Additive value of contrast echocardiography for the diagnosis of noncompaction of the left ventricular myocardium

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Abstract Noncompaction of the left ventricular myocardium is a rare and unclassified cardiomyopathy that remains frequently overlooked, even by experienced echocardiographers. This fact may be due to non-optimal imaging of the lateral and apical myocardium, and/or insufficient disease awareness by echocardiographers. We report a case of a young man with heart failure and severe left ventricular dysfunction, previously diagnosed of ischemic etiology, in which the contrast enhancement of left ventricular endocardium allowed us to reach the correct diagnosis of isolated left ventricular noncompaction.

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Noncompaction of the ventricular myocardium (NVM) is a rare and unclassified cardiomyopathy that remains frequently overlooked, even by experienced echocardiographers. This fact may be due to several factors: non-optimal imaging of the lateral and apical myocardium, similarity of left ventricular (LV) noncompaction to other disease of myocardium and endocardium, and/or insufficient disease awareness by echocardiographers. We report a case of a young man with heart failure and severe LV dysfunction, previously diagnosed of ischemic etiology, in which the contrast enhancement of LV endocardium allowed us to reach the correct diagnosis.
Case report

A 45-year-old white male with a history of dyspnoea on effort (NYHA functional class II) and LV dysfunction (LV ejection fraction = 28%) was referred to our echocardiography laboratory for reassessment of his LV function. His cardiac history began in 1996, when he was 37 years old. He started to complain of worsening dyspnoea on effort. He underwent transthoracic echocardiogram which showed severe LV systolic dysfunction (LV ejection fraction = 30%), an aneurysm of the LV apex with a large mural thrombus in it. The patient had never complained of chest pain, and had no cardiovascular risk factor except smoking. Coronary angiography did not show any coronary anomaly or significant obstructive coronary artery disease. The left ventriculography was not performed. The patient was treated with beta-blockers, ACE inhibitors, aspirin and warfarin. Warfarin was stopped in 1998 when he was 39, and after several echocardiographic studies that showed unchanged thrombus morphology. In 1999, he had a stroke. The patient did not complain of palpitations. A 24-h Holter monitoring, performed in February 2001, showed 0–10 isolated premature ventricular beats/h. In November 2001, when he was 42, he underwent treadmill exercise testing which demonstrated poor exercise capacity (test stopped after 4 min) and low maximal oxygen consumption (14 ml/kg/min). In 2003, when the patient was 44, he was moved to our city and his new general practitioner referred him to our outpatient clinic for re-evaluation.

The patient was symptomatic for dyspnoea on effort (NYHA functional class = II). At the physical examination, the pulse was 62 bpm. Blood pressure was 140/80 mmHg. A 2/6 holosystolic murmur was heard at the apex and a loud S4 was heard on the precordium. The peripheral pulses were present and were symmetric; there were thin rales heard over the lung bases at the end inspiration.

Electrocardiography demonstrated sinus rhythm, first degree atrio-ventricular block (PQ = 0.218 s), incomplete left bundle branch block (QRS duration = 0.214 s), and non-specific T wave abnormalities in V5 and V6.

The transthoracic echocardiogram was performed with a commercially available echocardiograph (Sonos 5500, Philips, Andover, U.S.), using second harmonic imaging mode. It demonstrated a markedly dilated left ventricle with severely impaired systolic function. A large mass which showed the same echogenicity of the adjacent myocardium was detectable in the LV apex (except the septal segment) (Fig. 1). Color Doppler imaging showed areas of flow in both the apical mass and the surrounding myocardium (Fig. 2). In addition, aberrant bands were visualized on anterior interventricular septum.

Images with second-generation echocardiographic intravenous contrast agent (Sono Vue™, Bracco srl, Italy) were used to obtain a more detailed evaluation of these findings. Contrast enhancement was achieved using 1 ml bolus of Sono Vue™ followed by immediate flush with 10 ml of normal saline. This technique revealed that the apical mass was composed of several prominent trabeculations and deep intertrabecular recesses which had a synchronous movement of contraction with ventricular myocardium. Contrast clearly

Figure 1 Apical 4-chamber view showing a large mass extending from mid lateral wall to lateral apex. The mass showed the same echogenicity of the adjacent myocardium.

Figure 2 Color Doppler study showing communication between left ventricular cavity and the intertrabecular spaces of the mass.
showed the direct communication between the interventricular spaces and LV cavity (Fig. 3). In addition, contrast echocardiography allowed us to assess precisely the depths of the trabeculations, otherwise faintly seen using Color Doppler, and to calculate the ratio between noncompaction and compaction layer, which was 2.5 (Fig. 4).

Isolated NVM was the final diagnosis.

**Discussion**

Noncompaction of the LV myocardium is an unclassified cardiomyopathy thought to be caused by the arrest of normal embryogenesis of the myocardium and endocardium. This abnormality is frequently associated with other congenital cardiac defects or extracardiac disorders, particularly neuromuscular. The prognosis of NVM remains controversial and clinical manifestations of the disease are highly variable ranging from no symptoms, to congestive heart failure, to ventricular arrhythmias, and to systemic thromboembolism. Echocardiography is the most widely used diagnostic technique and echocardiographic diagnostic criteria include: (1) the absence of any coexisting cardiac anomalies such as aortic or pulmonary valve obstruction and coronary artery anomalies; (2) the identification of a 2-layered structure of the left ventricular wall, characterized by a thin compacted epicardial and endocardial with numerous prominent trabeculations and deep intertrabecular recesses. The end-systolic ratio of noncompacted to compacted layer is > 2; (3) the involvement especially of the mid-lateral or apical or mid-inferior areas of the left ventricle; (4) the detection of a blood flow directly from the LV cavity into the deep intertrabecular recesses. Isolated NVM is diagnosed when these 3 criteria are satisfied. Detection of all these signs using echocardiography depends critically on image quality. Suboptimal imaging of the apical and mid-ventricular myocardium may prevent the echocardiographer to identify intertrabecular recesses, and to accurately measure thickness of the noncompacted to compacted layers. This imaging problems and lack of echocardiographer awareness of NVM make the correct diagnosis difficult. As a consequence, many cases of NVM have been misinterpreted as hypertrophic cardiomyopathy, endocardial fibroelastosis, dilative cardiomyopathy, restrictive cardiomyopathy, apical thrombosis, and endomyocardial fibrosis.
Echocardiographic contrast agents are useful to delineate endocardial borders and in this way, they improve the accuracy and the reproducibility of assessment of heart chambers morphology and function. With contrast enhancement, the LV endocardial borders are sharply demarcated allowing an optimal visualization of the prominent myocardial trabecular recesses, a intertrabecular flow from the LV cavity and an accurate assessment of the noncompacted/compacted layer thickness ratio.

References