Neuroimaging Highlight

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Amyotrophic Lateral Sclerosis

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Patient 1 (Figure 1) is a 48-year-old female who presented with a three to four year history of fatigue and right leg cramps, which progressed to severe muscle spasms and difficulty walking. EMG suggested Amyotrophic lateral sclerosis (ALS). She has subsequently developed bilateral upper and lower extremity and truncal weakness, and some difficulty swallowing.

Patient 2 (Figure 2) is a 31-year-old male who presented with right index finger weakness. This progressed over one month to involve the entire right arm, the neck, and the left arm. Physical examination also demonstrated truncal and lower extremity weakness, as well as mildly impaired speech and swallowing. Upper and lower motor neurons were affected, with no sensory involvement. EMG suggested ALS.

ALS, the most common form of motor neuron disease, is a progressive neurodegenerative disorder of unknown etiology. Most cases are sporadic but a small percentage are inherited in an autosomal dominant fashion. Diagnosis is clinical, and based on El Escorial criteria, which include the presence of upper and lower motor neuron signs and the progression of abnormalities over at least six to 12 months after disease onset. No autonomic, sensory, or cognitive involvement should be present.

Although the primary role of imaging is to exclude other diseases mimicking ALS, some characteristic MR findings have been described in this patient group. These are not present in all patients. Abnormal signal on T2-weighted images, including proton density and FLAIR, can be visualized extending throughout the course of the corticospinal tracts, usually in a bilateral symmetric fashion. Although this focal abnormal signal is best visualized in the posterior limb of the internal capsule and the cerebral peduncles, abnormally increased T2 signal can be seen extending from the precentral subcortical white matter to the anterolateral columns of the spinal cord. Abnormally decreased T2 signal has also been described in the motor cortex, possibly secondary to iron deposition. In normal studies, T2-weighted MR images demonstrate slightly increased signal in the corticospinal tract within the posterior limb of the internal capsule, relative to surrounding structures. In many ALS patients, however, this signal difference is markedly accentuated. Although pathologically there is asymmetric and variable involvement of the corticospinal tracts, patients with positive MR studies tend to demonstrate markedly symmetric abnormalities. This may reflect the severity of disease in patients with abnormal MR examinations.

Differential diagnosis of abnormal T2 signal in the corticospinal tracts is limited. Similar symmetric findings have

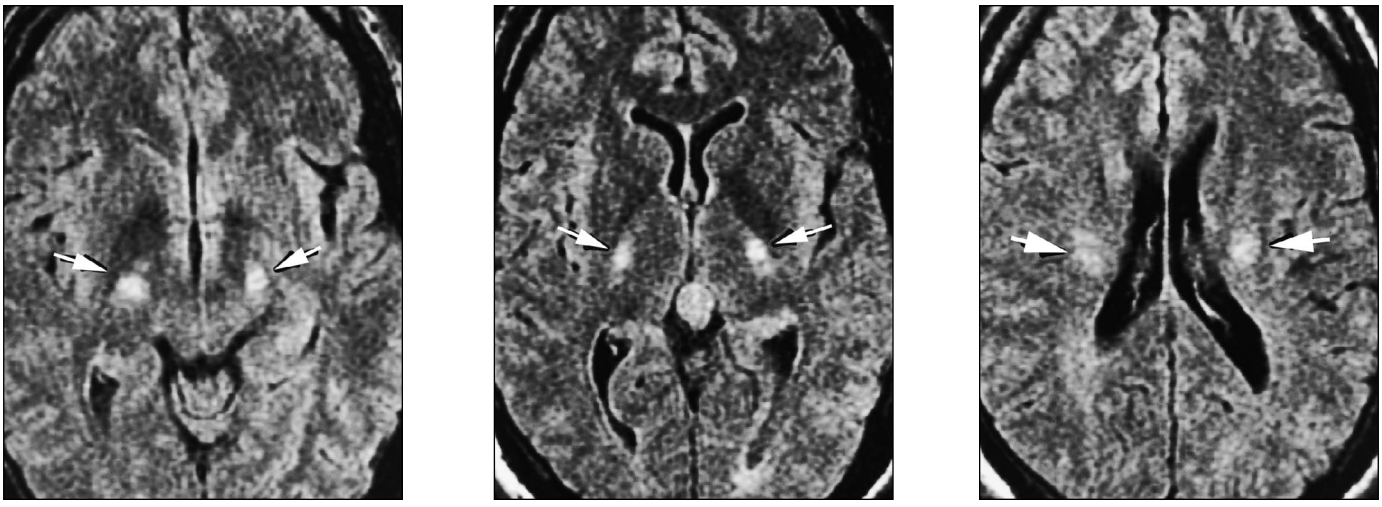

Figure 1 (A – C): *Axial FLAIR images from Patient 1, demonstrate focal symmetric increased signal (arrows) in the corticospinal tracts, in the cerebral peduncle (A), the posterior limb of the internal capsules (B), and the corona radiata (C). An incidental pineal cyst is noted in Figure 1B.*

been described in vitamin B12 deficiency, Friedreich's ataxia, and normal subjects, but signal changes are usually limited to the internal capsule. Asymmetric corticospinal tract changes may represent Wallerian-like degeneration, secondary to multiple potential etiologies. Bilateral symmetric increased FLAIR and T2 signal in the corticospinal tracts, extending from the corona radiata to the brainstem, however, when present, appears to be highly suggestive of ALS.

Submitted by W. Morrish

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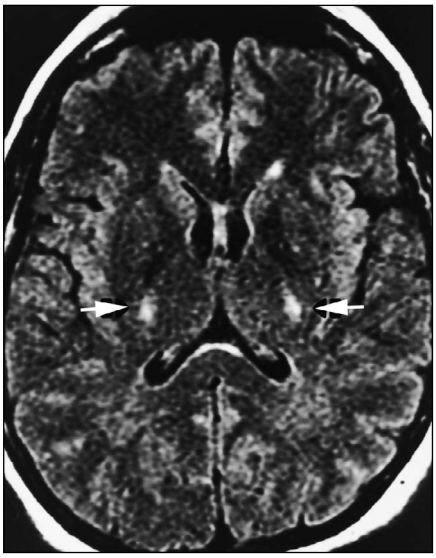

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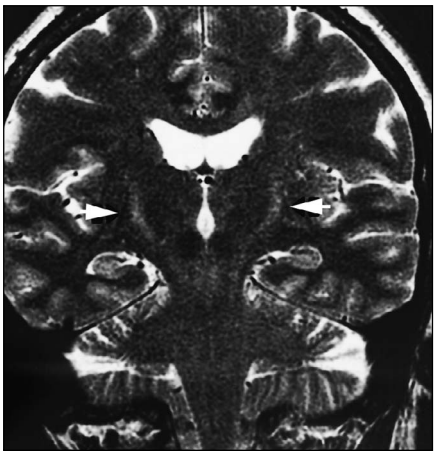

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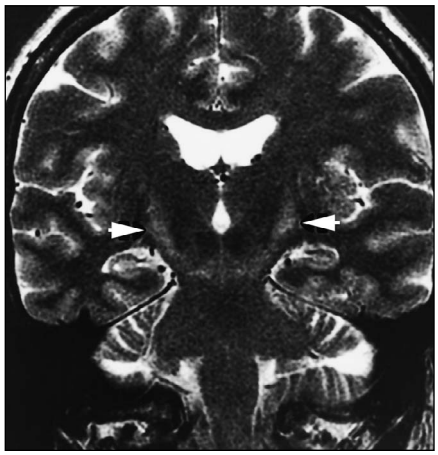

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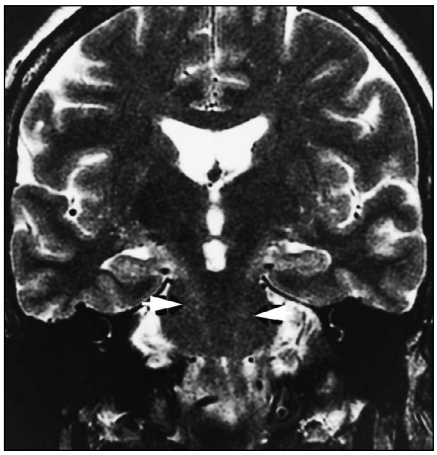

D

Figure 2 (A – D): *Axial FLAIR (A) and coronal T2 (B - D) images from Patient 2 demonstrate symmetric increased T2 signal in the corticospinal tracts (arrows). This is visualized in the posterior limbs of the internal capsules (A), and extending from the posterior limbs of the internal capsules into the pons (B, C, and D).*

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