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Noncompaction of the ventricular myocardium mimicking ischemic cardiomyopathy

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A 68-year-old woman was admitted to our hospital because of left ventricular failure. Myocardial perfusion single-photon emission computed tomography demonstrated a non-reversible perfusion defect in the anterolateral left ventricular segments and reduced ejection fraction, findings consisted with ischemic cardiomyopathy accompanied by lateral wall infarction. Coronary angiogram was normal, but the left ventriculogram showed prominent trabeculations in the apical and anterolateral segments. The left ventricular ejection fraction was 28%. Cine magnetic resonance imaging demonstrated prominent trabeculations and deep intertrabecular recesses, findings consistent with noncompaction of the ventricular myocardium. Myocardial hypoperfusion or necrosis in the noncompacted myocardium may be the cause of myocardial damage and possibly the basis of left ventricular failure.

Key words: isolated noncompaction of the ventricular myocardium, single-photon emission computed tomography

INTRODUCTION

ISOLATED noncompaction of the ventricular myocardium (INVM) is a rare disorder characterized by numerous, prominent ventricular trabeculations and deep intertrabecular recesses.[1] The pathogenesis of INVM is believed to be an arrest in endomyocardial morphogenesis.[1] Prognosis is apparently grim, with a high mortality and morbidity from heart failure, ventricular arrhythmias and systemic embolization.[1,2] The reason for depressed left ventricular function in INVM is unknown, but regional myocardial hypoperfusion has been observed by positron emission tomography[3] and gadolinium-enhanced first pass magnetic resonance imaging.[4,5] We describe a patient with INVM in whom myocardial perfusion single-photon emission computed tomography (SPECT) revealed myocardial necrosis.

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CASE REPORT

A 68-year old woman with a long history of diabetes mellitus was admitted to our hospital complaining of shortness of breath on effort. A chest X-ray revealed cardiomegaly (the cardiothoracic ratio was 63%) and mild pulmonary congestion. Her 12-lead electrocardiogram showed complete right bundle branch block and low voltage, but there were no abnormal Q waves. Laboratory examinations disclosed increased plasma brain natriuretic peptide (1180 pg/m*l*) and norepinephrin (905 pg/m*l*) levels. She had no family history of cardiomyopathy or sudden death. Echocardiography was non-diagnostic because of poor image quality, but it showed a dilated and hypocontractile left ventricle. With a provisional diagnosis of ischemic cardiomyopathy, she underwent rest ^{201}Tl (111 MBq)/stress ^{99m}Tc-tetrofosmin (740 MBq) separate acquisition dual-isotope myocardial perfusion single photon emission computed tomography (SPECT) using a dual detector gamma camera (E-CAM, Siemens Medical Solutions, Erlangen, Germany). Pharmacological stress was performed using adenosine triphosphate (0.16 mg/kg/min).[6] It revealed a moderate sized, non-reversible

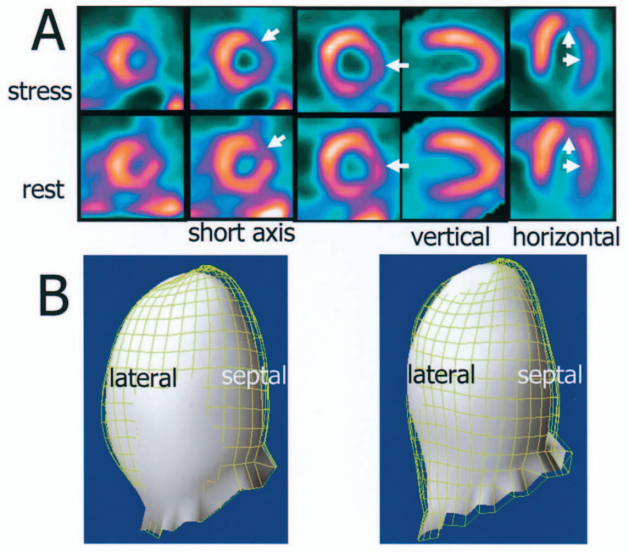

Fig. 1 (A) Single-photon emission computed tomographic images showing non-reversible perfusion defect in the anterolateral segments (*arrows*), (B) Quantitative gated SPECT showing akinesis in the lateral segments after stress (*left*) and at rest (*right*).

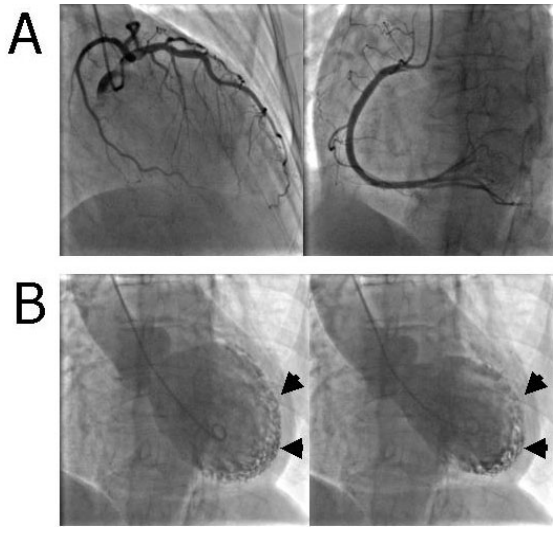

Fig. 2 (A) Left (*left*) and right (*right*) coronary angiograms showing normal coronary arteries. (B) Left ventriculogram from the frontal projection showing a honeycomb-like appearance in the lateral segments.

myocardial perfusion defect in the base to distal anterolateral segments and reversible defect in the inferolateral segments (Fig. 1A). Quantitative ECG-gated SPECT showed a markedly decreased left ventricular ejection fraction (18%) with akinesis in the lateral wall (Fig. 1B). The end-diastolic volume at rest was 93 m*l* and the transient ischemic dilation ratio was 1.08. These findings were consistent with ischemic cardiomyopathy with myocardial infarction in the territory of the left circumflex artery.

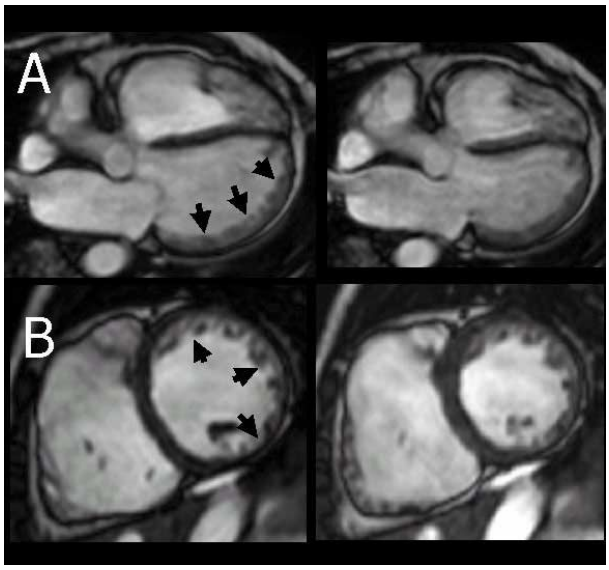

Fig. 3 Cine magnetic resonance images showing a double-layered appearance in the lateral wall (*arrows*) on the 4-chamber view (A) and marked trabeculations and deep intratrabecular recessess in the anterior, lateral and inferior segments (*arrows*) on the short-axis view. Left = diastole, right = systole.

Coronary angiography was performed and it revealed normal coronary arteries (Fig. 2A). Left ventriculography demonstrated a honeycomb-like appearance in the lateral segments (Fig. 2B). The left ventricular ejection fraction was 28%. An endomyocardial biopsy from the left ventricle showed mild endocardial thickening and interstitial fibrosis, but no evidence of inflammation. ECG-gated cine magnetic resonance imaging was performed with an Intra Achieva (1.5 T, Philips Medical Systems, Best, Netherlands) using steady-state coherent sequence, and showed a double-layered appearance with noncompacted endomyocardial segment and compacted epimyocardial segment in the lateral wall on the 4-chamber view (Fig 3A). Marked trabeculations and deep intratrabecular recesses in the anterior, lateral and inferior segments were observed on the short-axis view (Fig. 3B). There was severe hypokinesis in the lateral segments.

Thus, the diagnosis of INVM was established and the patient underwent medical treatment including beta blocker, furosemide and warfarin administration. She has been uneventful during a 6-month follow-up period.

DISCUSSION

Although INVM has been known for nearly 2 decades, it is still an "unclassified" cardiomyopathy by the World Health Organization classification of the cardiomyopathies. Consequently, the diagnosis of INVM is often

missed because of lack of knowledge, despite its unfavorable prognosis. Diagnostic criteria for this disorder are 1) the absence of coexisting cardiac abnormalities, 2) two-layer structure with a compacted thin epicardial band and much thicker noncompacted endocardial layer of prominent trabecular meshwork with deep endomyocardial spaces, resulting in a maximal end systolic ratio of noncompacted to compacted layers of > 2[2,7] and 3) evidence of deep intratrabecular recesses by color Doppler,[2,6] or magnetic resonance imaging.[3,4,8] Major complications of INVM are ventricular dysfunction, ventricular arrhythmia and systemic embolism.[1,2] The mechanisms of left ventricular dysfunction are not well understood. The pathological studies demonstrated a continuous layer of endothelium from the ventricular cavity into the recesses without coronary communications to the ventricular cavity[1] and evidence of increased subendomyocardial fibrosis within areas of noncompacted myocardium.[1,9] Reduced myocardial perfusion in the noncompacted endomyocardial segment has been observed by positron emission tomography[3] and contrast enhanced first pass magnetic resonance imaging.[4,5] In addition, myocardial necrosis in the noncompacted area has been documented previously by delayed enhancement on contrast-enhanced magnetic resonance imaging.[10–12] Although the left ventricular apex and inferolateral segments are the most affected regions of noncompaction,[1,2] and the areas with decreased myocardial perfusion and necrosis correspond to these regions, the vast majority of adult patients present with diffuse left ventricular hypokinesis without regional wall motion abnormalities, presumably due to ventricular remodeling.[2] In these patients with global left ventricular dysfunction, differentiation of INVM from other forms of cardiomyopathy such as dilated cardiomyopathy and ischemic cardiomyopathy is obscure, especially when echocardiography is non-diagnostic. At present, magnetic resonance imaging is the most reliable method for the diagnosis of INVM because it allows not only visualization of endomyocardial texture with high spatial resolution, but also assessment of myocardial perfusion abnormalities.

In our case, global left ventricular dysfunction as well as regional wall motion abnormality in the lateral wall was detected by quantitative ECG-gated SPECT, a finding consistent with that obtained by left ventriculogram and magnetic resonance imaging.

In conclusion, regional myocardial perfusion and wall motion abnormalities may occur in patients with INVM, which may lead to misinterpretation of myocardial perfusion SPECT and QGS wall motion analysis. Being aware of this rare disorder is of utmost importance for the differential diagnosis from coronary artery disease.

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