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MAGNETIC RESONANCE FINDINGS IN AMYOTROPHIC LATERAL SCLEROSIS USING A SPIN ECHO MAGNETIZATION TRANSFER SEQUENCE

PRELIMINARY REPORT

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ABSTRACT - We present the magnetic resonance (MR) findings of five patients with amyotrophic lateral sclerosis (ALS) using a spin-echo sequence with an additional magnetization transfer (MT) pulse on T1-weighted images (T1 SE/MT). These findings were absent in the control group and consisted of hyperintensity of the corticospinal tract. Moreover we discuss the principles and the use of this fast but simple MR technique in the diagnosis of ALS.

KEY WORDS: amyotrophic lateral sclerosis, pyramidal / corticospinal tract, magnetic resonance, magnetization transfer.

Alterações encontradas por ressonância magnética na esclerose lateral amiotrófica utilizando sequência *spin-echo* com transferência de magnetização: relato preliminar

RESUMO - Apresentamos as alterações do sinal de ressonância magnética (RM) encontradas em cinco pacientes com esclerose lateral amiotrofica (ELA) utilizando sequência *spin-echo* ponderada em T1 com pulso adicional de transferência de magnetização (T1 SE/MT). Essas anormalidades estão ausentes no grupo controle e consistem de hipersinal dos tratos piramidais. Discutimos os princípios e o emprego dessa técnica simples e rápida de imagens por RM no diagnóstico de ELA.

PALAVRAS-CHAVE: esclerose lateral amiotrófica, trato piramidal / cortico-espinhal, ressonânica magnética, transferência de magnetização.

Amyotrophic lateral sclerosis is a rare degenerative disease of unknown etiology that affects the motor neurons of the corticospinal tract and the spinal cord. Although the clinical and pathological abnormalities of ALS have been well described, its radiological findings are scarce and nonspecific.

The gross pathological changes include spinal cord atrophy secondary to neuron loss, which includes the motor nuclei of the inferior brainstem and the precentral cortex. Magnetic resonance (MR) imaging may depict these changes, mainly those of the brainstem and spinal cord. However,

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the involvement of the corticospinal tract cannot be properly demonstrated, although dubious T2-weighted images changes have been described. Other degenerative diseases of the motor neurons, such as progressive bulbar palsy and progressive muscular atrophy have a worse prognosis and do not show degeneration of the corticospinal tract.

Several papers have discussed the use of magnetic transfer (MT) on MRI. This technique is based mainly on the progressive loss of signal intensity of free water protons and on the consequent signal hyperintensity from tissues with protons bound to macromolecules given background suppression.

The purpose of this paper is to present the preliminary results of MRI as a diagnostic technique of ALS.

METHOD

Spin-echo MT sequence was performed on five patients(females; mean age 53,6) with progressive muscular weakness, brisk reflexes, fasciculation of tongue and/or limbs and bulbar symptoms suggestive of involvement of the cranial nerves.

MRI was performed on a 1.0 T Philips NT-10. Images were obtained in the axial plane, with the following parameters: 510 / 12 / 2 (TR / TE / NSA), FOV of 240x240 mm and MT pulse on resonance.

RESULTS

In the present study unambiguous findings in the corticospinal tract characterized by triangular areas of spontaneous signal hyperintensity on T1 SE/MT with extension from the semioval center to the motor cortex in the prefrontal gyrus, were demonstrated in all the patients (Fig 1). 1500 other brain MR exams were conducted in our division using the technique T1 SE / MT and no similar alterations have been reported in any other disease or even in normal individuals. The signal abnormalities observed indicate the selective lesion of the ALS over the pyramidal tracts. Thus the

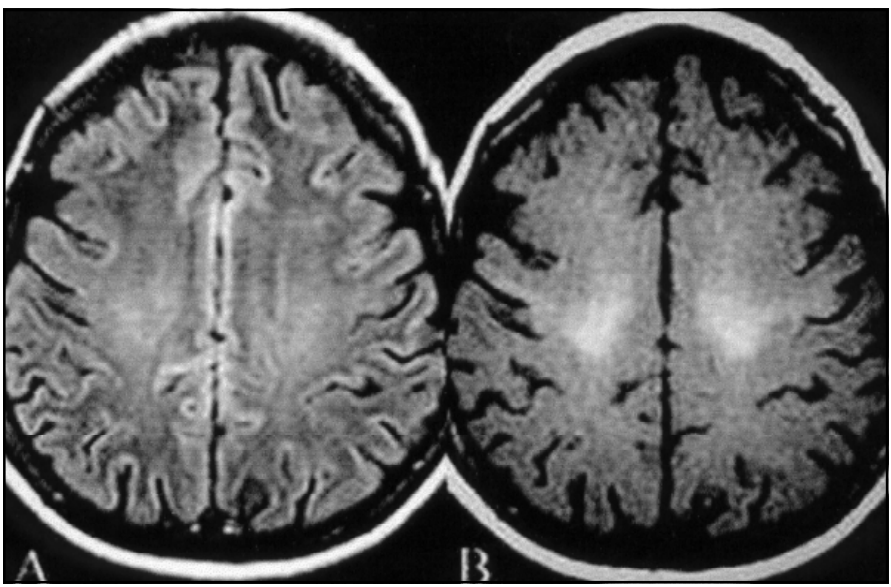

Fig 1. ALS patient. A- Axial FLAIR showing hyperintensity of the pyramidal tracts on the semioval center. B- Axial T1/SE/MT: marked hyperintensity of the pyramidal tracts, more evident than on FLAIR images.

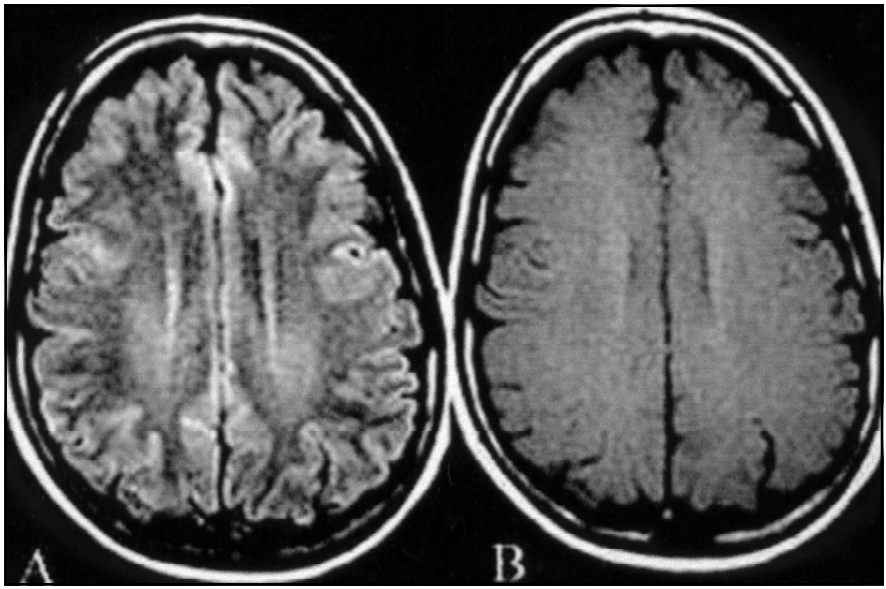

Fig 2. Healthy subject. A- Axial FLAIR showing hyperintensity of the pyramidal tracts extending from the internal capsule to the subcortical region of the precentral giri. B- axial T1/SE/MT: no abnormal findings are seen.

use of an additional magnetization transfer pulse allows, for the first time, to our knowledge, an unambiguous proof of this disease.

DISCUSSION

Areas of abnormal signal intensity on T1 and T2-weighted images in the corticospinal tract have been previously reported in patients with ALS[1-5]. However, these abnormalities were inconsistent and unreliable since they were observed frequently in normal patients[6], which poses a limitation to their usefulness (Fig 2). Other diseases such as ischemic processes, demyelinating diseases, infections, may involve the pyramidal tracts. However, demonstrating the bilateral symmetrical involvement associated with the characteristic clinical findings allows the diagnosis of ALS.

The additional use of MT aims at identifying of the affected brain tissue and can be used to alter or improve the image contrast between healthy and pathological tissues. Its use in the diagnosis of several other neurological diseases, including multiple sclerosis[7], tumors[8], among others, has been demonstrated both with and without the use of paramagnetic agents. The use of this technique to identify early lesions in the motor neuron disease has also been demonstrated[9,10]. However, the reports are based on the MT measurements, which require sophisticated equipment and takes much longer, since it calls for a post-processing procedure in order to obtain the variation rate among tissues.

Following previous reports, we decided to assess its usefulness through a simple method, observing only the alterations obtained from the usual sequences, which do not require post-processing procedure, and using only conventional MR equipment. The observed alterations are a result of the difference between the MT rates from the tissues from which the MR signal originate from free water protons or macromolecules. Tissues with free water have a greater MT rate, which results in a lower MR signal, whereas those of macromolecules, show a higher resulting signal.

The diagnosis of ALS has been based only on clinical and electromyographic data. The degeneration of motor neurons may result in a cellular loss and axonal edema as previously shown

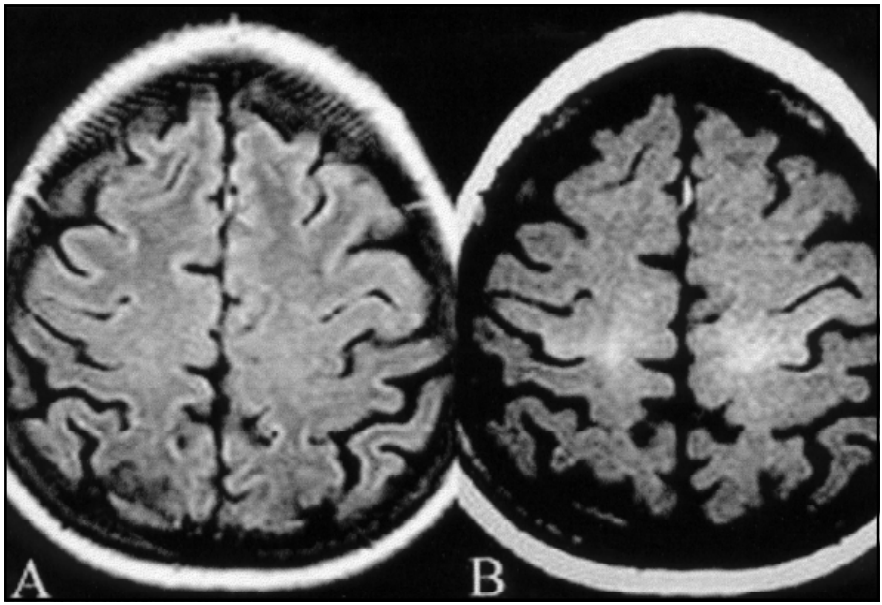

Fig 3. ALS patient. A- Axial FLAIR without abnormal findings. B- Axial T1/SE/MT showing the alterations more conspicuously than on FLAIR.

on electronic microscopy[11]. So far, imaging methods have been of limited use because of their unsatisfactory results.

In conclusion, in our small group of patients, T1 SE / MT MR imaging has been shown to be a valuable method of diagnosis of this rare disease. The findings of a bilateral symmetrical hyperintensity of the pyramidal tract are suggestive of ALS denoting degeneration of corticospinal tracts (Fig 3), although the nature of these findings require further investigation. This study is a preliminary report of a much larger investigation of this motor neuron disease and it serves to show for the first time,to our knowledge, the description of this abnormality.

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