Stroke and Ventricular Dysfunction in a Patient with Isolated Left Ventricular Noncompaction

Makoto Nakajima¹, Teruyuki Hirano², Hideki Doi³ and Makoto Uchino²

Abstract

A 44-year-old woman developed a cardioembolic stroke. Transthoracic echocardiography demonstrated isolated noncompaction of the ventricular myocardium. Left ventricular systolic function was mildly depressed, which severely decreased during 3 months after discharge. The embolic stroke might occur when the ventricular systolic function had begun to deteriorate. The proper time to start anticoagulation in isolated noncompaction of ventricular myocardium patients may be when left ventricular systolic function decreases below normal.

Key words: stroke, young adult, ventricular noncompaction

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Introduction

Isolated noncompaction of the ventricular myocardium (INVM) is a rare cause of stroke in young adults (1-5). Although embolic complications of this disease have been reported, there is a lack of evidence dealing with the relationship between changes in ventricular systolic function and embolic events. We report a case of INVM who developed an ischemic stroke during the time period that left ventricular systolic function deteriorated.

Case Report

A 44-year-old woman was admitted to our hospital due to a mild disturbance of consciousness. She had a past history of bronchial asthma and an ovarian cyst excision. She was never smoker; and had taken no medicine. Her mother had a history of ischemic stroke, and her elder sister had a cardiomyopathy and had also had an ischemic stroke. The patient developed drowsiness, unilateral hemispatial neglect, and mild hemiparesis on the left side; these symptoms fluctuated in the emergency room. At the time that the patient’s symptoms had deteriorated the most, the patient’s National Institutes of Health Stroke Scale score was 7. Diffusion-weighted magnetic resonance imaging demonstrated evidence of a new infarction in the right striatum and spotty ischemic lesions in the cerebral cortex. On magnetic resonance angiography, branches of the right middle cerebral artery were demonstrated incompletely (Fig. 1). Chest radiography showed no abnormalities except for mild cardiomegaly (Fig. 2). EKG showed sinus rhythm and some premature ventricular contractions (PVCs). On carotid ultrasonography, no plaques were seen, and no significant laterality of flow velocity was demonstrated in the carotid arteries or the vertebral arteries. Cerebrospinal fluids were not xanthochromic. All hematological and laboratory data, as well as markers of thrombosis and fibrinolysis, were within normal limits: prothrombin time-international normalized ratio was 0.89, activated partial thromboplastin time was 30.4 seconds, anti-thrombin III was 70%, D-dimer was 0.5 μg/mL, and fibrin degradation products was 1.0 μg/mL. The patient was started on unfractionated heparin and oral warfarin, and her initial symptoms improved soon thereafter.

On transthoracic echocardiography, noncompaction of the left ventricle was demonstrated: hypertrabeculation and an abnormally thickened myocardium identified as a 2-layered structure were seen in the infero-lateral wall from the level of the papillary muscle to the apex (Fig. 3A). Left ventricular end-diastolic diameter was 55.0 mm, and the size of left atrium was 36.4 mm. Left ventricular systolic function was diffusely depressed: fractional shortening was 24.3%, and

¹Department of Medicine, Kumamoto Rosai Hospital, Kumamoto, ²Department of Cardiology, Kumamoto Rosai Hospital, Kumamoto and ³Department of Neurology, Graduate School of Medical Sciences, Kumamoto University, Kumamoto

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Correspondence to Dr. Makoto Nakajima, nakazima@fc.kuh.kumamoto-u.ac.jp
Figure 1. Diffusion-weighted magnetic resonance imaging (DWI; TR 6000, TE 69.9, b = 1000; A and B), fluid attenuation inversion recovery (FLAIR; TR 6000, TE 69.9; C and D) imaging, and magnetic resonance angiography (MRA; E) on admission. DWI demonstrates hyperintensity lesions in the right striatum and spotty ischemic lesions in the cerebral cortex (arrow heads). These changes are obscure on FLAIR image, but a branch of the right middle cerebral artery is demonstrated as a hyperintense signal (arrows). On MRA, a signal defect is seen at the distal site of the right middle cerebral artery, with incomplete visualization of the branches.

the ejection fraction was 45.0%. No valvular heart disease was seen. Transesophageal echocardiography revealed no thrombus or intracardiac tumor such as papillary fibroelastoma. Intracardiac right-to-left shunt was not detected by bubble studies. Intra-aortic plaques with an irregular surface were also not seen. On 24-hour Holter monitoring, PVCs were recorded 2,556 times and premature atrial contractions were recorded 20 times per 95,383 beats. Coronary flow velocity reserve as assessed on transthoracic color Doppler echocardiography was not decreased in the left anterior descending coronary artery. MRI demonstrated hypertrabeculation of the endocardium in the left ventricle (Fig. 3B). Since cerebral embolism associated with INVM was suspected, she was discharged home on oral anticoagulation. After 3 months, transthoracic echocardiography revealed that: fractional shortening had decreased to 15.1%; the ejection fraction had decreased to 41.7%; the left atrial diameter had expanded to 40.1 mm in the end-diastolic phase; and the left ventricular diameter was 57.0 mm in the end-diastolic phase.

Discussion

We reported a case of cardioembolic stroke presumably associated with INVM. Interestingly, 3 months after discharge, the patient’s left ventricular systolic function decreased rapidly (fractional shortening, which is thought to be suitable for the estimation of systolic function (5), a decrease from 24.3% to 15.1%), and the left atrium and were ventricle dilated. Previous observational studies of a small number of INVM patients had no cases that exhibited such a rapid course, although some embolic events were recorded
Figure 2. Chest radiography on admission. Mild cardiomegaly with a cardiothorax ratio of 60% was seen with no abnormal shadow in the lung field.

(1, 2, 6). In the present patient, as ventricular systolic function deteriorated, thrombi might have developed between the deep recesses. Although INVM is a malformation that is present since the fetal period, our patient’s first embolic event occurred at the age of 44 years when her ventricular systolic function had begun to deteriorate.

One of the important complications of INVM is systemic embolism, which can include cerebrovascular diseases, pulmonary embolism, or mesenteric infarction (7). Most patients with INVM develop embolic events after 40 years of age unless they have associated coagulation abnormalities (1, 5, 6). The proper time to start anticoagulation in patients with INVM is controversial. The mechanism of cardioembolic events in INVM is thought to be related to: the development of thrombi in the extensively trabeculated ventricle; depressed left ventricular systolic function; or the development of atrial fibrillation (3, 4, 7). Some authors recommend prophylactic anticoagulants regardless of the existence of thrombus or depressed left ventricular systolic function (2, 3). However, Stöllberger and Finsterer stated that left ventricular noncompaction does not cause systemic embolism unless associated with atrial fibrillation, systolic dysfunction or other risk factors for stroke or embolism (5). The present case suggests that the proper time to start anticoagulation in INVM patients may be when left ventricular systolic function decreases below normal.

Minor cardiac sources of embolism are important as causes of stroke in young patients (8). However, most neurologists would not likely be familiar with left ventricular noncompaction as a cause of embolic stroke. In a patient with ischemic stroke, if the left ventricular dysfunction is not recognized, antiplatelet agents may be given based on the diagnosis of cryptogenic stroke (9). Although the incidence of INVM appears to be not as high as previously reported (0.014%) (2), the correct diagnosis based on echocardiography may often be missed or delayed (7). Thus, the actual incidence has not been elucidated. It is important for neurologists to be aware of INVM as a potential source of adult embolic stroke.

Both familial and sporadic forms of INVM have been reported, and some inheritance of the disorder is suspected. Some genes responsible for several familial cases of INVM have been described such as a mutation in the G4.5 gene of the Xq28 chromosome region, or distal chromosome 5q deletion (7). Although we suspected some inheritance in the present case, the patient did not wish to undergo further exploration.

We reported a case of cardioembolic stroke associated with INVM. Further investigation is needed to determine the proper time to start anticoagulation in INVM patients.

References

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