Isolated left ventricular noncompaction in a patient presenting with a subacute myocardial infarction

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Isolated left ventricular noncompaction is a rare cardiomyopathy that is often not recognised. So far, it is not well established how best to manage this abnormality. We describe a patient in whom the diagnosis of isolated left ventricular noncompaction was made after presentation with a subacute myocardial infarction. Because of non-sustained ventricular tachycardias during hospitalisation, which were inducible and deteriorated into ventricular fibrillation on electrophysiological examination after coronary artery bypass grafting, he received an implantable defibrillator. Whether the ventricular tachycardias were due to the myocardial infarction or to the noncompacted myocardium remains uncertain. (*Neth Heart J* 2007; 15:109-11.)

Keywords: noncompaction (isolated), cardiomyopathy, echocardiography

A 73-year-old man without a cardiac history presented to the Emergency Department because of chest pain radiating to the throat for two days. On physical examination he measured 1.70 m and weighed 58 kg. His blood pressure was 110/60 mmHg and the pulse regular at a rate of 70 beats/min. The jugular venous pressure appeared normal. Cardiac auscultation revealed normal heart sounds without murmurs. The lungs were clear to auscultation. The electrocardiogram showed sinus rhythm with right bundle branch block, 1 mm ST-segment elevation in leads I, II, aVL and V₃ to V₆, and slightly terminally negative

T waves in leads V₃ to V₆. The chest X-ray was normal with a cardiothoracic ratio of 43%. Creatine kinase (CK) and CK-MB were increased at presentation (2263 and 356 U/l, respectively). The diagnosis of subacute anterolateral myocardial infarction was made and the patient received aspirin, clopidogrel, low-molecular-weight heparins, nitrates, β -blockers and a statin. The next day, the CK and CK-MB had fallen and transthoracic echocardiography revealed akinetic anterolateral wall segments, a left ventricular ejection fraction of 35 to 40%, no significant valve disease, and prominent trabeculation of the apex of the left ventricular myocardium consistent with noncompaction. The presence of noncompaction was confirmed by contrast echocardiography (1.5 ml SonoVue intravenously), showing direct communication between the left ventricular cavity and the deep intertrabecular recesses (figure 1). The patient's convalescence was complicated by the occurrence of recurrent non-sustained ventricular tachycardia from the fourth day after admission, and ST depression (without chest pain) during bicycle ergometry on the fifth day after admission. Coronary angiography showed significant two-vessel disease of the left anterior descending (LAD) and left circumflex (LCX) coronary artery. Three weeks after admission, the patient underwent coronary artery bypass grafting (CABG). The left internal mammary artery was anastomosed with the LAD, and a venous jump graft was anastomosed with the obtuse marginal branch of the LCX via the anterolateral branch of the LCX. At electrophysiological examination nine days after CABG, a sustained monomorphic ventricular tachycardia was induced, which deteriorated into ventricular fibrillation during over-pacing and had to be terminated by defibrillation. The patient received an implantable defibrillator before he was discharged from hospital.

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Isolated left ventricular noncompaction is categorised as an unclassified cardiomyopathy in the report of the 1995 World Health Organisation/International Society and Federation of Cardiology task force on the definition and classification of cardiomyopathies.¹ It is characterised by hypertrabeculation of the myo-

Figure 1A. Transthoracic echocardiography (apical four-chamber view) of the described patient, revealing compacted (*) and non-compacted (**) myocardium of the left ventricle. **B.** Contrast echocardiography (1.5 ml SonoVue iv) of the same patient showing direct communication between the left ventricular cavity and the deep intertrabecular recesses. LA=left atrium, RA=right atrium, LV=left ventricle, RV=right ventricle.

cardium with deep intertrabecular recesses communicating directly with the left ventricular cavity, resulting in two layers of myocardium with a ratio of noncompacted to compacted myocardium $\geq 2:1$. The affected segments are usually located in the apical, mid-lateral and mid-inferior regions and are often hypokinetic. By definition, coexisting cardiac anomalies have to be absent.² Isolated left ventricular noncompaction is believed to be a congenital disorder, due to an arrest of the normal process of compaction of the loose myocardial meshwork between weeks five to eight of foetal life.³ Hypertrabeculation of the left ventricle, not fulfilling the criteria of isolated left ventricular noncompaction, is sometimes seen in patients with other congenital cardiac anomalies or in patients with neuromuscular disorders.⁴ Isolated left ventricular noncompaction has been described in children as well as adults, including the elderly.⁵ The prevalence is low, probably 0.05%, but may actually be higher because of underdiagnosing or misdiagnosing as hypertrophic cardiomyopathy.⁶ Men appear to be affected more often (about 70%) than women.⁷ Transthoracic echocardiography is the diagnostic test of choice, while contrast echocardiography can help to establish the diagnosis.⁸ Magnetic resonance imaging is useful in cases with poor echocardiographic image quality.⁹ The three most common clinical manifestations of isolated left ventricular noncompaction are heart failure, arrhythmias (atrial and/or ventricular), and thromboembolic events (including cerebrovascular accidents, transient ischaemic attacks, pulmonary embolism and mesenteric infarction).¹⁰ Heart failure, occurring in about 65% of patients, should be treated according to standard guidelines. In refractory cases heart transplantation can be indicated.¹¹ Atrial arrhythmias (25% of patients) mainly involve atrial fibrillation, necessitating prophylactic

oral anticoagulation. Ventricular arrhythmias (45% of patients) may make implantation of a defibrillator inevitable to prevent sudden cardiac death. Thromboembolic complications (30% of patients), due to atrial fibrillation or stasis of blood in the dilated heart or possibly in the deep intertrabecular recesses, need to be prevented by oral anticoagulation. The four-year survival of isolated noncompaction appears to be quite variable, ranging from 60 to 95%. The most common causes of death are heart failure and sudden cardiac death.¹² High-risk features for death include higher left ventricular end-diastolic diameter on presentation, New York Heart Association class III-IV, permanent or persistent atrial fibrillation, and bundle branch block.¹³ Because of the supposed familial association of isolated noncompaction, screening of first-degree relatives by echocardiography is recommended.¹⁴

Conclusion

In the described case, the diagnosis of isolated left ventricular noncompaction was made by chance, during hospitalisation for a subacute myocardial infarction. The ventricular tachycardias during convalescence, inducible and deteriorating into ventricular fibrillation at electrophysiological examination after (complete) revascularisation, could have been due to the scar of the myocardial infarction, to the noncompaction itself, or to a combination of these conditions. Because of the inducible sustained ventricular tachycardia at electrophysiological examination after complete revascularisation, there was a class I indication for an implantable defibrillator. The defibrillator may in this case also prevent possible ventricular arrhythmias caused by the noncompaction in the future. ■

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