

Silent Lacunary Brain Infarctions Associated with Left Ventricular Noncompaction and Idiopathic Epilepsy: A Case Report

Mihaela Lungu¹, Violeta Sapira² and Manuela Arbune³

¹ EMERGENCY CLINICAL HOSPITAL, GALATI, ROMANIA

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Abstract

Non-compaction of the left ventricle is a rare cause of cardiomyopathy, sometimes with a family character that may complicate evolving with heart failure, heart rhythm disorders and systemic embolic events - including stroke. We present the case of a young patient in neurological dispensary for epilepsy where the neuroimaging evaluation of an acutely installed cephalalgia syndrome revealed multiple silent lacunar brain injuries. Paraclinic examinations of their etiology has led to the diagnosis of a non-compaction of the left ventricle, in this context the lacunar infarcts are considered embolic cerebral events with a cardiac starting point.

Index terms— left ventricular noncompaction, silent lacunar infarction.

1 I. Introduction

Left ventricular noncompaction (LVNC) is a rare cardiomyopathy, characterized by the presence of an excessive trabeculation of the myocardium, which mostly affects the left ventricle. There are numerous excessive trabeculations and intertrabecular recesses, which communicate with the left ventricular cavity.

Pathogenetically it is believed that the cardiomyocyte compaction process should stop during embryogenesis.

Clinical aspects are similar to other cardiomyopathies and include systolic and diastolic dysfunctions of the left ventricle, tachyarrhythmias and systemic embolism.

2 II. Case Report

A 35-year-old female patient that went through neurological dispensary for 10 years for idiopathic epilepsy with generalized tonic-clonic seizures controlled by anticonvulsant therapy with valproic acid and salts -1000 mg/day was admitted into our neurological department due to an acute onset of a headache.

The family history of the patient included sudden deaths of several members: mother, uncle mother's brother and maternal grandmother.

Personal pathological antecedents included: idiopathic epilepsy with generalized therapeutically controlled crises, erosive gastritis with secondary mild iron anemia and irritable bowel.

The overall clinical examination of the organism and systems was within regular limits with normal blood pressure values, and the neurological exam did not reveal any neurological signs of the outbreak.

The neuro-imaging evaluation was a cerebral magnetic resonance angiography done with gadolinium which showed lacunary images in the temporal right lobe in the posterior hippocampus around the temporal ventricular horn as well as deep in the right insular area and also in the right brain peduncle, without any contrast intakes. The appearance of multiple silent lacunar infarctions imposed for further investigations to find out the etiology.

Blood tests have reported a mild iron deficiency anemia, with hemoglobin of 10.4 g/dl, mean corpuscular volume -75 fL, iron deficiency -29 µg/dl and ferritin 5.9 ng/ml. Tumor markers for the digestive tract were

within ordinary limits. We performed a upper digestive endoscopy with an antral mucosa biopsy, which showed us the appearance of erosive gastritis.

The lipogram had normal values. We performed several tests to check for thrombophilia, for antiphospholipid antibody syndrome, for vasculitis and for thyroid disorders, which also proved negative, excluding these etiologies. The borreliosis, HIV and syphilis tests were also negative.

The extra cranial Doppler exam did not detect any carotid, vertebral or subclavian lesions.

The 24-hour Holter-electrocardiogram for rhythm didn't detect any paroxysmal rhythm disorders.

Cardiac echography revealed a mitral regurgitation-grade II, with eccentric jet towards the posterior left atrial wall, the lateral wall, the apex and the lower wall of the left ventricle with a trabecular structure: LV = 51/34; LA=31; RV=21; RA=27; IVS=8; AO desc=15; EF=55%. Conclusion: noncom action cardiomyopathy. Mild mitral regurgitation -fig. 1: Also, considering the hero-collateral history of the patient which includes three cases of sudden deaths in first and second degree relatives, one can assume a family component of these type of cardiomyopathy.

The patient is in the neurological dispensary, receives antiepileptic drugs: 1000 mg of Depakine Chrono/day -with therapeutic control of the seizures and anticoagulant treatment with gastric protection. Iron deficiency anemia was solved using specific treatment. The risk of the anticoagulant remedy in a patient with epileptic seizures remains in question.

3 III. Discussions

Left ventricular noncom action (LVNC) is a rare cardiomyopathy with an incidence of 0, 05-1, 3/100.000 births [1].

LVNC includes the persistence of the fetal spongiform structure with an excessive trabeculation of the myocardium, which mostly affects the left ventricle [2, 3], but possibly also the right ventricle [3].

There are numerous excessive trabeculations and intertrabecular recesses, which communicate with the left ventricular cavity [2], but it does not connect with the coronary circulation. It occurs in adults as well as in children.

In human hearts, the left ventricle has up to 3 prominent trabeculations, and it has fewer trabeculations than the right ventricle [4].

The condition can appear in two forms: an isolated and a non-isolated one, associated with other congenital diseases: ventricular septal defects, atrial septal defects, pulmonic stenosis, hypoplastic left ventricle, facial dysmorphism [5].

The American Heart Association has classified LVNC as primary genetic cardiomyopathy [6], while The European Society of Cardiology considers it as unclassified cardiomyopathy, based on the fact that LVNC may be a morphological manifestation of other severe distinct cardiomyopathies.

LVNC is not considered distinct cardiomyopathy, being included by some authors as a congenital or acquired morphological characteristic of other declarative cardiomyopathies.

Path genetically it was suggested that the most important fact might be an arrest of myocardial fibers in the intrauterine development, resulting in two different myocardial layers: one compacted and one noncompacted, trabeculated [3].

It can be either secondary to congenital ventricular outflow tract obstructions or familial (Barth's syndrome) [7].

Studies of the genetics of the LVNC have strongly suggested that the disease has an inheritance pattern (18% to 50% of cases are familial) [6].

Several studies suggest that LVNC is a genetical and heterogeneous disease with a sporadic and also a familial form, with pathogenic mutations in the genes encoding proteins such as cytoskeletal, mitochondrial, sarcomeric and Z-line proteins. Various autosomal dominant, recessive, X-linked or mitochondrial transmissions were described [8].

In sporadic cases which are common, there were detected new mutations [8].

There is the absence of a specific genotype-phenotype association in LVNC, the mutation of the same gene can determine LVNC as well as declarative or hypertrophic cardiomyopathies, which make the genetic testing have a restrained utility.

There were three genes identified as correlating with NCV: dystrobrevin-alpha (DTNA), cipher/ZASP (Z-line component which is both found in the skeletal muscle as well as in the cardiac muscle), TAZ (a gene with unknown dysfunction which is involved in the Xlinked declarative cardiomyopathy) [8].

Clinical aspects are similar to other cardiomyopathies and include systolic and diastolic dysfunctions of the left ventricle, tachyarrhythmias and systemic embolism [9].

The asymptomatic period varies significantly, but average duration from the onset of the symptoms to the diagnosis is, on average, of three years [10].

The triad formed by the symptoms of heart failure arrhythmia and cardio embolic events is the clinical manifestation in patients with diastolic dysfunction of the left ventricle [11]. Different types of arrhythmias can occur, from atrial fibrillation -7 -26% to sustained ventricular tachycardia [1].

The most important symptom is dyspnea, due to low cardiac output. Tachyarrhythmias in Wolf-Parkinson-

White syndrome, ventricular tachycardia's, atrioventricular blocks, bundle branch blocks and even sudden death have been reported [5] .

Over a mean follow-up of four years, the 36-year-old patient had cardiac events. There were five cardiac deaths, 16 heart failure hospitalizations, ten ventricular arrhythmias and five thromboembolic events [12].

Another study made by Chin et al. on 8 patients with LVNC state, revealed a cardio embolic event of a stroke type in a 2,3-year-old child, that led to his death [2] .

Two series reported transitory ischemic attacks or stroke in 25% of all patients with LVNC. [1,2] . Because the incidence of atrial fibrillation in these patients in 29% of the cases, systemic anticoagulation is recommended. The patients with LVNC have a different prognosis. Some studies associate the disease with high mortality due to heart failure and sudden cardiac death [5] . Others patients have a better prognosis [8] . The prognosis depends on the stage of the disease at the moment of the diagnosis, by the severity of the heart failure and of the improvements due to treatment.

The most significant preclinical diagnosis is the echocardiogram-transthoracic echocardiogram TTE (two-dimensional TTE and three-dimensional TTE), but also heart computed tomography and magnetic resonance detect left ventricular trabeculations and recesses, which serve as a nidus for mural thrombus [6,13].

For the positive diagnosis, the ratio between non-compacted and compacted myocardium must be above 2 (Jenny Criteria) [3] .

There was a high rate in the prevalence of LVNC in the last years, due to a better diagnosis with more performant echocardiography [13] .

There is no specific therapy for LVNC. The treatment addresses the three types of clinical manifestations: heart failure -beta-blockers, angiotensin converting enzyme inhibitors, diuretics, arrhythmias-ant arrhythmic drugs and systemic embolic events-anticoagulation [9] .

IV. Conclusions 1. Multiple silent lacunar infarctions in a young epileptic patient, without any vascular risk factors and with a family history of sudden death is a challenge, and it requires extensive paraclinical investigations, both for finding out the etiology as well as for the differential diagnosis. 2. In the clinical case of the patient we presented, the essential examination was cardiac echography, this being the one that detected the LVNC, which can explain through embolic mechanism the occurrence of silent cerebral lacunar infarctions. 3. The case is clinically significant because the incidence of LVNC cases is very low and out of these cases only 25% are quoted as having during their evolution a stroke episode which occurred through the embolic mechanism. 4. The familial history is suggestive for a potential familial cardiomyopathy. 5. Preventing systemic embolic recurrence involves, even the absence of associated atrial fibrillation the administration of anticoagulant medication. 6. The anticoagulant medication in a patient which also takes epileptic treatment may be risky, but the therapeutic control of the seizures has allowed the choice of this therapeutic option. 7. It is necessary to follow up the patient to apply a requisite treatment promptly in the case of a possible complication (heart failure, atrial fibrillation, neurological complications-ex.: head trauma). This article does not contain any studies with human participants, performed by any of the authors.

Informed consent: the informed consent of the patient was obtained and written in the observation sheet.

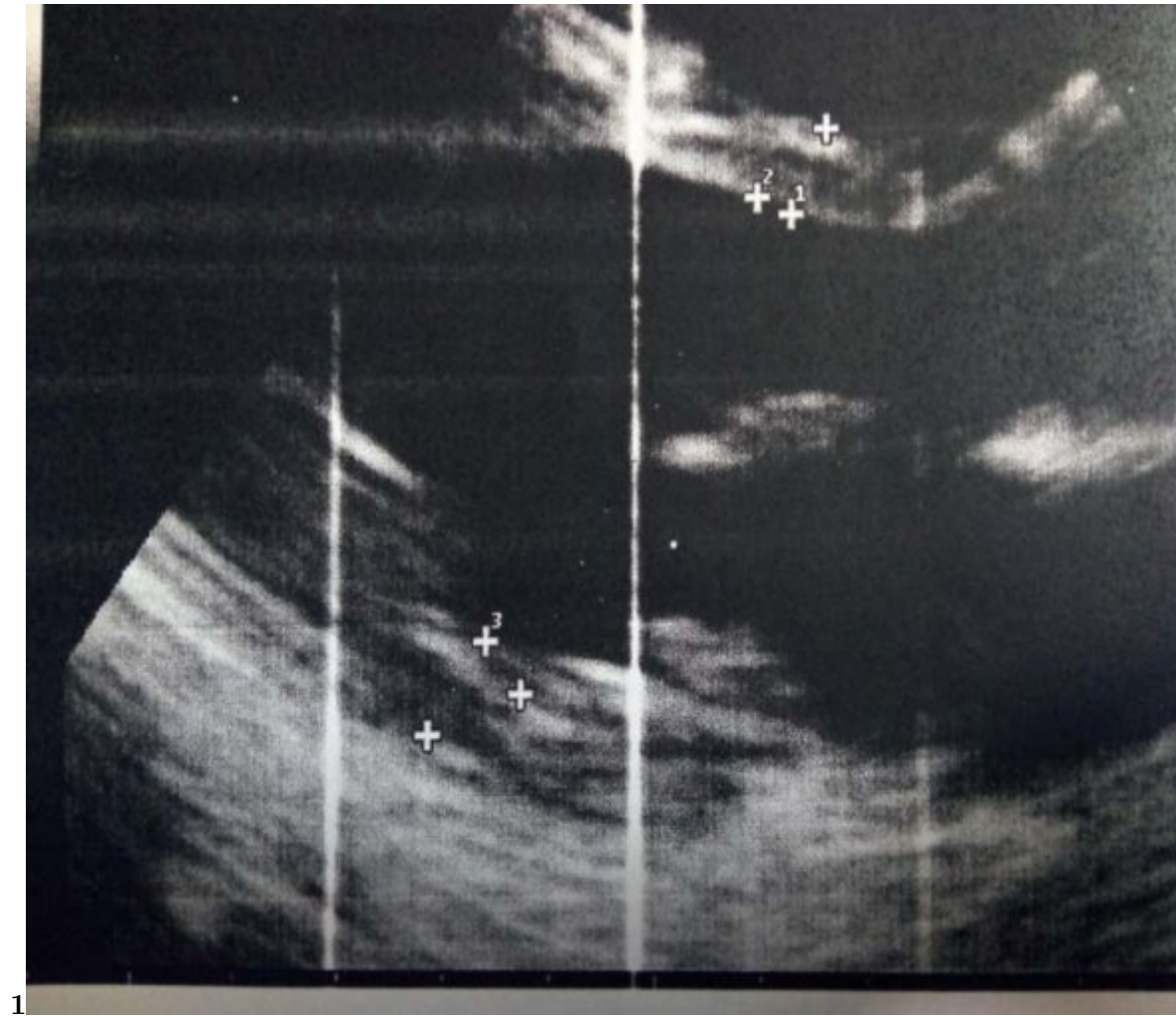


Figure 1: Fig. 1 :

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