Paroxysmal choreoathetosis after head injury

Sir: Paroxysmal choreoathetosis is an involuntary movement disorder which is often precipitated by an attempt to initiate motor activity, and which may resemble an elementary partial seizure.1 Most cases have been familiar in character, and a variety of pathological changes in the basal ganglia2 3 have been described. Occasional acquired cases have been reported after brain injury,4 and sporadic cases of no apparent cause have occurred, sometimes without historical-pathological abnormality.5 6 A pathological basis for the occasionally encountered dystonic movements after initiation of motor activity has recently been suggested.7 Paroxysmal choreoathetosis after head injury has previously been reported,8 but distinction between these involuntary movements and partial seizures may be difficult, and is important because of the controversies regarding post-traumatic epilepsy and its prophylaxis.9 10 We have recently studied three patients with paroxysmal choreoathetosis after closed head injury.

Case 1, a 22-year-old male sustained closed head injury in a head-on automobile accident. He had small right and left epidural hematomata which were evacuated within 72 hours, and was comatose for two weeks and lethargic and quadriparetic for another eight weeks. He made full recovery except for labile affect and mild spasticity of the right arm and leg. Four weeks after recovering consciousness, he developed paroxysmal involuntary movements of sudden onset and one to two minutes' duration, which could be precipitated by abduction and external rotation of the right shoulder; similar movements of the right leg were less frequently precipitated by extension of that limb. Attacks consisted of clonic jerking at the shoulder, writhing of the fingers, and spasmodic flexion or rotation of the leg. They could be aborted immediately by returning and lifting the arm to its previous position, or by grasping the arm or leg with the left hand.

Pattern-reversal visual and brainstem auditory evoked responses were normal six weeks after onset of the movements. EEG was unremarkable, and showed no change during a characteristic attack precipitated by right shoulder rotation. Diazepam produced minimal change and sedation. Phenytoin brought temporary relief but the attacks returned, and increasing dose requirements led to toxic symptoms at high-therapeutic serum levels. The attacks have been relieved with phenobarbitone 120 mg at bedtime.

Case 2, a 30-year-old male fell from a telephone pole and sustained closed head injury and four linear skull fractures. He was comatose for two weeks and minimally responsive for six weeks. He had a dense spastic right hemiparesis which gradually improved, a dense nonfluent aphasia which improved to permit nonverbal communication, and intact comprehension. Involuntary right arm movements began six weeks after regaining consciousness; he had no leg involvement but occasional jerking of the intercostal muscles, and prodromal sensations of pain and fatigue in the right arm. Attacks involved several minutes of clonic arm jerking, alternating with pronation or supination, and similar periods of fisting in alteration with writhing of the fingers. They were precipitated by attempts to lift the arm or bend the elbow, and usually did not respond to pressure but gradually improved if he grasped the right arm or concentrated intensely on the arm.

Approximately one year after injury, pattern-reversal visual evoked responses were normal, and brainstem auditory evoked responses showed attenuation of wave V from right ear stimulation in the presence of normal auditory thresholds, suggesting a mid-brain disturbance. EEG was normal, and a characteristic attack occurred spontaneously and without correlate. He was excessively sedated with diazepam, did not improve on phenytoin, and was fatigued on phenobarbitone. Carbamazepine stopped the attacks but he developed nausea and diplopa. The movements have been controlled with lorazepam 2 or 3 mg as needed at the onset of prodromal symptoms.

Case 3, an 18-year-old male sustained closed head injury and suspected brainstem contusion in a moped accident. He was unconscious for three days, and was then confused and agitated for ten days, followed by a complete recovery except for subjective mental slowing. Five weeks after recovery of consciousness he developed ballistic flailing of the right arm and less frequent clonic jerking of the right leg, which lasted for several minutes and could not be precipitated but were worsened by attempts to move after the attacks began, and were lessened by grasping or squeezing the involved limbs.

Pattern-reversal visual and brainstem auditory evoked responses were normal about one year after injury. EEG showed intermittent left centrotemporal slowing; a characteristic attack was not recorded, but was subsequently observed and could be terminated by pressure on the right arm. The attacks have been relieved by phenobarbitone 100 mg nightly.

These patients developed attacks typical of paroxysmal choreoathetosis four to six weeks after recovery of consciousness from head injury. They showed no cerebral correlate on EEG to movement-induced or modified clonic or choreoathetoid spasms, but responded well to anticonvulsants although phenytoin and carbamazepine were less successful than lorazepam and phenobarbitone. There was no evidence by examination or CT scan of identifiable injury to the basal ganglia or elsewhere.

The basal ganglia have been felt clinically to be the site of origin for these movements,1 although they have been classified in some reports as subcortical or reflex-induced seizures.2 3 The infrequent cases associated with acquired brain disease have been characterised clinically by both diffuse and focal basal ganglia dysfunction.4 Pathologically, asymmetry of the substantia nigra5 or melanin pigmentation of the locus ceruleus suggesting neuronal loss6 have been described, while sporadic cases with normal necropsy or brain biopsy findings were described by Lance7 and Gilroy.8 Sunohara et al.9 reported a patient with action-induced dystonic movements resembling our patient's, who had an infarct in the right posterior lateral ventral thalamus. Paroxysmal choreoathetosis after closed head injury was reported by Robin,5 also in a patient with no previous basal ganglia disease and no evidence of subcortical dysfunction after recovery from a diffuse encephalopathy. Clinical evidence of focal basal ganglia signs after diffuse brain injury is well described.11 Closed head injury might cause more discrete subcortical dysfunction through Duret haemorrhages.12 Although one patient was initially suspected of having brainstem contusion and another had attenuation of wave V of the brainstem auditory evoked responses, none of the patients had identifiable brainstem injury.

The heterogeneity of our patients' movements, lack of discernible EEG alteration, and susceptibility of attacks to change by pressure and movement might argue that the choreoathetosis is a movement disorder of subcortical origin, while a consistent latency of onset after injury may suggest aberrant reorganisation of motor patterns after injury. The consistent response to anticonvulsants could support the possibility of partial seizures, and partial seizures with repetitive movements.
normal ictal EEG have been described previously.13 The anticonvulsants effective in our patients have widespread membrane effects14 and have been used successfully in a number of paroxysmal symptoms other than seizures.15 The distinction between seizures and involuntary movement disorder in this condition may be artificial.

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References

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Behcet syndrome presenting as cerebrovascular disease

Sir: Initially described as a triad of recurrent oral aphthous stomatitis, genital ulceration and uveitis, Behcet disease is now known to be a systemic illness that in 10% to 25% of patients involves the nervous system.1 2 The neurological symptoms usually begin six months to several years after the mucocutaneous manifestations, and often occur concomitantly with exacerbations of the mucocutaneous lesions.3 4 In rare patients, however, the neurological manifestations may precede other signs of Behcet disease.4 6

Meningoencephalitis, often affecting the brain stem, is the most common neurological involvement. It is characterised by fever, headache, stiff neck, and focal neurological deficits such as hemiparesis, quadriparesis, cerebellar deficits, cranial nerve palsies, and aphasia. Other neurological manifestations include seizures, encephalopathy, pseudotumour cerebri, myelitis and peripheral neuropathy.1–13 Occlusion of cerebral arteries is either considered not to be a feature of this disease2 or to occur only in rare instances.14 We report a patient with Behcet disease who presented with cerebral transient ischaemic attacks that antedated mucocutaneous manifestations of the disease by several years.

A 43-year-old white man was first examined in November 1971 when, in the course of one week, he suffered three transient episodes of inability to talk and weakness and paraesthesias of the right upper extremity that each lasted approximately three hours. In the prior two years he had experienced migratory arthralgia of large joints as well as occasional myalgia. There was no history of hypertension or cardiovascular disease, and no risk factors for vascular disease. Blood pressure was 100/60 mm Hg, pulse 80 and regular, temperature 37·1°C. He ran an intermittent and low grade fever, and had left axillary and bilateral inguinal lymphadenopathy. Neurological examination was normal. Complete blood counts, erythrocyte sedimentation rate, serum electrolytes, glucose, urea nitrogen, creatinine, calcium, phosphorus, bilirubin, cholesterol, triglycerides, uric acid, lactic dehydrogenase, aldolase, transaminases, creatine kinase, coagulation time, prothrombin time, partial thromboplastin time, fibrinogen, and protein electrophoresis were normal. Venereal disease research laboratory (VDRL), fluorescent treponemal antibody absorption test (FTA-ABS), latex rheumatoid factor (RF) and fluorescent antinuclear antibody test (ANA) were negative. Urinalysis was normal. Creatine and creatinine in serum and urine were also normal. Lumbar puncture showed an opening pressure of 150 mm H2O. The cerebrospinal fluid (CSF) contained no red cells, 20 leukocytes/mm3 (90% lymphocytes, 3% monocytes, 7% polymorphonuclear leukocytes) and normal glucose.

Total protein was 65 mg/dl (upper limit of normal: 45 mg/dl), and protein electrophoresis showed prealbumin 3·4 mg/dl (5·2%), albumin 23·2 mg/dl (35·7%), alpha-1-globulin 2·0 mg/dl (3·1%), alpha-2-globulin 3·3 mg/dl (5·1%), beta globulin 5·3 mg/dl (8·2%) and gamma globulin 27·8 mg/dl (42·7%) (upper limit of normal: 13%). VDRL and FTA-ABS in the CSF were negative. Blood, urine and CSF cultures were negative. The EEG showed a focal theta activity over the left parietal temporal regions that was accentuated by hyperventilation. CT scan was not available at the time. Pneumoencephalogram, muscle and lymph node biopsy specimens were normal. Left carotid angiography demonstrated minor irregularities of the internal carotid artery and, with the first injection of contrast, a very high grade stenosis of the left middle cerebral artery approximately 1·5 cm from its origin. With a second contrast injection, a total occlusion of the previously stenotic area was observed. The patient immediately developed non-fluent aphasia and a right hemiparesis. Examination showed decreased verbal output with inability to repeat and write but relatively spared auditory and written comprehension, mild right lower face weakness, tongue deviation towards the right, marked weakness of the right upper extremity involving more severely the distal than proximal muscles, mild right leg weakness and slight right-sided hyperreflexia. Recovery began ten minutes after the onset of symptoms and was almost complete in three hours. Mild weakness of the right deltoid, triceps, supinator