TO THE EDITOR

45-Years Between Skin Lesions and CNS Symptoms in a Patient with Scleroderma

A 50-year old woman presented to her family doctor after a single episode of weakness in her left hand and numbness in her left hand and face. The symptoms resolved five minutes after onset and she was not left with any residual deficits. She was subsequently referred to our Stroke Clinic for evaluation. The patient had never experienced similar symptoms in the past nor did she have a history of migraines, seizures, traumatic brain injury, stroke, heart disease, or cancer. The patient had one sister with multiple sclerosis. On examination she was alert and oriented. Inspection revealed an area of second skin above the right eyebrow and widening of the right orbit (Figure, A). A linear scar was palpable from the right eyebrow to the right parietal bone. The patient recalled that the lesion developed spontaneously when she was five and had remained static since that time. Her family physician at that time referred to the lesion as ‘en coup de sabre’ although no subsequent neurological investigations were performed. A neurological examination performed in the Stroke Clinic was within normal limits. Because seizures have frequently been reported in patients with linear scleroderma ‘en coup de sabre’ (LScs), we considered the possibility that the recent episode was a partial seizure and a computed tomogram (CT) scan was performed to look for a potential focus. The CT scan showed a hypodensity and thickening of the cortex in the right frontal lobe and an enlarged right ventricle (Figure, B). A CT angiogram did not reveal any vascular abnormalities. A magnetic resonance image (MRI) revealed an extensive area of T2 (Figure, C) and fluid-attenuated inversion recovery (FLAIR) (Figure, D) hyperintensity involving the right frontal and temporal subcortical white matter, and the periventricular white matter. Abnormal sulcal and intravascular effacement was also observed. A cerebral angiogram revealed no abnormalities consistent with vasculitis. A follow-up MRI with gadolinium performed three months later did not show any enhancement or changes from the previous scan. An electroencephalogram performed was within normal limits. Based on the results of these investigations, the patient was started on carbamazepine for seizure prophylaxis and five months after presenting to our clinic she has not experienced any new events.

This report describes a 45-year delay between the development skin lesions and the onset of neurological symptoms in a patient with LScs. To the best of our knowledge, this is the longest delay ever reported. A recent review of patients with LScs and central nervous system (CNS) involvement found an average delay of 4.3 years between skin and neurological symptoms and that the average age of CNS symptom onset was 13.8. The neurological symptoms experienced by our patient were most likely a focal seizure which is consistent with previous reports. Indeed seizure activity is the most common neurological finding in patients with LScs. Because our patient had not undergone any brain imaging in the past, we considered the possibility that her neurological symptoms occurred as a result of a newly acquired brain lesion unrelated to her scleroderma. However a cerebral angiogram ruled out the presence of vasculitis and the MRI findings were inconsistent with a tumor. In contrast, her imaging findings were highly typical of patients with LScs. For example, focal atrophy and T2 hyperintensities are found in the majority of patients. Brain lesions are ipsilateral to the skin lesions in 88% of patients; most occur in the subcortical white matter. The presence of seizure activity after so many years could signal the development of new lesions, however we did not find any evidence of an acute process and the patient did not have any further episodes.

LScs shares many features with a related disorder called Parry-Romberg syndrome (PRS) and controversy exists as to whether the two conditions are in fact distinct entities. While our patient was originally diagnosed with LScs, MRI results demonstrated muscle and bone atrophy underlying the sclerotic lesion, a finding that is more consistent with PRS. However this delineation may be only academic as the two disorders coincide in up to 34% of patients. Although our patient did not require

Figure: Skin and brain lesions in a patient with linear scleroderma ‘en coup de sabre’. (A) The characteristic linear scar can be seen running from the right orbit to the right parietal region. Subdermal atrophy was also noted under the scar. (B) A non-contrast CT scar showing a region of hypodensity and thickening of the cortex in the frontal lobe underlying the skin lesion (B). The right ventricle was also enlarged. (C and D) MRI revealed hyperintensities on T2 (C) and FLAIR (D) images in the right frontal and temporal subcortical white matter and periventricular white matter.
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.

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References

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 MCA and the left ACA. MRI, magnetic resonance imaging; DWI, diffusion-weighted image; MCA, middle cerebral artery; ACA, anterior cerebral artery.

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.