



Self-Injurious Behavior Revealing Advanced Primary Progressive Multiple Sclerosis with a Massive Right Temporal Lesion

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Dear Editor,

A 40-year-old left-handed carpenter working in an art gallery was admitted for investigation of disabling walking difficulties due to imbalance and shaky legs that had insidiously evolved over the previous year. Physical examination found extended hyperkeratotic skin lesions consistent with lichenification (lichen simplex chronicus) on both upper extremities with radial distribution (Fig. 1). Clinical observation uncovered compulsive self-biting of both arms during unobserved moments. Further medical history-taking revealed that the self-injurious behavior (SIB) had started around 7 years previously. Not only did the patient bite his hands and arms, but he also poked his eyes, banged his head, and shouted loudly when he was alone, to such an extent that he was expelled from his apartment due to nocturnal noise. He explicitly denied itching in his arms, and related his behavior to intermittent feelings of loneliness, but denied being depressed.

Neurological examination revealed an attentive and friendly man with unremarkable behavior showing cerebellar dysarthria and ataxia and tetrapyramidal signs that caused his unsteady ataxospastic gait, but with no sensory difficulties in the arms, legs, or trunk. Brain and spine MRI showed extensive periventricular white-matter damage in the right temporal lobe (Fig. 2A), as well as multiple demyelinating lesions juxtacortical in the corpus callosum, pons, midbrain, and cerebellum (Fig. 2B), some of which showed also Gd+ contrast enhancement (Fig. 2C). However, there were virtually no lesions in the frontal lobe.

An extended infectiological and immunological workup did not find any evidence of acute or chronic infection (including PCRs of the CSF), nor of autoimmune connective-tissue disorder or paraneoplastic disease. The only deficit in vitamin levels was in vitamins B₁₂ and D, for which supplements were administered. Flow cytometry findings were unremarkable. CSF analysis showed a white blood cell count of 15,000/ μ L (lymphocytes), elevated proteins (0.61 g/L) with a highly elevated albumin quotient at 8.36×10^{-3} , positivity for oligoclonal bands (IgG index 1.3), and normal interleukin-6. A neuropsychological evaluation revealed memory problems (visuospatial, verbal, short-term, and working memory), and marked executive difficulties (mental inflexibility and psychomotor slowing). Based on the pattern of the brain and spine lesions fulfilling McDonald's revised criteria, we diagnosed primary progressive multiple sclerosis (PPMS) with radiological progression based on the neurological symptoms (notably walking difficulties) for longer than 1 year combined with progressive accumulation of handicap without recovery. An offline treatment with rituximab was introduced.

SIB is an intriguing human behavior that is seen in a range of neurological and psychiatric disorders but remains poorly understood. It occurs most frequently in developmental disorders and in psychiatric or neurological conditions involving striatofrontal dysfunction,

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such as in Tourette's, Lesch-Nyhan, Rett's, Prader-Willi, or fragile-X syndromes.¹ However, anterior temporal dysfunction has also been reported as an SIB-causing condition in patients with frontotemporal dementia² or temporal lobe epilepsy.³ Very little is known about SIB in multiple sclerosis. One neuroimaging study described damage to the right inferior and middle temporal gyri and the inferior frontal cortex as being associated with compulsive behavior in 16 patients with multiple sclerosis.⁴ Another study suspected that right temporo-parietal injury can result in actions related to SIB by disconnecting the networks of agency and body ownership, often in the presence of an existing focal neurological deficit (e.g., limb paresthesia).⁵

In our case, SIB over a period of years resulted in extended chronic lichenification on the patient's arms, hands, and fin-



Fig. 1. The patient's arms, hands, and fingers were covered with hyperkeratotic skin lesions consistent with lichen simplex chronicus.

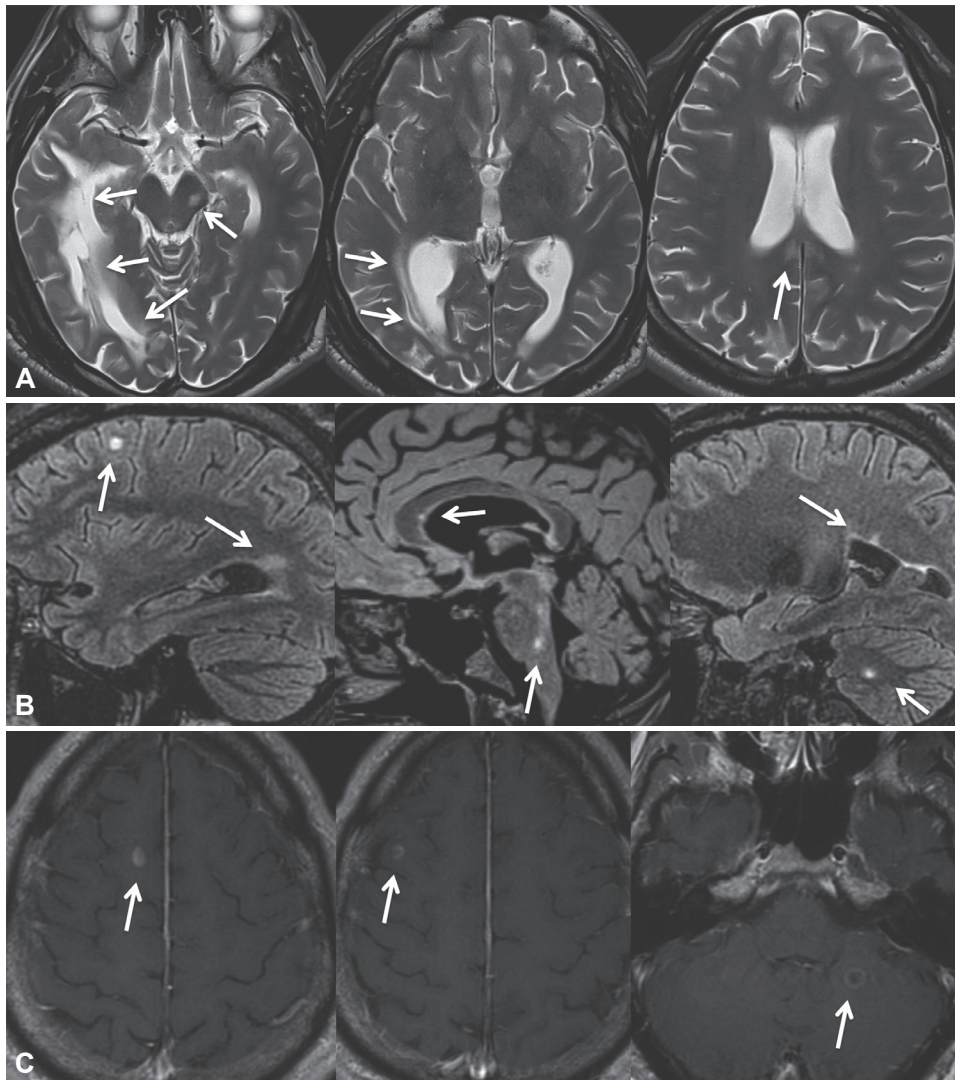


Fig. 2. Magnetic resonance imaging of the patient's brain. A: Transverse T2-weighted images show an extensive periventricular white-matter damage in the right temporal lobe (arrows). B: Sagittal FLAIR images show multiple demyelinating lesions juxtacortical in the corpus callosum, pons, midbrain, and cerebellum (arrows). C: T1-weighted images with Gd+ show lesions with contrast enhancement (arrows). Images displayed in accordance with the usual radiological convention. FLAIR: fluid attenuated inversion recovery.

gers. He only sought medical advice after walking difficulties became disabling. The first brain MRI led to the discovery of advanced PPMS with an unusually massive white-matter lesion in the right temporal lobe. Evidence can be found in the literature that establish a causal link of this lesion to the patient's SIB.

Our case highlights the utility of early brain MRI in cases of newly appearing SIB, since there is increasing recognition that focal brain lesions—due to potentially treatable conditions—can be implicated in SIB.

Conflicts of Interest

The authors have no financial conflicts of interest.

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