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Left ventricular non-compaction cardiomyopathy and ischaemic stroke

Cătălina Elena Bistriceanu[1, 2], Florentina Anca Danciu[3], Dan Iulian Cuciureanu[2, 3]

[1]Elytis Hospital Hope, Iasi, Romania
[2]Neurology Department, Faculty of Medicine, Grigore T. Popa University of Medicine and Pharmacy, Iasi, Romania
[3]Prof. Dr. N. Oblu Neurosurgery Clinical Emergency Hospital, Iasi, Romania

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To the Editors

Non-compaction cardiomyopathy is a rare congenital myocardial disorder that results due to the arrest of left ventricle compaction during embryogenesis. Other terms such as "spongy myocardium" or "persistent embryonic myocardium" have been used, but most frequently this disorder is known as left ventricle non-compaction or non-compaction cardiomyopathy [1].

The 'compaction' of the spongy myocardium (with a trabecular appearance) is during weeks 5–8 of embryogenesis, and the interruption of this process leads to persistence of these trabeculations continuous with ventricular cavity and a cessation of the communication with epicardial circulation [2]. More than 40 genes coding for sarcomeric, cytoskeletal, ion channels and desmosomal proteins have been identified in this disorder [3].

Atrial fibrillation, decreased systolic function and intratrabecular thrombus formation found in the left ventricle are common clinical features of left ventricular non-compaction, and can be involved in thromboembolic events [2]. These events can be stroke, transient ischaemic attack, mesenteric, myocardial and renal infarction, or peripheral embolism [4].

We here report the case of a 47-year-old man who was admitted to the neurological department for sudden onset of weakness in left limbs and slurred speech of 3.5 hours' duration. On admission, the patient showed no signs for mesenteric, myocardial, renal or peripheral embolisation. He had a history of alcohol drinking and cigarette smoking, without any additional vascular risk factors. His father had died young due to myocardial infarction.

The neurological examination revealed left hemi-sensory deficits and left-sided hemiparesis (strength was 3+/5 on the left hand and 4+/5 on the left foot, and the Babinski reflex was extensor on the left and flexor on the right), oculocephalic deviations to the right, and dysarthria with an NIHSS score of 6 points. The CT scan performed in the emergency department revealed an acute hyperdense thrombus in M1-M2 segments of right middle cerebral artery. The ASPECTS score was 9 points. The routine blood exam was unremarkable. Thrombolysis with Alteplase was performed, and the NIHSS score during hospitalisation decreased to 3 points.

The cerebral MRI performed two days after thrombolysis confirmed ischaemic lesions in the right middle cerebral artery territory and a small contralateral parietal lesion (Fig. 1A–C). We did not identify cervical or intracranial atherosclerosis.

The cardiovascular investigations such as transthoracic echocardiography revealed an obvious trabecular aspect of the left ventricle, with an EF (ejection fraction) < 20% (Fig. 1D–E). Stöllberger echocardiographic criteria [5] and Paterick criteria [6] for left ventricular non-compaction were met.

Stöllberger et al. [5] defined the echocardiographic criteria for LVNC as: three or more trabeculations protruding from the LV endocardial border in end-diastole; trabeculations are moving synchronously with the compacted myocardium; these trabeculations are non-compacted part of the two-layered

Address for correspondence: Catalina Elena Bistriceanu; University of Medicine and Pharmacy, Iasi, Romania; e-mail: catalina_nastac@yahoo.com

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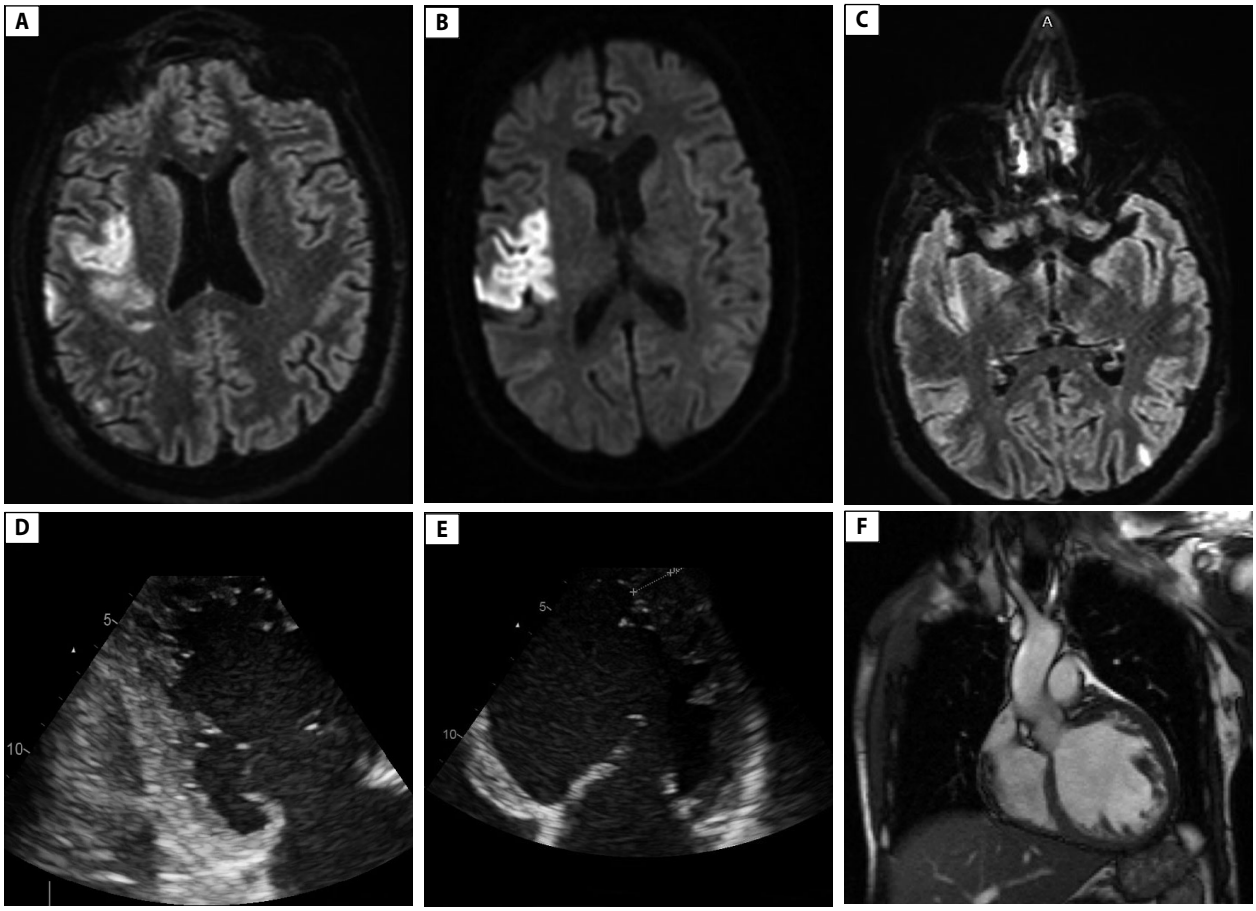

Figure 1. A. axial FLAIR: high signal in right MCA territory; **B.** Axial diffusion: confirming acute right superficial MCA ischaemic stroke; **C.** Axial FLAIR: small left parietal area with high signal; **D.** Echocardiography: apical two-chamber view: > 3 prominent trabeculations with deep intertrabecular recesses (Stöllberger criterion) [5]; **E.** Apical four-chamber view, end-diastole, left ventricle zoom, 2-layered myocardium with non-compaction/compaction ratio = 1.3 cm/0.7 cm = 1.85 (Paterick criterion: ratio > 2) [6]; **F.** Cardiac MRI: myocardial non-compaction of left ventricle

myocardial structure; and perfusion of the intertrabecular spaces from the ventricular cavity is present at end-diastole on colour-Doppler echocardiography.

According to Paterick et al., the echocardiographic criteria for LVNC are: an evaluation of the trabeculations' sizes (non-compacted myocardium NC) in relation to compacted (C) wall thicknesses in multiple imaging windows; identification of the bilayered myocardium (C and NC), in the short-axis views at the mid- and apical levels, and in the apical 2- and 4-chamber and apical long-axis views; thicknesses of the C and NC sections of the myocardium are best measured in the short-axis views at end-diastole, with NC/C ratio > 2 being diagnostic of LVNC; and abnormal ventricular function and abnormal myocardial mechanics, along with the above noted features, to diagnose LVNC cardiomyopathy [6].

A cardiac MRI was performed, confirming the diagnosis of non-compaction cardiomyopathy (Fig. 1F).

In the literature, we found common opinions about complications, but there was a lack of consensus in specific guidelines about the type of anticoagulation needed for secondary ischaemic stroke prevention.

Non-compaction cardiomyopathy may remain undiagnosed until adulthood when complications like cardiac arrhythmias, cardiac failure, thromboembolic events or sudden death can be seen. The management of this disorder is dependent on clinical manifestations. The treatment is addressed to cardiac failure management, comprising an implantable defibrillator or, in some cases, cardiac transplant [2].

Because this cardiac disorder has a low prevalence, there have been no large randomised trials regarding the clinical management of this disorder, especially regarding the anticoagulation treatment [4].

According to the American Heart Association 2021 guidelines for secondary stroke prevention, five randomised trials have evaluated the effects of antithrombotic therapy on clinical outcome, including stroke, in patients with heart failure and reduced LV EF in sinus rhythm.

The WARCEF trial (Warfarin *vs.* Aspirin in Reduced Cardiac Ejection Fraction) documented no benefit of warfarin therapy compared to acetylsalicylic acid at a mean follow-up of 3.5 years for the primary outcome (i.e. death, ischaemic stroke, or intracranial haemorrhage), although patients on warfarin

had reduced incidence of stroke, particularly patients with an EF ≥ 15. The only randomised trial evaluating the benefit of DOACs (dabigatran) in stable patients after LVAD (Left Ventricular Assist Device) implantation was halted prematurely because of an excess of thromboembolic events [7].

In our case, we supposed that cardio-embolic mechanism was implicated in the stroke aetiology (supported also by imaging examination), and we decided to start the treatment with DOACs.

Some studies have recommended routine anticoagulation for primary thromboembolic prevention in left ventricular non-compaction, while others have suggested anticoagulation when systolic dysfunction (EF < 40%), atrial fibrillation, or intracardiac thrombus exist [8]. In our patient, prolonged cardiac monitoring put into place post-treatment did not reveal atrial fibrillation or any other cardiac arrhythmia.

Hypertrabeculation and affected left ventricular function predispose patients to cerebrovascular events, and the data from the literature recommends oral anticoagulation when such a diagnosis is confirmed.

Because this is a rare disease, evidence-based recommendations for preventing thromboembolic events in this disorder are not well established. The utility of echocardiography can be considered to estimate the duration of anticoagulation [2, 8].

New insights into understanding the genetic aetiology and the utility of screening relatives are promising areas for further research. It would be most valuable to establish consensus guidelines [8].

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