treatment, steroids and oral immunosuppressants are the first line therapy for both LScs and PRS.[2,3]

While most patients with LScs involving the CNS experience their first neurological symptoms within a few years of the development of the sclerotic lesion, this report demonstrates that the first symptoms can occur even decades later. Accordingly, LScs needs to be considered when a patient with a long standing sclerotic lesion on the scalp and face presents with new onset neurological symptoms.

Jason K. Wasserman
University of Ottawa, Ottawa, Ontario, Canada

Rafael Glikstein, Michael Sharma
The Ottawa Hospital, Ottawa, Ontario, Canada
Email: msharma@ottawahospital.on.ca

References

1. Kister I, Inglese M, Laxer RM, Herbert J. Neurologic manifestations of localized scleroderma: a case report and literature review. Neurology. 2008 Nov 4;71(19):1538-45.
2. Korkmaz C, Adapinar B, Uysal S. Beneficial effect of immunosuppressive drugs on Parry-Romberg syndrome: a case report and review of the literature. South Med J. 2005 Sep;98(9): 940-2.
3. Kreuter A, Gambichler T, Breuckmann F, et al. Pulsed high-dose corticosteroids combined with low-dose methotrexate in severe localized scleroderma. Arch Dermatol. 2005 Jul;141(7):847-52.

To the Editor

Thrombolysis for Acute Ischemic Stroke in a Patient with Moyamoya Disease

Moyamoya disease (MMD) is an uncommon cerebrovascular condition that is characterized by progressive stenosis or occlusion of the terminal part of the internal carotid artery (ICA) together with abnormalities of the anterior and middle cerebral arteries.[1] A hallmark of this condition is the formation of fine net-like vessels at the base of the brain resulting from the compensatory development of a collateral vascular network.[1] The majority of affected adults and children present with ischemic symptoms, but there is still no specific treatment. Although surgical revascularization for MMD is an accepted and effective form of treatment for the prevention of further ischemic attacks,[2] it is not a fundamental treatment because the pathogenesis of this disease remain obscure.[2] In addition, there has been no consensus guideline nor well-organized clinical trials that assess the acute treatment options for MMD. Here, we describe a case of ischemic stroke in a patient with MMD who was treated with thrombolytic therapy. Thrombolysis in patients with MMD has rarely been reported.

Case Presentation

A 37-year-old woman presented to the emergency room with sudden-onset aphasia and right hemiparesis. Neurological examination revealed global aphasia, flattening of the left nasolabial fold, and weakness of the left arm and leg (4/5 strength). Her symptoms developed 1 hour prior to presentation and her National Institutes of Health Stroke Scale (NIHSS) score was 11. There was no remarkable history of medical illness. Her husband documented that both her parents had a stroke in their 50s. Urgent blood analysis and magnetic resonance imaging (MRI) were performed. The inclusion and exclusion criteria for the safe implementation of thrombolysis in hyperacute ischemic stroke were verified. The MRI of the brain showed increased signal in diffusion-weighted images (DWI) and low signal intensity in apparent diffusion coefficient maps in the territory of the left middle cerebral artery (MCA) and the left anterior cerebral artery (Figure 1). Intracranial magnetic resonance angiography (MRA) revealed occlusions of both supraclinoid ICAs and no visualization of their proximal branches. The patient was administered a 3.36 mg intravenous (IV) bolus of IV recombinant tissue plasminogen activator (rtPA) followed by a

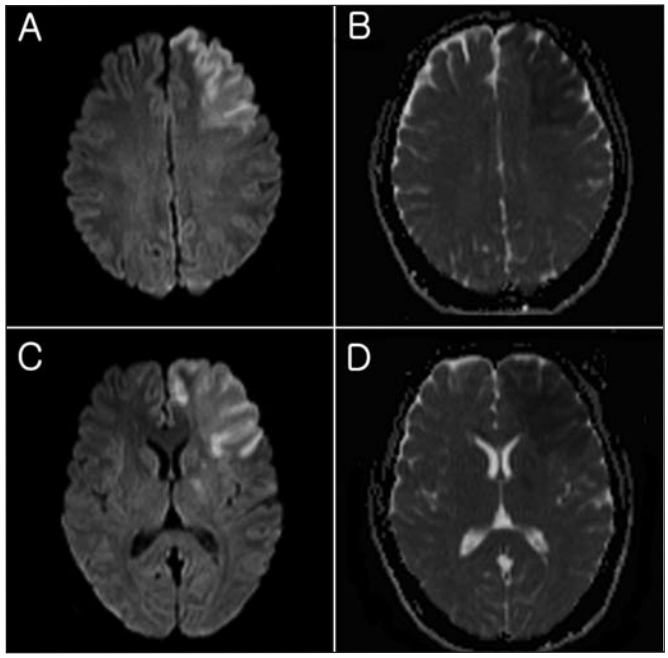

Figure 1: *MRI of the brain showing increased signal on DWI (A, C) and low signal intensity on ADC (B, D) maps in the territory of the left MCA and the left ACA. MRI, magnetic resonance imaging; DWI, diffusion-weighted image; MCA, middle cerebral artery; ACA, anterior cerebral artery.*

https://doi.org/10.1017/S0317167100018266 Published online by Cambridge University Press

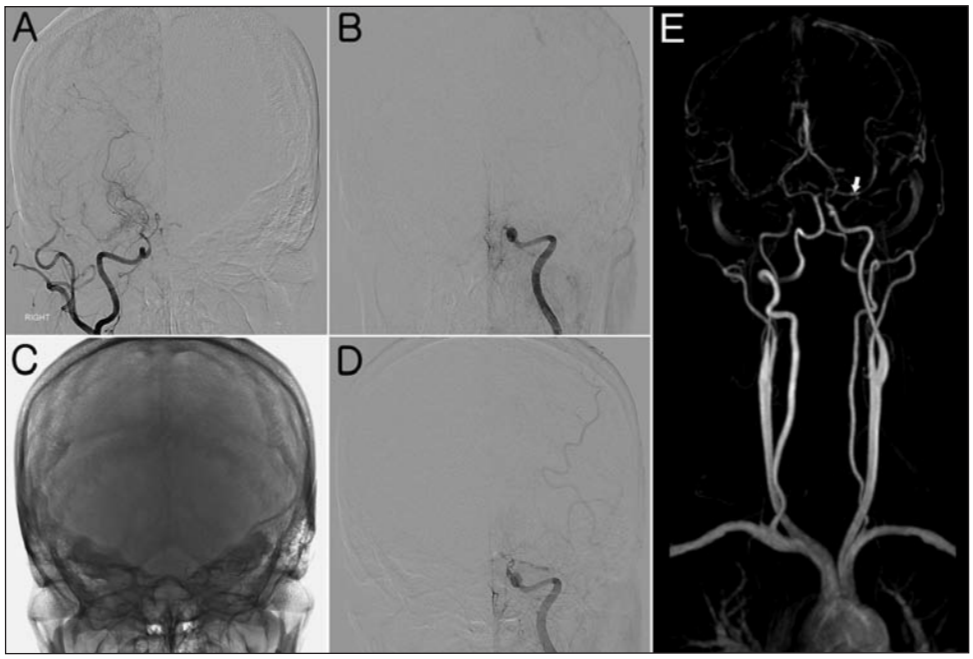

Figure 2: *(A) The right carotid angiogram showed occlusion of the terminal portion of the ICA with abundant fine vessels. (B) The left carotid angiogram showed occlusion of the supraclinoid ICA with a few collateral vessels. (C) Microcatheter guided IA mechanical thrombolysis was performed. (D) After thrombolysis, the terminal portion of the left supraclinoid ICA and proximal MCA were minimally recanalized, but intraluminal filling defects were noted. (E) Contrast-enhanced MRA of the brain showing revascularization of the occluded left ICA and left MCA (arrow). ICA, internal carotid artery; MCA, middle cerebral artery; MRA, magnetic resonance angiography.*

30.24 mg IV based on her weight (0.6 mg/kg for 60 minutes) in preparation for intraarterial (IA) thrombolysis. A subsequent cerebral angiography was performed after IV thrombolysis. The right carotid angiogram showed an occlusion of the terminal portion of the ICA with an abnormal network of fine vessels at the base of the brain (Figure 2). The left carotid angiogram also showed an occlusion of the supraclinoid ICA with a few collateral vessels (Figure 2). The above findings were compatible with MMD. We performed microcatheter guided IA mechanical thrombolysis on the assumption that the left ICA was the symptomatic occlusion (Figure 2). We did not perform stent assisted thrombectomy or other procedures, due to the risk of hemorrhage from the fragile vessels. On final angiogram, the terminal portion of the left supraclinoid ICA and the proximal MCA were minimally recanalized, but intraluminal filling defects were noted (Figure 2). The patient showed improvement immediately after the IV and IA thrombolysis. Her hemiparesis was fully recovered, and her post thrombolysis NIHSS score was 8. DWI performed 24 hours after the thrombolytic therapy showed acute infarction in the previously mentioned area and did not show hemorrhagic transformation or brain edema. Two days later, although her facial palsy had not resolved, her aphasia was considerably improved. An MRA of the brain was performed 4 days after the onset of symptoms, and it confirmed revascularization of the occluded left ICA and left MCA (Figure 2). After the acute ischemic stroke period, the patient was treated conservatively with 100 mg/day of aspirin. Her NIHSS score and modified Rankin score after one month were 1/1, due to aphasia.

Discussion

The cerebral angiographic findings presented in our patient are typical of MMD.[2] The patient was relatively young and had no cardiovascular risk factors. Her brain MRA showed no atherosclerotic changes of the proximal ICA and posterior circulation. Moyamoya syndrome, characterized as moyamoya vasculopathy with associated conditions, such as infection, autoimmune disease, meningitis, brain neoplasm, Down's syndrome, neurofibromatosis, head trauma, or irradiation to the head, was excluded.[2] All of the findings confirmed the diagnosis of MMD in our patient. As already noted, there are extremely few reported cases of IV rtPA use in acute ischemic stroke patients with MMD. This treatment might be considered a dangerous attempt because of the theoretical concerns regarding increased bleeding risk from the fragile collateral vessels. However, vascular abnormalities such as MMD were not excluded as initial indication for rtPA thrombolysis because the landmark NINDS-rtPA trial used computed tomography as the screening imaging modality.[3] Therefore, the true risk of systemic thrombolysis-precipitated intracranial bleeding is unknown in patients with MMD.

Our patient experienced no complications, and thrombolysis improved her neurological deficits. It is uncertain whether IV

https://doi.org/10.1017/S0317167100018266 Published online by Cambridge University Press

rtPA thrombolysis or IA thrombolysis played a larger role in the recanalization of the affected vessel in this patient, but the role of IA thrombolysis seemed to be limited because the arterial occlusion was minimally recanalized after IA thrombolysis. These findings suggest that the clotting system and fibrinolysis are associated with MMD, although the mechanism of action of rtPA in MMD is unclear. The pathophysiological mechanisms of MMD have not been fully elucidated. Pathological analysis has proposed that both MMD and cerebral atherosclerosis shared the same histopathological bases.[4] In MMD, the thrombotic components, such as fibrin, platelets and plasma constituents, formed microthrombi, which were considered to be a possible factor in the development of a thickened intima.[4,5] This pathogenesis of the intimal thickening in MMD was closely related to that of atherosclerosis and might lead to cerebral ischemia. Although further investigation is needed to determine the precise pathogenesis of MMD, the resolution of an ischemic stroke related to MMD with thrombolytic therapy can partially explain MDD pathogenesis.

Conclusions

Although proper long term treatment of MMD after the acute stroke period should be determined on a case-by-case basis, in acute ischemic stroke patient with MMD confirmed by an imaging modality, such as MRA or angiography, it is important that rtPA thrombolysis not be delayed due to our hemorrhagic concerns. Early diagnosis and prompt management are essential for minimizing the neurological deficits and lead to favorable outcomes. Although there can be limited generalization of our experience, thrombolysis should be considered as a treatment option for MMD patients who present with acute ischemic symptoms.

Hye Mi Lee, Mi-Yeon Eun, Woo-Keun Seo, Sang il Suh
Department of Neurology (HML, MYE, WKS) and Radiology (SIS), Korea University College of Medicine, Seoul, Korea
Email: nukseo@korea.ac.kr

References

1. Suzuki J, Takaku A. Cerebrovascular "moyamoya" disease. Disease showing abnormal net-like vessels in base of brain. Arch Neurol. 1969;20(3):288-99.
2. Kuroda S, Houkin K. Moyamoya disease: current concepts and future perspectives. Lancet Neurol. 2008;7(11):1056-66.
3. The National Institute of Neurological Disorders and Stroke rt-PA Stroke Study Group. Tissue plasminogen activator for acute ischemic stroke. N Engl J Med. 1995;333(24):1581-8.
4. Weinberg DG, Arnaout OM, Rahme RJ, et al. Moyamoya disease: a review of histopathology, biochemistry, and genetics. Neurosurg Focus. 2011;30(6):E20.
5. Yamashita M, Oka K, Tanaka K. Cervico-cephalic arterial thrombi and thromboemboli in moyamoya disease--possible correlation with progressive intimal thickening in the intracranial major arteries. Stroke. 1984;15(2):264-70.

To the Editor

Multiple Brain Cysts: An Unusual Form of Radiologically Isolated Syndrome

We have recently reported a patient with multiple brain cysts who after extensive investigations, proved to be have unusual form of radiologically isolated syndrome (RIS).[1]

However due to relatively limited follow up (six months) and in absence of confirmation of new disease activity (clinically or by MRI) we could not with certainty exclude an unusual case of ADEM. Although ADEM patients almost always have abrupt clinical presentation with disturbance of consciousness and/or seizures, it remains possible that some ADEM presentations could be subtle.[2] Follow-up of such patients is important because in cases of MS, the clinical course (developing of new symptoms) and MRI appearance (shrinkage of lesions and appearance of new lesions, usually more consistent with MS) will change.

Here we report follow-up MRI findings of our patient after 15 months, showing two new lesions, one in the white matter of the right frontal lobe, and other in subcortical white matter of the left postcentral region (Figure). Both lesions show irregular rim enhancement after application of the contrast medium. There were no infratentorial lesions and previously present cystic

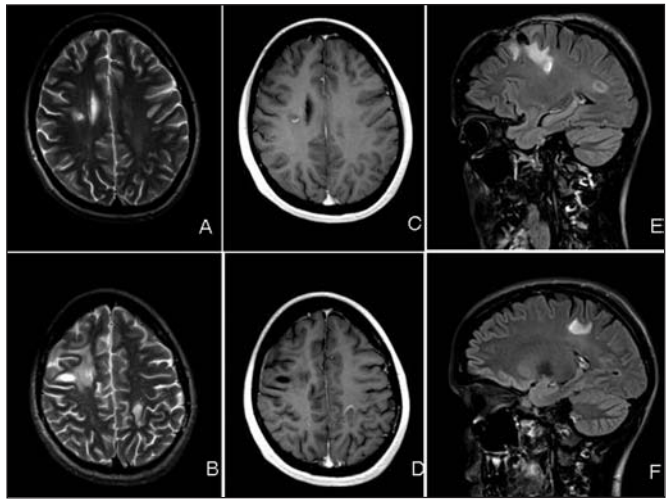

Figure: *Brain MRI; A) and B) T2-wighted images; C) and D) T1-weighted sequences following gadolinium application; E) and F) FLAIR images; Comparing to previous MRI (see reference 1) two new lesions are seen, one in the white matter of the right frontal lobe, and other in subcortical white matter of the left postcentral region. Both lesions are showing irregular rim enhancement after application of the contrast medium. Previously present cystic lesions are significantly smaller and paler.*

https://doi.org/10.1017/S0317167100018266 Published online by Cambridge University Press