Becker muscular dystrophy and severe congestive cardiac failure. Cardiac magnetic resonance imaging revealed subendocardial late gadolinium enhancement, similar to that seen in myocardial infarction. He had no risk factors for atherosclerotic coronary artery disease and coronary angiography was normal. We propose that regional subendocardial myocardial fibrosis which has been described histologically in the cardiomyopathy associated with Becker muscular dystrophy may resemble previous infarction at contrast enhanced cardiac magnetic resonance imaging.

Key Words: Cardiomyopathy; Heart failure; Magnetic resonance imaging; Becker muscular dystrophy

1. Case report

A 39-year-old man with Becker muscular dystrophy (BMD) presented with NYHA IV congestive cardiac failure. His EKG showed sinus rhythm, left axis deviation, intraventricular conduction delay and non specific anterior ST flattening. Transthoracic echocardiogram demonstrated a severely dilated, hypokinetic left ventricle (left ventricular end diastolic diameter 8 cm) in keeping with dilated cardiomyopathy secondary to BMD. Although no wall motion abnormality was obvious at echocardiography, ventricular function was so poor that further analysis of regional myocardial function was difficult. Serological investigations including virology and iron studies were normal.

Further assessment was undertaken with cardiac magnetic resonance imaging (CMR). Imaging was performed on a 1.5 Tesla scanner (Siemens Sonata, Erlangen, Germany). A fast-imaging with steady-state precession (true FISP) sequence was used to acquire cine images in long axis planes (vertical long axis, horizontal long axis, left ventricular outflow tract) followed by sequential short axis LV cine loops (8 mm slice thickness, 2 mm gap between slices) from the atrioventicular ring to the apex. Further images were acquired 10 minutes after a peripheral intravenous bolus of Gd-DTPA (0.2 mmol/kg) using a breath-hold segmented turbo fast low angle shot (FLASH) inversion-recovery sequence, using identical slice positions as the cine images.

CMR showed severe biventricular systolic dysfunction with wall thinning and akinesis of both the basal- and mid-anterior and anterolateral left ventricular segments (1) (end diastolic wall thickness 2.7 mm, end systolic thickness 3.4 mm; Fig. 1). Ejection fraction measured by CMR was 15%, end diastolic volume 298 mL, end systolic volume 253 mL, end diastolic diameter by CMR 7.92 cm). These corresponding segments also demonstrated extensive subendocardial late gadolinium enhancement in keeping with images described in previous myocardial infarction (2) (Fig. 2). Approximately 50–75% of the wall thickness was involved in this area of enhancement. Late gadolinium imaging also confirmed the presence of mid wall septal gadolinium enhancement, as has been previously described in both BMD (3) and idiopathic dilated cardiomyopathy (4). Cardiac catheterization demonstrated entirely normal, non obstructed coronary arteries. Although the CMR images are highly suggestive of previous myocardial infarction, there was no clinical or angiographic evidence to support this (Fig. 3).

The patient has been treated with a diuretic, beta blocker, angiotensin converting enzyme inhibitor, anticoagulation and cardiac resynchronization therapy and has made good clinical progress, currently functioning at NYHA class II.

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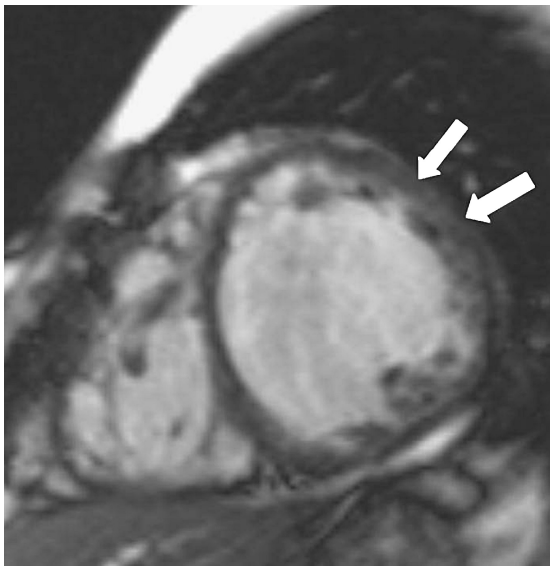

Figure 1. Short axis end diastolic view, from the cine images (fast imaging with steady state precession sequence) of the mid cavity of the left ventricle demonstrating dilated end diastolic cavity with thinning of the lateral wall (arrowed). Wall thickness of the mid anterolateral segment is 2.7 mm compared to 4.2, 4.3, 6.1, 4.5 and 4.2 mm for the mid-anterior, anteroseptal, inferoseptal, inferior and inferolateral segments respectively.

2. Discussion

BMD is a rare X-linked recessive disorder with a prevalence estimated at 2.38 per 100,000 live births characterized by progressive skeletal muscle weakness and cardiomyopathy.

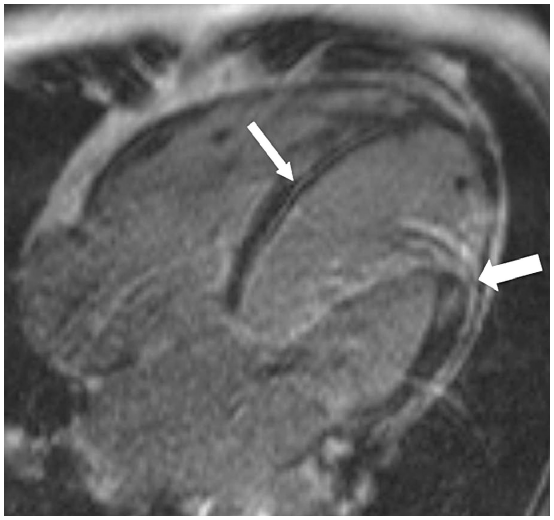

Figure 2. Horizontal long axis (4-chamber) view demonstrating mid wall late gadolinium hyperenhancement in the septum (thin arrow) with subendocardial hyperenhancement in the lateral wall (thick arrow). Unfortunately, motion artefact is also present on this image, seen in the basal portion of the left ventricular cavity. The patient was struggling to maintain breath holds for the duration of acquisition of this image.

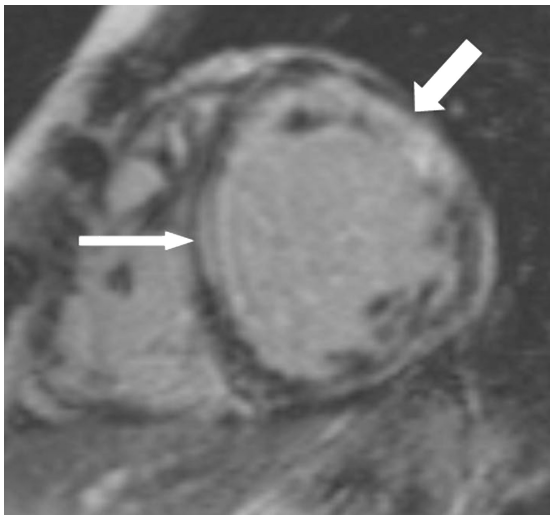

Figure 3. Short axis end diastolic view of the apical left ventricle demonstrating subendocardial late gadolinium enhancement (thick arrow) in the thinned lateral wall, with mid wall late gadolinium enhancement in the septum (thin arrow) corresponding with Fig. 2.

The gene involved codes for a 427 kDa protein called dystrophin (5). Although cardiac symptoms preceding skeletal muscle involvement is rare, cardiac involvement may be the presenting feature (3), typically presenting in the third decade. Cardiac myocyte dystrophin deficiency leads to myocardial cell necrosis causing replacement of the myocardium with connective tissue or fat. Cardiac involvement is invariable in BMD and is the cause of death in approximately 50% of patients with the condition (6). EKG findings such as non specific ST changes, varying degrees of atrioventricular block, pathological Q waves have been described, progressing to left bundle branch block and ventricular arrhythmias in advanced cases (7). Screening for cardiac involvement in BMD with EKG and echocardiography has been recommended, and although no large prospective studies of pharmacological therapy in the condition exist, favorable response to modern heart failure treatment has been reported (8), while cardiac transplantation may have a role in selected cases (9).

Pathological studies of the explanted hearts in BMD have demonstrated normal coronary arteries with thickened left ventricular myocardium, while histological examination of endomyocardial biopsies have shown interstitial and endocardial fibrosis. No characteristic histological features distinguish the cardiomyopathy associated with BMD from other cardiomyopathies, without the use of immunohistochemistry (10). These findings offer a histopathological correlate of our clinical, angiographic and CMR results. CMR offers an attractive screening tool for affected individuals and may be able to identify patients at high risk of ventricular arrhythmia based on the presence of either myocardial fibrosis or fatty change. However as our case illustrates, caution should be exerted in interpreting subendocardial late

gadolinium hyperenhancement as being due to myocardial infarction in the presence of severe left ventricular dysfunction and a diagnosis of heritable cardiomyopathy, such as BMD should be considered.

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