Amyotrophic lateral sclerosis and neurocysticercosis

Martínez Héctor R,*,*** Caro Enrique,*Gil-Valadez Alfonso,*,***
Moreno Cuevas Jorge,*** González-Garza María Teresa,*** Molina-López Juan Francisco,***
Treviño-Manllo Sergio A,*** Hernández-Torre Martín****

* Servicio de Neurología, Hospital San José Tecnológico de Monterrey.
** Servicio de Neurocirugía, Hospital San José Tecnológico de Monterrey.
*** Tecnológico de Monterrey, School of Medicine, Servicio de Terapia Celular, CITES, Monterrey N.L., Mexico.
**** Biotechnology and Health, Tecnológico de Monterrey.

Revista Mexicana de Neurociencias — Mayo-Junio, 2010; 11(3): 240-242

INTRODUCTION

Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disorder characterized by rapid deterioration and selective death of motor neurons in the cerebral cortex, brain stem and spinal cord.[1,2] Despite advances in understanding the molecular basis of ALS, the etiology of sporadic cases remains unexplained.[3,4] Neurocysticercosis (NCC) is produced by CNS infestation with cysticerci, the larvae of *Taenia solium*. The spectrum of neurologic syndromes in NCC is broad and depends on the number, size and location of the cysts in the CNS and host immune response. The diagnosis of NCC is supported by neuroimaging and CSF immunodiagnostic assays.[5] ALS incidence is approximately 2 per 100,000 persons per year.[6] In Mexico, the prevalence of NCC may be as high as 3% based on autopsies performed at third level hospitals.[5] The brain is frequently involved in NCC, whereas the spinal cord is rarely involved. The association between ALS and NCC has rarely been described in the literature.[7] We describe a patient with both disorders.

CASE REPORT

A 55-year old male without family history of neurological disease began 17 months before evaluation with bradylalia and transient periods of speech arrest. A cranial CT and brain MRI revealed a hypointense area in the left temporal lobe and right motor strip, surrounded by a hyperintense rim compatible with brain parenchymal cysts. CSF measurement of antibodies against cysticercus antigens was positive as determined by ELISA and the NCC diagnosis was established. After Praziquantel (50 mg/kg/day for 15 days) treatment, right hand weakness ascended to the arm and shoulder. Cervical MRI was normal and no treatment was instituted. Five months before first evaluation, fasciculations in upper and lower limbs, left upper limb weakness, sialorrhea, dysphagia, and loss of strength in cervical musculature were observed. After a second period of Praziquantel (50 mg/kg/day for 15 days) and steroids, patient experienced weakness in the right lower leg and sialorrhea during sleep with consequent dyspnea. Neurology Service (Clinical)

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RESUMEN

La asociación entre la Esclerosis Lateral Amiotrófica (ELA) y la Neurocisticercosis (NCC) rara vez se ha descrito en la literatura. Presentamos un paciente con ambos trastornos. Se diagnosticó NCC en un paciente de 55 años de edad, masculino con tomografía axial y pruebas de inmunoensayo en líquido cefalorraquídeo (LCR). Después del tratamiento con Praziquantel, desarrolló bradilalia, disartria llegando a lenguaje incomprensible. También mostró sintomatología bulbar y de neurona motora superior e inferior. La electromiografía apoyó el diagnóstico ELA definida, la resonancia magnética de cerebro confir-

ABSTRACT

The association between Amyotrophic Lateral Sclerosis (ALS) and Neurocysticercosis (NCC) has rarely been described in literature. We describe a patient with both disorders. NCC was diagnosed in a 55 year-old male patient with positive CT scan and cerebrospinal fluid (CSF) immunoassay tests. After Praziquantel treatment, he developed slurred speech, bradylalia and periods of speech arrest. He also demonstrated bulbar, upper and lower motor neuron symptomatology. Electromyography supported definite ALS diagnosis, and brain MRI confirmed the presence of NCC in motor cortices. Although the association between NCC and ALS may have occurred by chance, we hypothesize that autoimmune, apoptotic and circulatory

mó la presencia de NCC en corteza motora bilateral. Aunque la asociación entre el NCC y ELA puede haber ocurrido por casualidad, proponemos una hipótesis en la que mecanismos autoinmunes, de apoptosis y circulatorios contribuyeron en el desarrollo de ELA en este paciente con NCC.

Palabras clave: Esclerosis lateral amiotrófica, neurocisticercosis, praziquantel.

mechanisms contributed in the development of ALS in this patient.

Key words: Amyotrophic lateral sclerosis, neurocysticercosis, praziquantel.

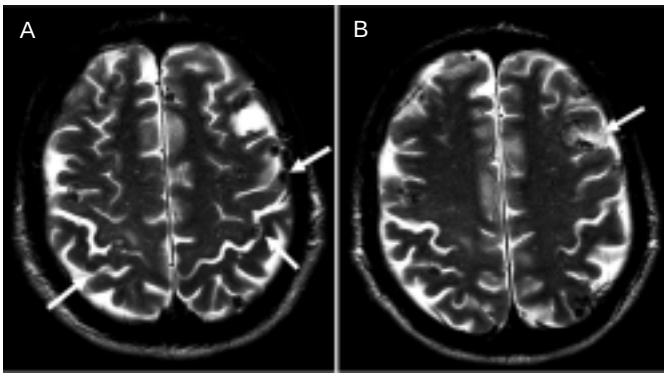

Figure 1. Admission brain MRI shows on T2 weighted images. A. Multiple calcifications in frontal motor strip (arrows). B. Cysticercus in granular stage in left frontal lobe (arrow).

evaluation showed non-comprehensive speech, sialorrhea, neck weakness, tongue with atrophy and fasciculations. Upper (1/5) and lower (3.5/5) limb weakness with muscular atrophy and fasciculations also presented with generalized hyperreflexia, bilateral Babinski, Chaddock, Hoffman and Trommer signs. Electromyography demonstrated positive sharp waves, fibrillations and fasciculations in four limbs. A nerve conduction study revealed velocities within normal range. Diagnosis of definite ALS in association with NCC was then established. On admission, MRI showed multiple hypointense lesions in frontal lobes including bilateral motor strip (Figure 1).

DISCUSSION

The association of ALS and NCC has rarely been described in literature.[7] We describe an NCC patient who presented ALS after receiving cysticidal agent Praziquantel. The cause of ALS remains unknown although the identification of mutations in the SOD1 gene[6] is relevant. Other etiologic hypotheses have been proposed and include exposure to heavy metals, virus, prions, endogenous cytotoxic factors, age, apoptosis, abnormal neurotrophic factors or axonal transport and autoimmunity.[1-3,6] None of these mechanisms alone explain the cascade of events that lead to selective motor neuron destruction.

In our patient, NCC and ALS association may have occurred by chance. We consider that inflammatory reaction against the cysticercus promoted the release of substances such as peripherin which is known to cause axonal injury, disorganization of neurofilaments and axonal strangulation.[3] Inflammatory molecule release may have up-regulated transcriptional factors leading to activation of apoptotic paths, thus creating and amplifying cascade of caspases which led to degradation of DNA and cell death.[8] An autoimmune cross reaction between NCC antigens and dystonin may have produced upper motor neuron destruction by losing neuronal cytoskeleton integrity.[3] Adhesive leptomeningitis and meningeal fibrosis in the brain and spinal cord is frequently described in NCC.[5] Perivascular involvement in spinal cord by fibrosis or meningeal inflammation can produce vascular insufficiency with consequent lower motor neuron death.

Since NCC produces a broad spectrum of neurological manifestations, we suggest that ALS patients in developing countries should undergo CSF immunoassay evaluation to corroborate its association with NCC. Autoimmunity, apoptosis and ischemia induced by NCC and/or Praziquantel can play a significant role in the pathogenesis of this ALS patient.

CONFLICT OF INTERESTS

The authors declare that they have no conflict of interest.

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Corresponding: Héctor R. Martínez MD, FACP
School of Medicine. CITES 3rd floor
Morones Prieto No. 3000
Col. Pte Monterrey
C.P. 64710, Nuevo León, México
Tel.: 52(81) 8888-2177,
Fax: 52(81) 8888-2148
E-mail: hector.ramon@itesm.mx