QUESTION-ANSWER

What is your diagnosis?
Quel est votre diagnostic?

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Observation

This 55-year-old man was admitted for investigation into his 2-year history of progressive motor weakness of the upper and lower limbs. Neurological examination showed spastic paraparesis with bilateral Babinski sign, muscular atrophy of the upper extremities and severe atrophy of the tongue. Routine blood tests were normal, syphilis serology was negative and there was no vitamin B12 deficiency.

Magnetic resonance imaging (MRI) of the brain was performed (Fig. 1).

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Commentary

MRI of the brain showed bilateral symmetrical high signal intensity in the lateral parts of pons and cerebral peduncles, posterior limbs of the internal capsules, lower part of the corona radiata and white-matter fibers of the motor cortex (Fig. 1). These findings are suggestive of corticospinal tract degeneration as seen in amyotrophic lateral sclerosis (ALS).

We performed an electromyography, which suggested generalized lower motor neuron involvement of the head, arms and legs, supportive of a diagnosis of ALS.

The clinical findings, associated with the results of MRI and electromyography, confirmed the diagnosis.

Discussion

ALS is a degenerative disease of unknown cause that affects upper and lower motor neurons. It is the most common form of motor neuron disease. The prevalence of sporadic ALS is about five cases in 100,000 and primarily affects middle-aged to elderly adults with a male predominance [1,2].

Histopathological 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 [3]. The pathogenesis of ALS is still unknown [2], and its clinical presentation is highly variable, including signs of upper and lower motor neuron compromise [4].

Several types of MRI abnormalities have been found in patients with ALS, and novel MRI features have been described in the last few years. The most important and consistent finding is degeneration of the corticospinal tracts presenting as hyperintense signals on T2-weighted images and Fluid Attenuated Inversion Recovery (FLAIR). Large, round, hyperintense signals that are visible bilaterally in several slices along the pyramidal tracts, as were seen in our case, are typical of ALS [4]. Such hyperintensity suggests axon lysis and myelin degradation into proteins and lipids with local release of water contents. However, this MRI finding should not be read in isolation, and good clinical correlation is needed to consider it suggestive of ALS. On the other hand, T2 hypointensity has also been described in the pre-central motor cortex of ALS patients. Some authors have also described atrophy of the superior parietal gyrus, and yet another abnormality is high signal intensity in fibers of the corpus callosum on T2-weighted spin-echo images [4,5]. In addition, hyperintense signals in the anterolateral columns of the cervical spinal cord on T1-weighted images have been described [4]. One study has also reported that T1-weighted spin-echo magnetization transfer contrast sequence is sensitive and accurate in depicting corticospinal tract lesions in ALS as hyperintense areas along the corticospinal tract [6]. However, MR spectroscopy is more sensitive than MRI in the detection of motor neuron abnormality in ALS [5].

No cure has yet been found for ALS, a disease that invariably leads to death, usually within 3–5 years of its onset [6]. This means that treatment can only relieve symptoms and improve the quality of life for ALS sufferers.

In conclusion, MRI can detect degeneration of corticospinal tracts in ALS. Correlation with clinical and electromyographic findings is necessary to arrive at the diagnosis.

References