Corticospinal Tract Degeneration in Motor Neuron Disease

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Summary: Long-repetition-time spin-echo MR images showed symmetric hyperintensity of the intracerebral corticospinal tracts in two patients with clinical and neurophysiologic diagnosis of primary lateral sclerosis and amyotrophic lateral sclerosis. In both, axial low-flip-angle gradient-echo images of the cervical spine showed hyperintensity of the lateral columns of the cord consistent with antegrade degeneration of the crossed corticospinal tracts.

Index terms: Degenerative disease; Sclerosis; Spine, magnetic resonance

Two variants of motor neuron disease, primary lateral sclerosis and amyotrophic lateral sclerosis, can be distinguished from each other on the basis of pathologic, clinical, and neurophysiologic findings (1). In primary lateral sclerosis, cellular loss and damage is confined to the motor neurons in the pyramidal cortex, and the clinical picture is that of progressive spastic paresis with delayed conduction in the corticospinal tracts on motor evoked potentials. In amyotrophic lateral sclerosis, spinal cord motor neurons also are damaged, and muscular wasting with fasciculations and abnormal potentials at electromyography are added to the clinical and neurophysiologic picture.

Magnetic resonance (MR) features of antegrade degeneration of the corticospinal tracts in the brain of patients with motor neuron disease are well established (2–4). We report the MR appearance of degeneration of the cervical corticospinal tracts in two patients with motor neuron disease.

Case Reports

Case 1

A 42-year-old man was referred for an MR examination because of progressive spastic tetraparesis, more prominent on the left side. He had no familial history of neurologic disorders and had been well until 1987, when he noted progressive weakness of the left lower limb. At that time, MR of the cervical spine showed herniation of the C5-6 intervertebral disk without cord compression. Weakness extended to the left upper limb, and in 1990 he underwent diskectomy at C5-6 without any benefit. Clinical examination in 1992 showed spastic paraparetic gait with diffusely increased deep tendon reflexes, bilateral Babinski sign, dysphagia, dysarthria, and inappropriate crying and laughing. No sensory deficit was present. Blood and cerebrospinal fluid analyses were normal. Electromyography and sensory evoked potentials were normal. Examination of motor evoked potentials showed bilateral slowing of conduction in the corticospinal tracts consistent with a cliniconeurophysiologic diagnosis of primary lateral sclerosis. Cranial and cervical spine MR examination at 0.5 T showed symmetric signal changes in the corticospinal tracts from the centrum semiovale to the lower portion of the cervical spinal cord (Fig 1).

Case 2

A 47-year-old woman without familial neurologic disorders had progressive dysarthria and dysphagia. In addition, she had noted diffuse muscular twitching and cramps in the lower limbs. Neurologic examination revealed wasting and weakness of the small muscles of the hands, diffusely increased deep tendon reflexes, clonus of the jaw, and frequent fasciculations. No sensory deficits were detected. Electromyography showed mild chronic denervation with abundant fasciculation potentials. Bulbar palsy, wasting of the hand muscles, and a spastic paraparetic gait were noted 4 months after the neurologic examination. Examination of motor evoked potentials showed bilateral slowing of the conduction in the corticospinal tracts. She was referred for MR with a cliniconeurophysiologic diagnosis of amyotrophic lateral sclerosis. MR examination of the cervical spine at 0.5 T showed symmetric signal changes in the crossed corticospinal tracts (Fig 2). Cranial MR examination (Fig 2) showed extension of the abnormality through the brain stem to the centrum semiovale.

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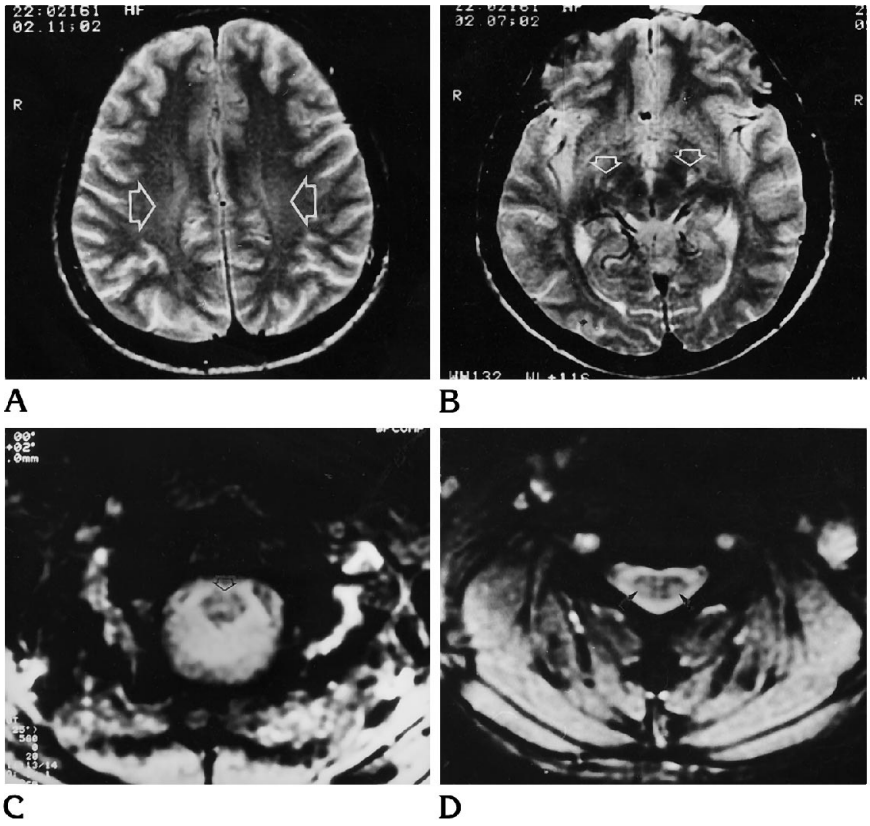

Fig 1. Case 1 (primary lateral sclerosis).

A and *B,* Axial T2-weighted spin-echo MR images (1800/100/2 [repetition time/echo time/excitations]) show symmetric hyperintensity (*arrows*) in the centrum semiovale (*A*) and in the cerebral peduncles (*B*).

C and *D,* Axial flow-compensated gradient-echo images (560/20/4) with a 25° flip angle at C-1 (*C*) and C-5 (*D*) show abnormal symmetric hyperintensity in the anterior portion of the cervicomedullary junction (*arrow*) and in the lateral columns of the spinal cord (*arrowheads*).

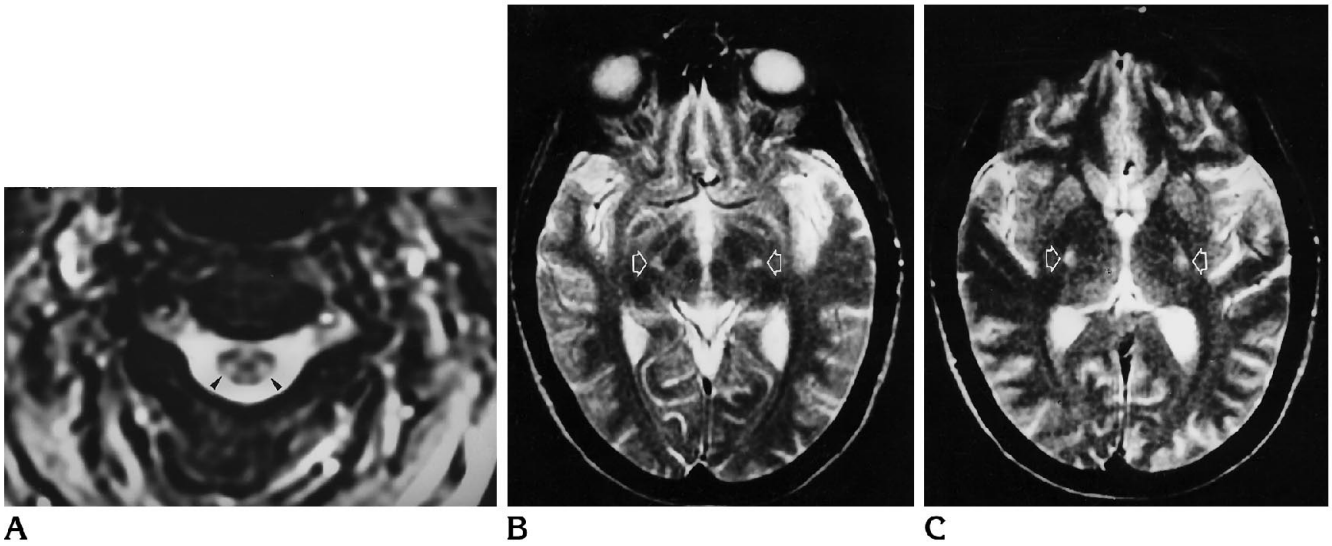

Fig 2. Case 2 (amyotrophic lateral sclerosis).

A, Axial flow-compensated gradient-echo MR image (400/30/6) with a 25° flip angle at C-3 shows symmetric hyperintensity in the lateral columns of the spinal cord (*arrowheads*).

B and *C,* Axial T2-weighted spin-echo images (2240/100/2) show symmetric circumscribed hyperintensity (*arrows*) in the cerebral peduncles (*B*) and in the internal capsules (*C*).

Discussion

Histopathologic examination of motor neuron disease shows extensive, variable, antegrade degeneration of the corticospinal tracts that can be tracked from the cerebral cortex to the conus medullaris (5). Recently, the potential of MR to show in vivo degeneration of the white matter tracts in the cervical spinal cord of patients with vitamin B_{12} deficiency (6), with Friedreich ataxia (7), or after intracerebral hemorrhage (8) has been cited. Our observations extend these potentials of MR to motor neuron disease and indicate that MR can contribute to the diagnosis of this condition not only by excluding other causes of spastic paraparesis with or without amyotrophy but also by demonstrating intramedullary white matter signal abnormalities matching the histopathologic changes typically observed in the spinal cord in cases of motor neuron disease.

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