Letters

Improvement of idiopathic torsion dystonia following dystonia-induced cervical subluxation

Sir: Idiopathic torsion dystonia is an involuntary movement disorder consisting of twisting, pulling, and sustained contractions that may be extremely powerful and painful. The adult-onset form usually remains restricted to one section of the body, sparing the legs. The treatment of idiopathic torsion dystonia has included thalamic surgery and medical therapy with various muscle relaxant and neurotransmitter drugs. We present a patient with adult-onset dystonia with a unique clinical course.

A 50-year-old non-Jewish white male presented in June 1977 with the onset of dystonic movements of the neck and upper extremities. His perinatal history and early development were normal, and there was no history of any known precipitating illness or exposure to medications known to provoke torsion dystonia. Family history was negative for dystonia or other neurological disorders.

He first presented to our hospital in April 1980. Examination revealed dystonia of the trunk, neck, and proximal upper extremities. No intellectual, pyramidal, cerebellar, or sensory deficits were observed. Reflexes were normal and the Babinski response was bilaterally absent. Evaluation including an electroencephalogram, CT scan of the head, electromyography, nerve conduction velocity studies, antinuclear antibody levels, coeruleoplasmin levels, intravenous pyelography, and thyroid function studies were all within normal limits. Treatment with benztpnone, physostigmine, haloperidol, carbiridopa-levodopa, diazepam, amantadine, and artane failed to improve his dystonic movements.

In May 1981 he noted increased involuntary movements of his neck, trunk, and upper extremities. Examination at that time was again normal except for the presence of dystonia. During that admission he experienced the sudden onset of pins and needles sensation in both arms and legs, and complete inability to move all extremities. There was no neck trauma other than from his severe dystonic movements. On physical examination he was hyperreflexic in the lower extremities and the Babinski responses were present bilaterally. Motor strength was graded at three out of five and his sensory deficit was at the T6–T7 level. Spinal CT scan and myelogram revealed cervical spondylitis and a C2–C3 subluxation. He was treated initially with dexamethasone and cervical restraints. Subsequently he underwent a C3 laminectomy with fusion of vertebrae C2 to C5, decompression of nerve roots C2, C3, and C4 bilaterally, and crushing of the C3 nerve roots bilaterally. After the operation he remained quadriplegic at the C5 level. There were no dystonic movements.

He was discharged to a nursing home and returned one year later for follow-up. The patient stated that his motor strength had steadily improved without return of his dystonic movements. Medical treatment at that time included benztpnone and baclofen. His motor strength was four out of five in the extensors of the upper extremities, but otherwise five out of five. In the lower extremities strength was four out of five, except for planter flexion which was five out of five. Reflexes were increased in the upper extremities and normal in the lower extremities. There was dysmetria in finger to nose testing bilaterally. The Babinski and Hoffman signs were present bilaterally. Mental status and sensory functions were not impaired. Only minimal dystonic movements of the neck were observed consisting of some twisting of his head to the right. The patient's drug regimens were discontinued without increase of his involuntary movements.

Our patient meets the criteria of idiopathic torsion dystonia as described by Marsden et al. To our knowledge this is the first report in which a patient's dystonic movements produced a cervical subluxation and eventual quadriplegia. In addition, one year later the patient's motor strength improved with only a minimal return of his involuntary movements. There are several possible explanations for this occurrence. First, the patient may have undergone spontaneous remission of his disease. Many patients with idiopathic torsion dystonia have gone through periods of partial remission, but this explanation seems unlikely because the patient has remained improved for a period of one year without exacerbation of his symptoms. Second, he may have been adequately controlled on medical therapy, namely benztpnone and baclofen. However, discontinuation of these medications did not result in an increase of his involuntary movements. Third, the involvement of his cervical spinal cord by the subluxation and/or surgery may have affected certain neural structures necessary to produce the involuntary movements of idiopathic torsion dystonia. This explana-

References


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Focal paroxysmal kinesigenic choreoathetosis preceding the development of Steele-Richardson-Olszewski syndrome

Sir: Precipitate falls in progressive supranuclear palsy (the Steele-Richardson-Olszewski syndrome) have been previously attributed to visual difficulties such as with
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