Improvement of idiopathic 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, coeruloplasmin levels, intravenous pyelography, and thyroid function studies were all within normal limits. Treatment with benzotropine, physostigmine, haloperidol, carbipoda-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 spondylit 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 benzotropine 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 plantar 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 benzotropine 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 explanation seems to be the most plausible in this patient especially in view of reports that cervical spinal cord stimulation with epidural electrodes may improve the involuntary movements in some dystonic patients.

Further studies in this area are warranted.

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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
downward gaze and/or cycloplegia, mild ataxia, truncal ataxia, sudden loss of tone on one side, falling backward due to severe neck dystonia in extension.\(^1\) However, Behrman\(^2\) observed that these falls are often among the first symptoms, and may occur when neurological examination reveals no abnormality. Therefore in most cases the cause of this paroxysmal dysyequilibrium remains unexplained. Here, a case is described at which the abrupt falls were clearly due to left sided paroxysmal dystonia that was induced by movement.

A 56-year-old African female from Meru district in Kenya first presented to the teaching hospital in Nairobi on 9 June 1983 with a history of falls. The resident on duty diagnosed heart block and referred her to a cardiac clinic. At this clinic an important detail of her presenting symptom was elicited, namely, the falls were always preceded by criss-crossing of her left leg across the right leg and therefore the patient would trip herself and fall down without losing consciousness. This symptom had been present for three years. She was then referred to a neurology clinic where she was noted to be mentally slow and her left deep tendon jerks were brisker than right but both plantar reflexes were flexor. A diagnosis of drop attacks was made; EEG showed a normal reactive alpha rhythm 8–9 Hz over the bifrontal areas. There was no sharp or slow wave activity.

Her condition became worse so that five months later she had difficulty in walking, her speech became slow and there was blurring of her vision. She was admitted for investigations. An ophthalmological examination, including a slit lamp examination, showed normal optic media with no Kayser/Flesher ring and normal fundi. The visual acuity was also normal; 6/6 bilaterally. Serum for universal syphilis reagent was positive with a titre of 1:8 and the specific treponema haemaglutination test was reactive. The CSF was completely normal. Her neurological disease was wrongly diagnosed as general paresis of the insane and the patient was given a total of 21 million units of intramuscular procaine penicillin over a period of five weeks. She was discharged from the hospital six weeks later.

After this prolonged and vigorous antisyphilitic treatment her symptoms were never arrested and they showed the same relentless progression so that by November, 1984, the patient could neither walk nor stand. Her dysarthria and visual difficulties were also becoming worse. The patient was again referred to the neurology clinic and was seen by the author on 5 November, 1984. During this visit the following points were noted: the paroxysmal dystonia was precipitated by movement, so that it would occur on standing or walking after sitting or lying down. The dystonia was brief lasting for about 10–15 seconds without loss of consciousness. There was no aura and it was not accompanied by facial grimacing nor incontinence during the attack. The patient was unable to abort an attack voluntarily. There were no post-ictal phenomena. The posture of the leg was plantar-flexion and inversion of the foot, knee flexion, hip flexion and adduction. The upper limb was adducted at the shoulder, flexed at the elbow, wrist metacarpophalangeal and interphalangeal joints, during the dystonia. These attacks were initially paroxysmal. In between attacks there was no evidence of the dystonic posture. The frequency of these attacks was about four to six episodes a day. She was not aware of any other factors such as, starlette, coffee, hypertension, that could induce an attack and was not aware of a refractory period. There were no attacks in the right sided limbs. However, the illness had progressed so that for six months prior to this consultation the dystonia was more or less permanent so that there was hardly any moment she could remember that her left limbs relaxed from the dystonic posture.

She also complained of dysphagia. There was no reversal in sleep rhythm and in particular no history of oculargic crisis. She denied also history of tremor, myoclonic jerks, convulsions and headache. There was no history to suggest involvement of the sphincters, and there was no past history to suggest encephalitis. She was not on any drugs at that time. Both her parents died when she was very young for causes unknown to her. One of her brothers was killed during the war of independence. The remaining brother and sisters are healthy, one of her daughters died of carcinoma of cervix and another has goitre. The other four daughters and four sons are healthy.

Examination revealed a short, thin middle age woman with a spastic face and an intense staring appearance. She rarely blinked and when she did so the movement was slow. There was no sialorrhoea. She had many traumatic scars on her left sided limbs because of the repeated falls. She was well oriented in person, place and time. She had a normal attention span. Short term and long term memory was intact. She could not do simple addition, and she could not interpret simple proverbs from her own language. She had no hallucinations and no delusions, grandiose or otherwise. She had severe slurring dysarthria and bradykinesia. Vertical gaze both upward and downward for willed and pursuit eye movements was absent, although these eye movements could be induced by oculocephalic reflex. Bell’s phenomenon was absent. The only horizontal eye movements present were the pursuit eye movements and even these were jerky. There was no convergence. The pupils were small and regular. They reacted to light but not to accommodation. Cold caloricity in either ear induced only tonic deviation of both eyes to the same side. There was no nystagmus. Simultaneous irrigation of both ears with cold water caused only tonic deviation of both eyes downwards. Vertical and horizontal optokinetic nystagmus was also absent. The rest of the cranial nerve examination was normal including both fundi. The jaw jerk was absent and the gag reflexes were normal. The primitive reflexes were present such as the glabellar tap, sucking, pouting, palmo-mental and bilateral tonic grasp reflexes.

The neck was held rigidly erect. All movements of the neck were possible except flexion, which was difficult because of the hypertonicity of the next extensors. The left limbs were held in a dystonic posture described above during a paroxysmal attack, and they showed plantar rigidity. There was no tremor. The motor system revealed normal bulk of muscles and normal power. The deep tendon jerks were symmetrically exaggerated the abdominals were absent and the plantar responses were consistently flexor. She could neither stand nor walk because of the dystonia. There were no cerebellar signs. Sensation was intact to all modalities. There was no parietal sensory loss. The other systems revealed no abnormalities and there was no postural hypotension.

A repeat of blood serology for syphilis after the prolonged course of penicillin showed that the treponema haemaglutination test had become non reactive and the universal syphilis reagent titre had fallen to 1:2. A repeat CSF study again revealed a completely normal fluid. All the other haematological, biochemical and simple radiological investigations were normal. The leucocyte glutamate dehydrogenase activity was normal at 0.768 μmol/h/mg protein with controls of 0.719 and 0.695 μmol/h/mg protein. This enzyme activity was assayed according to the method of Duvois et al.\(^4\)

Her left sided rigidity resolved on levodopa and the paroxysmal attacks of her left sided limbs were abolished by carbamazepine.
This is the first case of progressive supranuclear palsy in which falls, which is a characteristic early symptom of the disease, are clearly due to focal paroxysmal kinesigenic choreoathetosis and they have many of the criteria proposed by Kertesz. These criteria are: duration and character of the paroxysms, response to anticonvulsants even though the dystonic illness was at an advanced and more complex in this case, the precipitating factor and absence of EEG abnormalities. The late age of onset of this focal paroxysmal kinesigenic choreoathetosis and the absence of a positive family history is explained by the “symptomatic” nature of this disorder. However, later in the evolution of the tonic disorder in this case of progressive supranuclear palsy, the dystonia became more or less permanent. This may explain the inability of levodopa alone to abolish this more complicated dystonia as in the case described by Loong although it effectively resolved the persistent plastic rigidity component. At this point the paroxysmal component of this disorder became apparent again as in earlier stages and addition of carbamazepine effectively abolished the focal paroxysmal kinesigenic choreoathetosis.

This is also a first report of progressive supranuclear palsy in Eastern Africa, which partly explains the delay in the diagnosis of this patient.

The occurrence of paroxysmal kinesigenic choreoathetosis in progressive supranuclear palsy is not surprising as most recent authors of the former consider it to be a disorder of basal ganglia function despite the only necropsied case showed no definite pathological findings, while in the later, pathological lesions in the basal ganglia are prolific. Finally, it is recommended that in patients with Steele-Richardson-Olszewski syndrome who fall, or in any obscure falls, focal paroxysmal kinesigenic choreoathetosis should be seriously considered and if this disorder is suspected a trial of carbamazepine should be instituted.

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References

Status epileptics due to abrupt diazepam withdrawal: a case report

Sir: Benzodiazepines are extensively used as anxiolytic and hypnotic drugs and are usually considered as having a low risk for pharmacological dependence. However, many authors have warned of the existence of a benzodiazepine withdrawal syndrome. We report a patient, heavily dependent on diazepam, who had a confusional psychosis and generalised tonic-clonic status epilepticus a few days after the abrupt discontinuation of the drug.

The patient, a 25-year-old right-handed man, was admitted to a psychiatric unit because of delusions and visual hallucinations experienced during the two preceding days. He also appeared anxious and confused, had tremors and excessive sweating and reportedly had experienced a seizure a few hours before, although further information was not available. Within one hour a generalised tonic-clonic attack was observed. Diazepam (10 mg IV) was administered and during the subsequent hours the patient was fully alert and felt quite well. During the third hour he again became restless and a second grand mal attack was observed. He was transferred to a medical unit. However, epileptic seizures occurred and became more frequent in spite of treatment with diazepam (50 mg IV within 12 hours) and phenobarbitone (200 mg im). The state of consciousness progressively deteriorated up to a comatose state. Routine haematocrit data revealed only a slight anaemia.

Twelve hours after admission the patient was transferred to our intensive care unit because of a generalised tonic-clonic status epilepticus. The duration of the seizures, which appeared every 15-20 minutes, was short. These were characterised by a brief tonic phase and mild clonic twitches of the