ISOLATED NONCOMPACTION OF THE VENTRICULAR MYOCARDIUM

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SUMMARY

A case of isolated noncompaction of the ventricular myocardium with MRI findings is presented. MRI features and a review of published reports of noncompaction of the ventricular myocardium are briefly discussed.

Key words: magnetic resonance imaging: isolated noncompaction ventricular myocardium

CASE REPORT

A 59-year-old-female had a cardiac arrest in July 2002 and was resuscitated. She was admitted to the hospital for further investigation which revealed increased creatinine kinase(CK) and a raised Troponin. An electrocardiogram performed demonstrated T-wave changes in the anterolateral leads suggestive of an infarct.

The patient initially presented in 1994 with a history of palpitation of 12 months duration. Electrocardiogram (ECG) showed evidence of atrial fibrillation and the patient was commenced on Warfarin. She had a past history of hypertension for a period of 10 years. In June 2002, one month prior to her cardiac arrest , the patient presented with chest pain and had a stress test which was normal. There was no evidence of ischaemic changes and her ECG showed atrial fibrillation with normal complexes.

The left ventriculogram demonstrated sponge -like appearance of the non-compacted ventricular wall and diverticular configuration of the anterolateral wall (Fig1). There was mild hypokinesia of the noncompacted ventricular wall and there was apical akinesia. Coronary arteriogram demonstrated normal left main coronary artery, left anterior descending artery and circumflex artery. There was minor irregularities of the dominant right coronary artery with a 30% stenosis of the proximal third of the vessel. A probable diagnosis of noncompaction of the left ventricle with apical akinesia and diverticulae of the anterolateral wall and mild coronary artery disease was made.

Magnetic resonance imaging (MRI) was performed to confirm the diagnosis. Electrocardiography gated half fourier single shot turbo spin-echo (HASTE) sequences in the axial, coronal and sagittal planes, and T1-weighted fast spin-echo(FSE) and T2-weighted fast spin-echo (FSE) in the short-axis view and axial T2-weighted fast spin-echo and cine gradient-echo (GRE) sequences in the short-axis and four chamber view were performed.

Cine MRI demonstrated a thickened inner endocardial myocardium of prominent trabeculation

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with deep intertrabecular recesses and a thin compacted epicardial myocardium.(Fig2A,B,C) IVNC involved the anterior, lateral and inferior segments from mid-ventricular level to the apex of the left ventricle. The largest trabeculation measured 2.6cm from the epicardial surface to the peak of the trabeculation, and 0.8 cm between the epicardial surface and trough of the trabecular recess. The ratio of the noncompacted/compacted myocardium was >2 which confirmed the diagnosis of IVNC. Cine gradient-echo sequences demonstrated hypokinesia of the left ventricular wall and there was a small focal area of thinned myocardium with akinesia of the apical lateral segment (segment16).

The T2-weighted sequences demonstrated a focal area of high signal intensity within the left ventricular cavity at the apex which was probably related to slow flow adjacent to a akinetic segment.

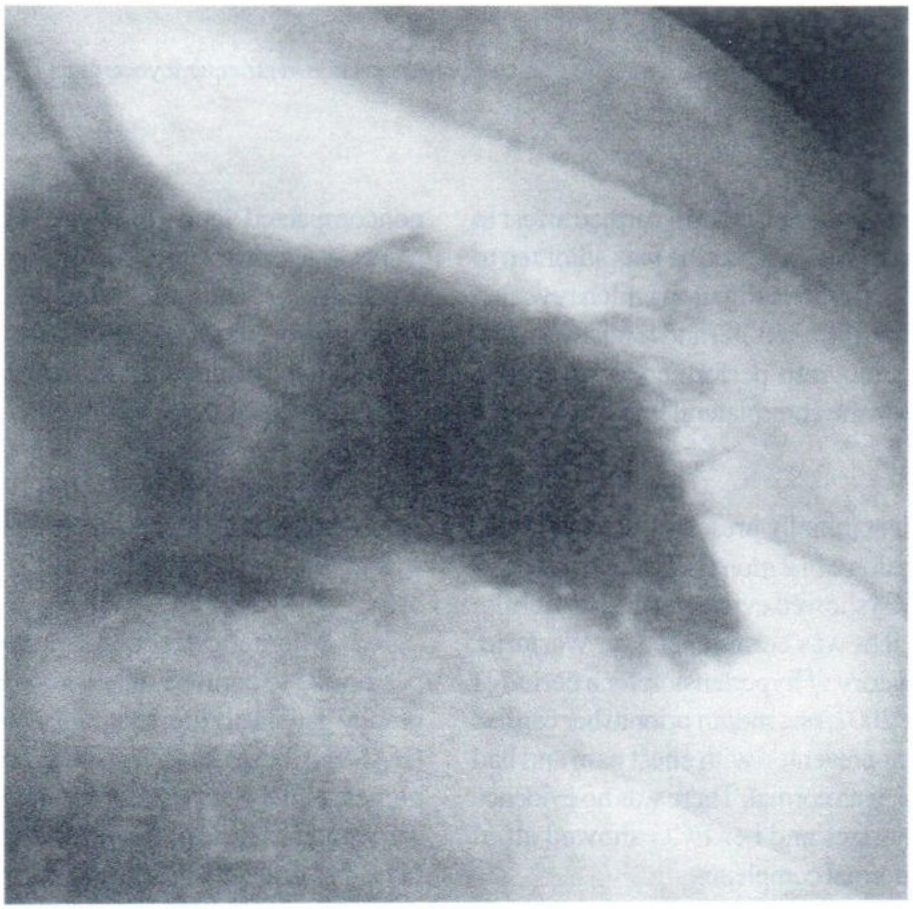
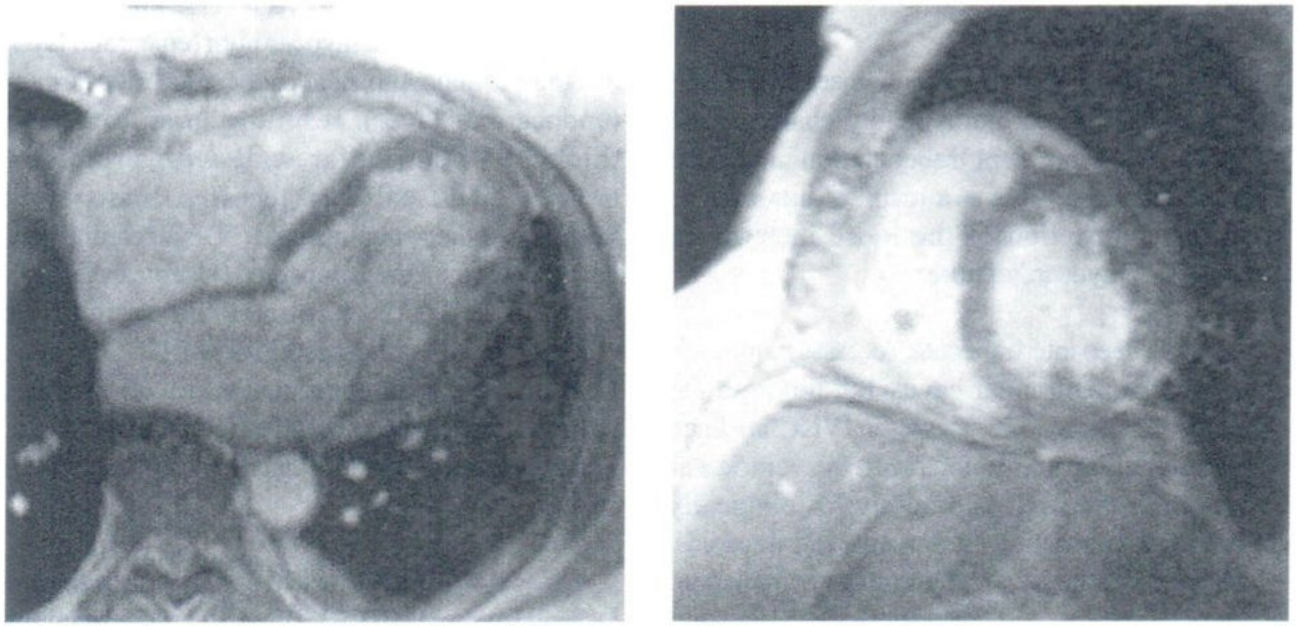
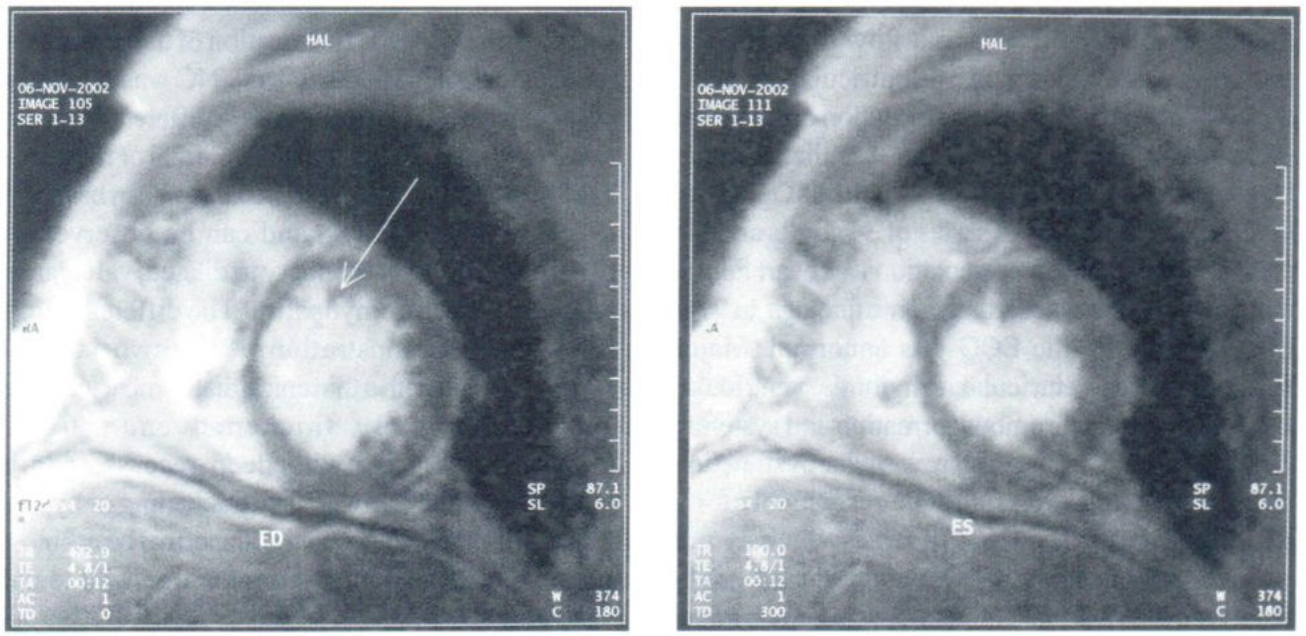


Fig.1 Left ventriculogram demonstrates sponge-like appearance of the non-compacted ventricular wall and diverticular configuration of the anterolateral wall.



A B

Fig.2 **A)** axial GRE –image at the level of the mitral valve **B)** mid-ventricular short-axis view



(a) (b)

Fig.2 **C)** short-axis view at the ventricular apex **(a)** end-diastolic **(b)** and end-systolic image demonstrates prominent trabeculations on the endo cardial surface of the left ventricle with deep intertrabecular recess filled with blood from the ventricular cavity.

DISCUSSION

Isolated ventricular noncompaction (IVNC) is a rare congenital cardiomyopathy characterised by numerous, prominent trabeculation and deep intertrabecular recesses as a result of intrauterine arrest of compaction of the loose interwoven meshwork of myocardial fibres.

The deep intertrabecular recesses communicate with the ventricular cavity but not with the coronary circulation. The recesses in IVNC are lined with endothelium continuous with the ventricular endocardial endothelium and are histologically different from persisting intramyocardial sinusoids, which are in continuity with the coronary circulation.

The ventricular noncompaction may occur in the left, right or both ventricles[1] and may be associated with other congenital cardiac malformation., including anomalous origin of the left coronary artery from the pulmonary trunk[2] and obstructive lesions of the left or right ventricular out flow tract, such as pulmonary atresia with intact ventricular septum.[3] Isolated left ventricular noncompaction occurs even more rarely.[4] The disorder may be familial and may be associated with facial dysmorphism.[4] Oechslin[5] et al reported the largest series of 34 adults and the most common clinical presentation was heart failure and characteristic echocardiographic findings. In 94% of their patients, the ECG was abnormal. Major cardiac risks are ventricular arrhythmia, ventricular hypokinesia with thrombus formation and systemic embolization. Coronary microvascular dysfunction associated with IVNC might be responsible for a decreased coronary flow reserve which is not confined to noncompacted segments, but extends to most segments with wall motion dysfuction and mural thrombus formation within the deep intertrabecular recesses abnormalities.[6]

The typical echocardiographic findings of noncompacted myocardium in IVNC is characterised by myocardium with extremely thickened, hypokinetic segments consisting of two layers. There is thin, compacted epicardial myocardium (epicardial layer) and thicker noncompacted endocardial myocardium (endocardial layer), resulting in an extremely thickened ventricular wall with prominent trabeculations and deep recesses. End-systolic thickness of the noncompacted (N) endocardial layer was thicker than the compacted (C) epicardial layer (ratio of $N/C \geq 2$). A ratio of noncompacted/ compacted ≥ 2 is diagnostic for IVNC. On colour Doppler imaging the trabeculation are both increased in prominence and excessive in number and deep recesses are filled with blood from the ventricular cavity.

IVNC is observed in one or more ventricular wall segments. In79% three and more segments were involved.[5] Most commonly, the apical and midventricular segments of both the inferior and lateral wall were affected in more than 80% of the patients and the midventricular anterior wall and septum and the basal segments was much less frequently involved.[5] The location of the prominent trabeculation in patients with IVNC was typically apical, inferior and lateral which is different from the prominent trabeculation found in normal or hypertrophied heart. Prominent LV trabeculation can be found in healthy hearts (68%) and can be observed in hypertrophic hearts secondary to dilated, valvular or hypertensive cardiomyopathy. The differentiation depends on demonstration of the two layered myocardial wall with a thin epicardial compacted zone and an extremely thickened endocardial noncompacted zone with deep recesses and a segmental rather than a diffuse thickening or hypertrophy. Prominent trabeculation in normal hearts most frequently (85%) course from the free wall to the ventricular septum.[7]

All noncompacted segments were hypokinetic. The normally compacted segments were occasionally hypokinetic despite normal wall thickness which was reflected by the impaired fractional shortening in 82%.[5] Oechlin et al[5] demonstrated

an enlarged left ventricular end-diastolic diameter (≥60 mm) in (67%), and reduced fractional shortening (<29%) and reduced left ventricular ejection fraction < 50% in 86% of their cases.

Cardiac catheterisation demonstrates normal left ventricular volume and increased left ventricular end-diastolic pressure, consistent with restrictive hemodynamic.[8] Hook et al[9] reported a case of IVNC presenting as restrictive cardiomyopathy. In contrast Chin et al found in their study decreased left ventricular systolic function similar to that of dilated cardiomyopathy. Discrepancy in the hemodynamic characteristic may represent the different stages of the disease process. The abundant trabecular network may limit distensibility of the left ventricle and cause restrictive hemodynamics.[8] Symptomatic patients with rapidly progressive clinical course may show hemodynamic properties similar to dilated cardiomyopathy, whereas asymptomatic patients may follow a slowly progressive course of restrictive hemodynamic physiology.

On left ventriculography Ichida et al[8] demonstrated the sponge-like appearance of the non-compacted ventricular wall during the diastolic phase and marked retention of the contrast material in the intertrabecular recesses during the systolic phase. In most of their cases, there was hypokinesia of the noncompacted ventricular wall and in one case diverticular configuration of the noncompacted ventricular wall was present.

Thallium-201 myocardial imaging[8] in 14 patients at rest demonstrated a hypoperfusion area in the left ventricle corresponding to the zones where noncompacted ventricular myocardium was localised.

Magnetic resonance imaging was able to distinguish inner zones of noncompacted myocardium from the thin outer zones of compacted myocardium. T2-weighted sequences revealed high signal intensity areas at the apex of the left ventricles in two cases, which thought to be due to disturbed microcirculation due to fibrosis, thrombus formation and hypokinesis of this area.[8]

Soler et al[10] reported the first case of first-pass MR perfusion imaging demonstration of subendocardial perfusion deficit at rest in the noncompacted myocardium of the anterior and septal walls and in the normal myocardium of the inferolateral wall.

ECG –gated spin-echo echo-planar images showed thickened myocardium and heterogeneous signal due to flow void areas. Fast gradient echo sequences demonstrated prominent trabeculations and typical blood-filled deep recesses.[10]

Compared to echocardiography MRI is less operator dependent and might be superior to echocardiography in case of impaired acoustic window and thrombus hidden in the sponge like myocardium of IVNC might not be detected.

Ichida et al[8] reported in computed tomography early defects and delayed enhancement of the noncompacted ventricular myocardium, implying fibrosis in this area. Endomyocardial biopsy demonstrated a wide range, interstitial fibrosis, endomyocardial thickening, subendocardial fibroelastosis, myocyte hypertrophy and intramural thrombosis.

Reported CT findings includes markedly thickened myocardium of the left ventricular wall with two zones of different attenuation. The thin outer portion of compacted myocardium consisted of uniform tissue attenuation iso-intense to that of muscle and an inner thicker layer composed of soft tissue attenuation of trabecular myocardium and contrast-enhanced ventricular blood filling the deep intertrabecular recesses.[11]

CONCLUSION

Isolated noncompaction of the ventricular

myocardium (IVNC) is a rare congenital cardiomyopathy with characteristic imaging findings and can be recognised on CT imaging. However it is suggested that MRI should be the modality of choice for diagnosis and follow-up of patients with IVNC.

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