Amyotrophic lateral sclerosis and neurocysticercosis

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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 neurologic disease began 17 months before evaluation with bradykiegia 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, salorrhea, 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 salorrhea during sleep with consequent dyspnea. Neurology Service (Clinical)
evaluation showed non-comprehensive speech, salorrhea, 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. 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 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. 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. 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. An autoimmune cross reaction between NCC antigens and dystonin may have produced upper motor neuron destruction by losing neuronal cytoskeleton integrity. Adhesive leptomeningitis and meningeal fibrosis in the brain and spinal cord is frequently described in NCC. 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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