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Kinesigenic Dystonia as the First Manifestation of Multiple Sclerosis with Cervical and Brainstem Lesions

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Paroxysmal symptoms, particularly paroxysmal dystonia (also referred to as ‘tonic seizure’), had been clearly related to multiple sclerosis (MS) by Matthews [1] in 1958, although such symptoms had been reported since 1928 [2]. Sometimes they may be the first manifestation of the demyelinating disease [3]. When they occur without other signs or symptoms they often present diagnostic difficulties.

We describe the case of a young man with laboratory-supported definite MS [4] in whom unilateral tonic spasms were the initial symptom.

A 27-year old man was admitted with a history of variable numbness affecting the right side of his body, beginning 2 months earlier; 1 month later he developed carpo-pe-da- spasms in his right hand and sometimes a dystonic involvement of his lower right limb, facial grimacing, torsion of the neck and dysarthria. The episodes lasted about 60s and were not accompanied by an alteration in consciousness, loss of sphincter control or postictal sequels.

During the dystonic posturing a Babinski sign was present on the right side. Such episodes occurred 4-5 times daily only during the waking-up period. They were always evoked by voluntary movements of the neck or of the right arm and by hyperventilation (while the patient was at rest, but not if he blew into a bag).

On admission physical examination disclosed brisk muscle stretch reflexes on the right, slight impairment of vibratory sensation in the right upper limb. Plantar re-

Fig. 1. Axial slice from the MR scan demonstrating a lesion in the left cerebral peduncle.

sponses were normal on both sides. Repeated EEGs during the attacks and interic-tally were normal. The visual evoked potentials and brainstem auditory evoked potentials were within normal ranges. Somatosensory evoked potentials of the upper limbs showed bilateral lowering of the cortical component. The cortical magnetic stimula-

Fig. 2. Spinal slice from MRI scan showing an area of altered signal located in the right cervical tract C4-C5.

tion showed an increased motor latency for the right limbs, whereas the cervical stimulation (C7) was within the normal range. The brain CT scan did not show any focal lesion. The MRI study, on T2-weighted images, displayed an area of increased signal involving the left subthalamic nuclei and the left cerebral peduncle (fig. 1) and the right

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cervical tract at the level C4-C5 (fig. 2). Cerebrospinal fluid showed an increased IgG level (9.1 mg/dl) with oligoclonal bands (IgG kappa-lambda) and free K chains.
Paroxysmal dystonia, also known as tonic seizures or tonic spasms, is characterized by the sudden onset of abnormal, involuntary posture of one or more limbs, sometimes with associated facial grimacing. Usually unilateral, abnormal dystonic postures can spread contralaterally or have a bilateral onset [4, 5]. They can be associated with painful sensations and autonomic symptoms [4, 5]. Active movements, sensory stimuli or hyperventilation can generate bouts of paroxysmal dystonia [6, 7]. Paroxysmal dystonia can occur many times a day (up to 30) and last from seconds to minutes. Antiepileptic drugs can readily suppress the attacks [6]. No EGG alteration is reported during paroxysmal activity [5-8].
In 1 case, autopsy revealed a demyelinat-ing spinal cord lesion, while in other cases computed tomography or magnetic resonance scans showed lesions in the contralateral internal capsule [5, 8, 9]. To our knowl-edge our case is the first in which there is a brainstem lesion in association with a cervical one.
Because of the presence of facial involvement we believe that most of the clinical symptoms are caused by the supraspinal lesion. The mechanisms causing the symptoms are not known; hyperventilation can induce alterations in blood flow and pH [10]. According to Berger et al. [6] paroxysmal dystonia related to MS must be correctly diagnosed and differentiated from ‘tonic seizures’ present in other diseases.

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