LETTER TO THE EDITOR

An unusual case of non-compaction cardiomyopathy associated with coronary artery disease

KEYWORDS
Non-compaction cardiomyopathy; Coronary artery disease; Pulmonary embolism

1. Introduction

Non-compaction cardiomyopathy (NCC) is a morphological abnormality of excessive myocardial trabeculation and deep intertrabecular recesses with or without left ventricular (LV) dysfunction.1,2 The European Society of Cardiology refers to NCC as an “unclassified cardiomyopathy”.3 In fact, current evidence suggests that it is unlikely a distinct cardiomyopathy.4 Traditionally, NCC is diagnosed by 2D echocardiography.1,5,6 It can be diagnosed at any age, and an association between NCC and additional cardiac anomalies and neuromuscular disorders has been identified.5 This report describes a case of NCC associated with complex three-vessel coronary artery disease (CAD).

2. Case presentation

A 71-year-old man, who travelled a week earlier by plane, was admitted to the pulmonology clinic due to severe dyspnea and fever. The working diagnosis based on both the clinical examination and the chest X-ray was pneumonia. On the tenth day of his hospitalization, the patient developed acute dyspnea and hypotension. At that point, a cardiology consultation was requested. Physical examination revealed a blood pressure of 80/50 mmHg, a pulse of 100/min, normal heart sounds, no fever, crepitations on the lower right lobe, and SpO2 = 85%. Laboratory findings showed increased total white blood cells of 27,700 (neutrophils: 89%), Hgb = 12 g/dl, troponin T levels of 0.45 ng/ml, and d-dimers = 1600–3200. The blood gas analysis showed the following: pH = 7.45, pO2 = 50 mmHg, pCO2 = 37 mmHg, and HCO3 = 27 mmol/l. Electrocardiography (ECG) showed sinus tachycardia, a new right bundle branch block, and S1Q3T3 compatible with a pulmonary embolism (Fig. 1b). The whole picture was suggestive of severe pulmonary embolism. Reviewing the ECG on admission showed QS in the precordial leads compatible with an “old” myocardial infarction (Fig. 1a). An urgent chest CT-scan confirmed the clinical diagnosis of severe pulmonary embolism (Fig. 2). In addition, a “deficit” in the left ventricular apex was noted (Fig. 2b). Pneumonia in the right lower lobe and pleural effusion were also noted (Fig. 2c,d).

The patient was then admitted to the intensive care unit. Urgent transthoracic echocardiography (TTE) revealed a dilated left ventricle, severely reduced ejection fraction with global hypokinesia (LVEF = 20–25%), two large thrombi and near normal right cavities (Fig. 3a–c). Furthermore, mild to moderate mitral valve regurgitation, moderate tricuspid valve regurgitation and moderate pulmonary hypertension (RVSP = 55 mmHg) were noted. In view of these findings (thrombi), thrombolysis was not performed due to the risk of a new thromboembolic event from the lysis of thrombi and the patient was treated with low-molecular weight heparin, oxygen, inotropes and antibiotics. During the first few hours, the patient suffered from two nearly fatal arrhythmological complications (sustained VT, cardiac standstill, both easily treated with precordial thumb). Amiodarone and a low dose of carvedilol were added to his treatment. Bed-side triplex of the lower limbs showed extensive severe left-sided deep vein thrombosis (DVT). After stabilization, the patient was transferred to the cardiac laboratory for a more detailed TTE. Surprisingly, the whole LV picture was typical of NCC. There was a thin epicardial layer and an extremely
thickened endocardial layer with prominent trabeculations and deep recesses. The predominant localization of the pathology was to mid-lateral and apical areas (Fig. 3d). The maximal end systolic ratio of non-compacted to compacted layers was greater than 2, which is diagnostic for NCC (Fig. 3c). Blood flow from the ventricular cavity into the deep recesses was also noted on color Doppler imaging (Fig. 3e). A further 3D study with contrast clearly demonstrated the abnormal trabeculations and the two large thrombi in the LV (one was attached to the apex and the other one was entrapped in the trabeculations of the intraventricular septum) (Fig. 3f).

In view of the presence of the q waves on the admission ECG and possible recent subacute MI, we investigated his history further. The patient never smoked or had any other risk factors for CAD, and he denied that he had any heart problems. When asked specifically about the start of his symptoms, the patient recalled that three days before travelling to Cyprus he visited his general practitioner due to acute onset of chest “discomfort”. At the time, the working diagnosis was chest infection that was treated with oral antibiotics. No ECG or chest X-ray was performed at that time. Despite his symptoms, the patient was allowed to travel to Cyprus for the holidays.

A series of diagnoses were thus established: a) NCC (not known until that day) with typical TTE findings, b) a recent anterior myocardial infarction (2–3 weeks previously) that was not diagnosed or treated, c) the presence of a severely impaired LVEF, d) left lower limb extensive DVT, e) a massive pulmonary embolism (provisionally DVT occurred during travel by plane), f) the presence of two large thrombi in the LV (one of them entrapped in the trabeculae due to the non-compacted LV, rather than the recent MI), and g) pneumonia.

In view of the above findings, the critical condition of the patient and the fact that the thrombi were organized,
after further stabilization of the patient, we proceeded to use a dobutamine stress echo (DSE) to prove the presence of viability before referring the patient for a coronary angiogram. In fact, there was viability during DSE of all the myocardial walls. The coronary angiogram that was performed showed extensive obstructions (LMCA: proximal stenosis (60%), LAD: occluded proximally, ramus intermedius: proximal stenosis, and RCA: occluded ostially) despite the absence of risk factors (Fig. 4). The patient was readmitted to the ICU after the coronary angiogram due to his fragile status and the wait for the urgent coronary artery bypass graft (CABG). A week later, he was transferred to a tertiary hospital for CABG (LIMA to LAD, SVG to ramus intermedius, SVG to PDA). The patient was discharged ten days after the CABG in good general condition. No further complications occurred during the peri- and post-operative period (the thrombi in LV and the extensive DVT were still present despite anticoagulation). The treatment on discharge was warfarin, aspirin, carvedilol, amiodarone, candesartan, atorvastatin and omeprazol. A week later, the patient was safely repatriated with the suggestion of further investigation for the possibility of CRT-D implantation and that his family be screened for NCC by echocardiography.

3. Discussion

The exact incidence of NCC is not known, although it is reported to be 1/10,000 per year in the general population based on echocardiographic studies. ECG is a reliable, noninvasive imaging technique for the diagnosis of patients with NCC. All criteria that are currently in use are based on morphological features and mandate the presence of excessive trabeculations with deep intertrabecular recesses communicating with the LV cavity and a two layered appearance to the myocardium (trabecular, and compacted myocardium). However, based on current criteria, patients are often misdiagnosed. Advanced echocardiography methods, such as 3D echocardiography and LV opacification with contrast administration, have been proposed to improve diagnostic accuracy. Cardiac MRI also has an emerging role in the diagnosis of NCC, especially when the echocardiographic image quality is poor. NCC is often complicated by ventricular dysfunction, rhythm disturbances and cardioembolism, which are the main clinical manifestations of the disease. Therefore, patients need to be properly treated with anti-failure, anti-arrhythmic, and anti-coagulation therapy and possibly with CRT-D implantation. Furthermore, comprehensive clinical screening is recommended in asymptomatic first-degree relatives of patients diagnosed with NCC. Although the association of NCC with CAD was once believed to be rare, there are a series of recent reports of an association between NCC and subclinical or acute myocardial infarction, and it is possible that this association has previously been undiagnosed. Furthermore, one study that performed cardiac catheterization in patients with NCC showed that 29% of the patients had significant CAD. Cardiac ischemia as a feature of NCC is considered to be a consequence of microvascular dysfunction. NCC patients exhibit decreased coronary flow reserves in both compacted and non-compacted segments.
NCC, ischemia could be present even if coronary arteries are angiographically normal.\textsuperscript{17}

In conclusion, we reported a case of NCC associated with complex three-vessel CAD. Furthermore, the combination of a massive pulmonary embolism and clots in the LV made the patient’s management even more challenging.

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**Disclosures**

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**References**


Stylianos Karvounaris, MD, PhD a
George Michas, MD, MRes, PhD a
Vasilis Karampetsos, MD
Department of Cardiology, Paphos General Hospital, Paphos, Cyprus

Stefanos Fousas, MD, FESC, FACC
Department of Cardiology, Tzaneio General Hospital of Piraeus, Piraeus, Greece

Petros Mavrommatis, MD, FESC *
Department of Cardiology, Paphos General Hospital, Paphos, Cyprus

*Corresponding author. Petros Mavrommatis, MD, FESC, Department of Cardiology, Paphos General Hospital, Paphos, Cyprus. Tel.: +357 26803463; fax: +357 26803481.
E-mail address: p.mavrommatis@gmail.com
(P. Mavrommatis)

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a Equal Contribution.