The "Motor Band Sign:" Susceptibility-Weighted Imaging in Amyotrophic Lateral Sclerosis

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The diagnosis of amyotrophic lateral sclerosis (ALS) requires detection of both upper motor neuron (UMN) and lower motor neuron (LMN) degeneration. Electromyography effectively detects LMN degeneration, but there is no definite neuroimaging technique for demonstrating UMN involvement. This is of concern because clinical UMN signs may not be evident until late in the disease course. Increased T2 signal changes have been reported along the course of corticospinal tracts from the subcortical precentral gyrus through the centrum semiovale, posterior limb of internal capsule, cerebral peduncles, pons, medullary pyramids, and anterior and lateral corticospinal tracts of the spinal cord.[1,2] These magnetic resonance (MR) changes have been thought to represent corticospinal degeneration; however, they are not sensitive or specific for ALS and may be associated with normal aging.[3,4] Susceptibility-weighted imaging (SWI) is a new high-resolution, three-dimensional gradient echo MR imaging (MRI) technique[5] that may be more effective in detecting changes in motor cortex and thus likely provide supportive evidence of UMN involvement.

We report two cases of ALS with striking SWI scans revealing an abnormal hypointense T2 signal in the precentral gyrus. We called this finding the "motor band sign" because of its band-like appearance in the motor cortex.

Case Reports

Clinical and Laboratory Findings: Case 1

A 60-year-old woman presented with a two-year history of progressive asymmetric painless weakness. In the first year, symptoms were limited to the left leg, and the clinical findings were felt to be consistent with a focal upper motor neuron disorder. In the second year, similar weakness appeared in the right leg, then to both arms. The patient also reported increasing dysarthria and dysphagia. There was no history of trauma or family history of neurological disease.

When examined in 2010 (at the time of MRI), the patient had a normal mental status. The only cranial nerve dysfunction was a spastic dysarthria and increased gag reflex. There was mild atrophy of the left leg. Infrequent fasciculations were noted in both arms and both legs. There was a spastic quadriparesis that was more pronounced on the left side. Weakness was graded as more severe in the legs (Medical Research Council grades in the range of 4/5 proximally, 2-3/5 distally) than in the arms (mostly Medical Research Council grade 4-/5). There was diffuse hyperreflexia, with bilateral ankle clonus, although plantar responses were bilaterally flexor on repeated assessments. The sensory examination was normal.

Motor and sensory nerve conduction studies were within normal limits, with the exception of reduced median nerve compound muscle action potential amplitude. Monopolar electromyelograph showed patchy abnormalities in keeping with active denervation (positive sharp waves, fibrillations, reduced recruitment) in the biceps, triceps, and tibialis anterior muscles. In the gastrocnemius and first dorsal interosseus, there were abnormalities suggesting chronic reinnervation (large amplitude, long duration motor unit potentials). Other muscles showed decreased voluntary activation. Fasciculation potentials were noted in three muscles.

The following blood tests were normal or negative: complete blood count, electrolytes, renal and liver function studies, Venereal Disease Research Laboratory, antinuclear antibodies, and HIV serology. Visual evoked potential latencies, ordered when demyelinating disease was an initial consideration, were also normal. MRI of the cervical, thoracic, and lumbar spine showed no significant central stenosis or abnormal cord signal.

At the time of her 2010 MRI study, the patient was felt to have motor neuron disease with a predominantly upper motor neuron clinical picture. She was classified as clinically probable ALS according to Revised El Escorial criteria.[6] The patient was still alive four years after her 2010 MRI, though she had progressed to anarthria and marked quadriparesis, requiring a wheelchair for ambulation and support for all activities of daily life.

Clinical and Laboratory Findings: Case 2

A 58-year-old man with a history of alcoholic cirrhosis presented with a two-year history of insidiously progressive and painless weakness of the right arm and leg. For the previous six months, he had also noticed frequent fasciculations in all of his limbs. He had not noticed any cognitive impairment and reported

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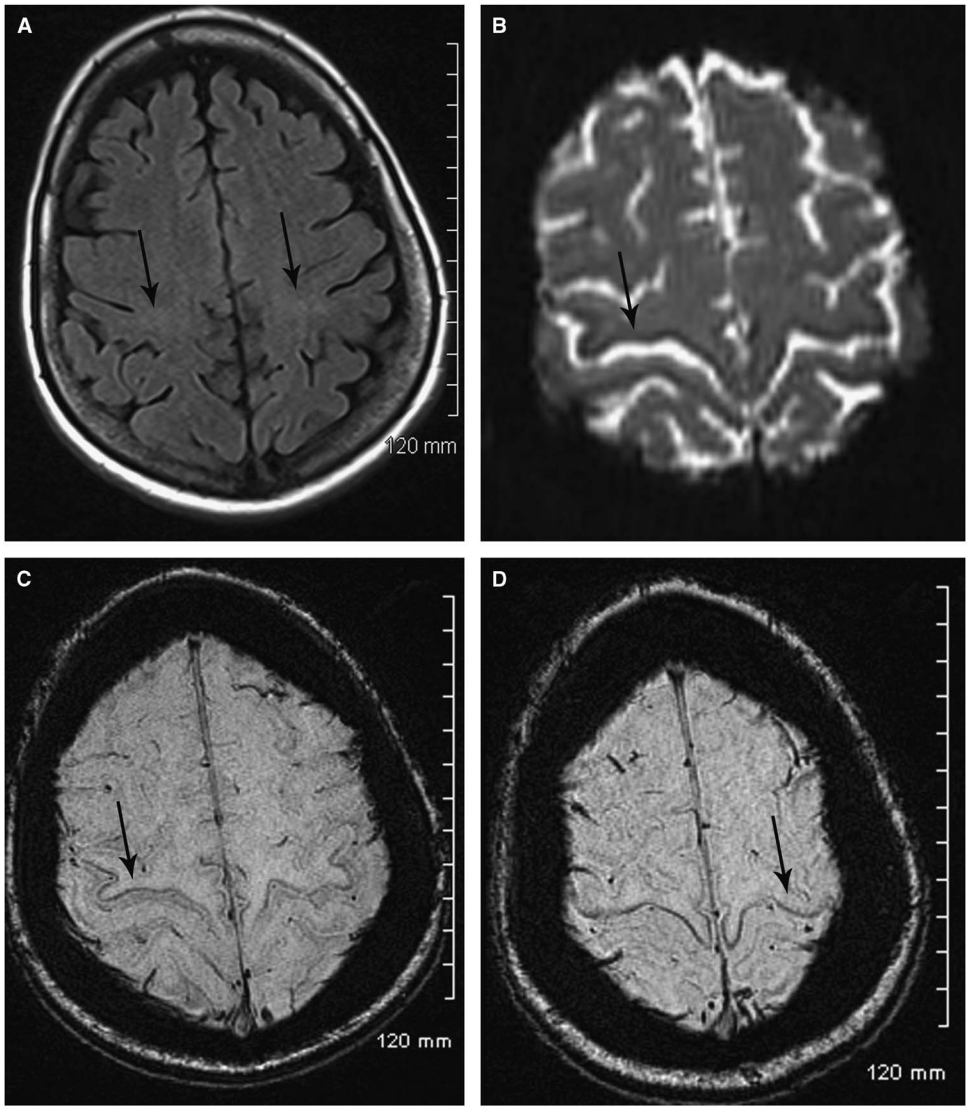

Figure 1: *Case 1. Axial fluid-attenuated inversion recovery image (A) shows subtle high signal along the corticospinal tracts bilaterally. Axial EPI T2 image (1B) shows an area of hypointensity along the motor cortex. Axial SWI images (C, D) show typical "motor band sign" with hypointensity exclusively along the posterior cortex of the precentral gyrus (black arrows). Please note adjacent gyri do not show this change.*

no visual, sensory, or autonomic symptoms. There was no relevant family history of neurological disease.

When examined at the time of his MRI study, the patient was lucid and coherent. The cranial nerve examination was normal except for slight relative atrophy of the left border of the tongue, mild effacement of the left nasolabial fold, and diminished left palatal movement. The sensory examination was normal. Infrequent fasciculations were noted in both arms and the right leg. There was marked spasticity of the right arm and leg. Motor strength testing revealed a right hemiparesis, in the range of Medical Research Council 4/5, following an upper motor neuron distribution pattern (deltoid, triceps, finger extension, hip and knee flexion, ankle dorsiflexion). There was asymmetric but diffuse hyperreflexia, more evident on the right side, with a right Babinski sign and flexor plantar response on the left.

Motor and sensory nerve conduction studies were within normal limits. Monopolar electromyelograph revealed mild but definite changes of active denervation (fibrillations, positive sharp waves, and reduced recruitment) in the following muscles: right first dorsal interosseus, right tibialis anterior, right medial gastrocnemius, and thoracic paraspinal muscles. Changes of chronic reinnervation were noted in several other muscles (right and left vastus medialis, medial gastrocnemius).

Routine biochemical and hematological studies were normal, with the exception of a mild elevation in aspartate aminotransferase (57 U/L), and total bilirubin (17 μmol/L). Computed tomography of the abdomen was consistent with cirrhosis of the liver. MRI of the cervical spine showed mild changes of degenerative disc disease without significant foraminal or central stenosis.

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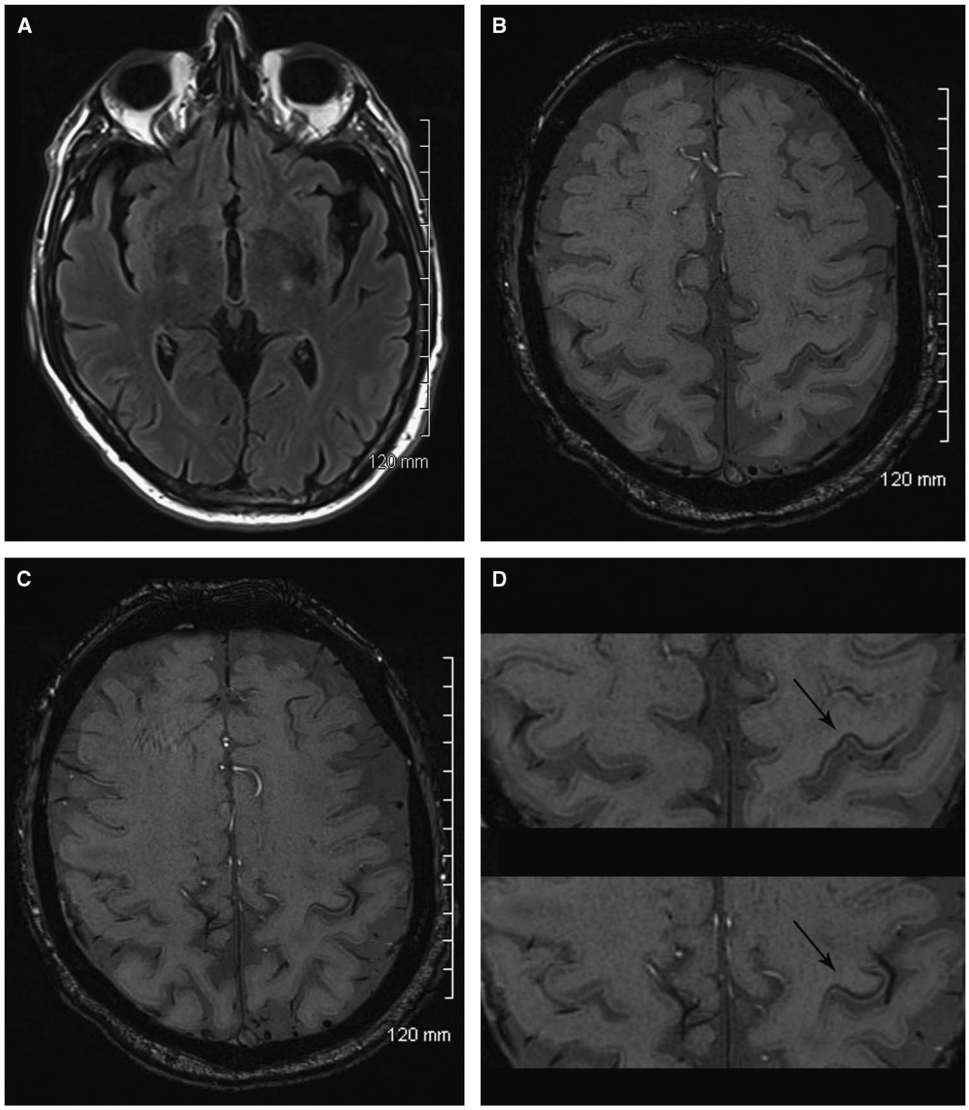

Figure 2: *Case 2. Axial fluid-attenuated inversion recovery image (A) shows high signal along the corticospinal tracts (internal capsule) bilaterally. Axial SWI images (B, C) show the typical "motor band sign" with hypointensity exclusively along the posterior cortex of the pre-central gyrus mainly involving the left side (patient had prominent right hemibody upper motor neuron involvement). Magnified images (D) highlight the findings on the left side (black arrows).*

At the time of the MRI, the patient was classified as clinically definite ALS according to Revised El Escorial criteria. He passed away from respiratory failure 18 months later.

Imaging Findings

In both patients (Figures 1, 2), fluid-attenuated inversion recovery and to a lesser extent T2-weighted MRI studies showed subtle bilateral hyperintensity along the expected course of the corticospinal tracts: subcortical frontal white matter, posterior limb of the internal capsule, cerebral peduncles and pons. Both computed tomography and MRI showed a variable degree of atrophy involving the primary motor cortex with prominence of the adjacent central sulcus.

More significantly, axial SWI images revealed linear hypointensity along the cortical margin of the precentral gyrus, distinctly limited to its posterior border. This was clearly present bilaterally (Figure 1C, D) in case 1, but affected mostly the left precentral gyrus (Figure 2D) in case 2. Corresponding images on axial echo-planar imaging (EPI) T2 sequences (Figure 1B) also showed this linear hypointensity, although less sharply demarcated. This discrepancy is expected because of the greater spatial resolution and signal-to-noise ratio of SWI compared with EPI. Adjacent gyri, notably the superior, middle, and inferior frontal gyri as well as the parietal postcentral gyri, did not show similar abnormalities.

Discussion

ALS is a progressive neurodegenerative disease characterized by LMN and UMN dysfunction. The diagnosis is currently based on clinical assessment, supported by electrodiagnostic studies and exclusion of other disease mimics such as compressive

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myelopathy. There is no reliable paraclinical marker for UMN involvement in ALS. A method that detects early UMN involvement and accurately monitors disease progression is highly desirable especially for future clinical trials proposing early institution of disease-modifying therapies.

Axonal degeneration in corticospinal tracts in patients with ALS is difficult to detect reliably using conventional MRI sequences.[3] Recently several authors have reported using newer techniques such as magnetic resonance spectroscopy, magnetization transfer, diffusion tensor imaging, and fiber tractography to detect corticospinal tract degeneration in patients with ALS.[7-9]

SWI is a new MRI technique[5] that uses the differences of susceptibility between tissues. It is a high-resolution, three-dimensional gradient echo sequence that uses magnitude and phase data both separately and added together after appropriate filtering to enhance the paramagnetic properties that underlie tissue susceptibility.[5] It has been shown to be very sensitive to iron in the form of hemosiderin, ferritin, intracellular methemoglobin, and deoxyhemoglobin. In this report, we described the ability of SWI to detect T2 hypointensity in the motor cortex in two patients with ALS that likely correlate with UMN involvement. T2 hypointensity in the motor cortex of ALS patients has been described before[10-13], but the increased resolution of SWI makes it visible within the motor cortex in a band-like fashion within the gray matter.

Interestingly, we found that SWI revealed hypointense T2 signal in the motor cortex bilaterally in the first patient and in the left motor cortex of the second patient, consistent with the clinical finding of bilateral spasticity in the first patient and predominantly right sided spastic hemiparesis in the second patient. The striking feature in SWI in our patients was exclusive involvement of the motor cortex (Brodmann's area 4 or primary motor cortex M1)[14] in a band-like fashion. The adjacent cortex (including the anterior lip of the precentral gyrus) did not show similar hypointensity. The presumed cause of low T2 intensity is iron deposition in the motor cortex[10,15].

Low T2 intensity is also often observed in aged patients and patients with various other neurological diseases. Imon et al[16] have reported that its incidence was significantly higher in patients with central nervous system disorders such as Alzheimer disease, Parkinson disease, and multiple cerebral infarctions and was thus not specific for any particular disease process. Nonetheless, a specific pattern of motor cortex involvement might be a clue to UMN involvement in ALS. In our experience, these changes are not invariably seen in every ALS patient and they may be variably expressed in relation to the disease course and the degree of clinical UMN involvement. This heterogeneity is an important challenge in the diagnosis of ALS, perhaps reflecting the fact that iron deposition is not a consistent neuropathologic finding.[17] Larger prospective studies will be needed to determine the incidence, sensitivity, and specificity of the motor band sign in ALS and whether it is a useful prognostic indicator.

Conclusion

We found very striking abnormal hypointense T2 signal exclusively in the posterior motor cortex of the precentral gyrus in two patients with ALS on SWI. SWI has the potential to recognize corticospinal degeneration in ALS; however, further studies are required to determine the accuracy of this finding in clinical practice.

Disclosures

Santanu Chakraborty, Aparna Gupta, Thanh Nguyen, and Pierre Bourque do not have anything to disclose.

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