Neuroradiology Case of the Day

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HISTORY

A 45-year-old man presented with progressive dysphonia of 2 years duration. After the onset of dysphonia, the patient had started developing dysarthria. Several months later, he began noticing progressive motor weakness, first in the hands and then in the legs. He also noticed fasciculations involving different muscles of the extremities and trunk. These were sometimes associated with muscle aches and cramps. The patient’s condition deteriorated to the point where he was unable to perform his job as a fisherman. His speech became unintelligible, and he was unable to chew solid food. Physical examination showed significant atrophy and fasciculations of the tongue and bilateral mild facial weakness. However, the pupillary reflexes were normal. Examination of the limbs showed pronounced atrophy of the muscles of the forearm and of the intrinsic muscles of the hands. Similar findings were seen in the feet and legs. Motor strength was diminished. Deep tendon reflexes were brisk with clonus and a positive Babinski sign on one side. There was no vitamin B12 deficiency, and protein electrophoresis did not show an abnormal band. Further work-up included electromyography and magnetic resonance (MR) imaging of the brain.

FINDINGS

Electromyography showed denervation potentials. Unenhanced brain MR imaging demonstrated abnormally high signal intensity on both proton-density–weighted and T2-weighted images. This high signal intensity extended from the subcortical white matter of the motor cortex and the corona radiata (Fig 1), through the most posterior aspect of the internal capsule (Fig 2), and into the cerebral peduncles (Fig 3).
The abnormality appeared symmetric and well-circumscribed. Coronal MR imaging demonstrated continuous linear involvement (Fig 4). Low signal intensity was seen along the precentral gyrus on T2-weighted images (Fig 5). Both the corresponding T1-weighted images and MR images of the cervical spine demonstrated normal findings.

**DIAGNOSIS:** Amyotrophic lateral sclerosis.

**DISCUSSION**

Amyotrophic lateral sclerosis, the most common form of motor neuron disease, is a devastating, progressive, neurodegenerative disorder that demonstrates both upper (hyperreflexia, spasticity) and lower (fasciculation, atrophy) neuronal symptoms. By definition, there should be no autonomic, sensory, or cognitive involvement. The El Escorial diagnostic criteria are typically used for clinical diagnosis (1). Amyotrophic lateral sclerosis demonstrates a male predilection with onset in the middle and late adult years (2).

At gross examination, the brain may show atrophy of the precentral gyrus. At microscopic analysis, there is loss of pyramidal and Betz cells in the motor cortex with loss of anterior horn cells in the spinal cord. The proximal axons of the neuronal cells show swelling. Ferric stains demonstrate strong staining for iron (2).

The exact cause of the disease is not well known. However, after the recent discovery of a mutation in the gene encoding the enzyme superoxide mutase-1 in some patients with the disease, the theory of free radical damage to
neurons would appear to be the most plausible. Other theories include an autoimmune process or heavy metal toxicity (2).

The primary role of imaging in amyotrophic lateral sclerosis is to exclude other causes such as cervical degenerative disk disease, Chiari malformation, or multiple sclerosis. Imaging is also helpful in atypical cases of the disease (3).

Fairly typical MR imaging findings have been described in patients with amyotrophic lateral sclerosis (4). High signal intensity is seen involving the corticospinal tract at both T2-weighted and proton-density-weighted imaging, with the latter being more sensitive (2). This abnormal signal intensity extends from the corona radiata, through the most caudal aspect of the posterior limb of the internal capsule, into the ventral aspect of the brain stem, and finally into the anterolateral column of the spinal cord (2,5). Involvement of the corpus callosum has also been reported (6). High signal intensity in the corticospinal tract has also been described in some healthy individuals, along with Friedreich ataxia and vitamin B12 deficiency (7). In such cases, however, the abnormal signal intensity is usually limited to the internal capsule and does not extend to the corona radiata or brain stem as in amyotrophic lateral sclerosis (2). This extended involvement distinguishes amyotrophic lateral sclerosis from the periventricular pattern of multiple sclerosis (1).

In addition, T2-weighted MR imaging typically demonstrates low signal intensity in the motor cortex in amyotrophic lateral sclerosis. This finding has been attributed to T2 shortening due to iron deposition (3). Use of MR spectroscopy and magnetization transfer techniques for early identification of amyotrophic lateral sclerosis has also been reported (8,9). However, further research is probably needed.

REFERENCES