Acquired focal dystonia following recovery from central pontine myelinolysis

Sir: Focal dystonia and hemidystonia are rare disorders of movement characterised by forceful and prolonged simultaneous contraction of agonist and antagonist muscles which distort the affected extremities into typical postures. Although the precise pathophysiological basis of hemidystonia is undetermined, symptomatic hemidystonia has been described in association with focal structural brain lesions including tumour, infarction, haemorrhage and arteriovenous malformations as well as several inflammatory and metabolic disorders (such as Wilson's disease, Leigh's disease, syphilis, and encephalitis). We describe a patient who developed monomeric dystonia following recovery from central pontine myelinolysis (CPM).

A 57 year old man presented to a community hospital with confusion after a prodrome of viral gastroenteritis. He had a long history of alcohol use without any associated deleterious medical complication. He had no prior history of any movement disorder, neuroleptic use, or focal dystonia. Initial neurological examination revealed an awake, disoriented man. Laboratory evaluation revealed a serum sodium of 100 mmol/l. Replacement therapy with normal saline was initiated at a rate of 1-4 mmol/l/hour over the first 24 hours to a level of 128 mmol/l. The patient's sensorium cleared. Two days later, the patient developed diplopia followed by quadriplegia and a pseudobulbar affect. Two CT scans during the first 2 weeks of admission were normal and CSF studies were unremarkable. The clinical impression of central pontine myelinolysis was confirmed by magnetic resonance imaging (MRI) performed 16 days following correction of the hyponatraemia (fig).

The patient was transferred to the Seattle Veterans Hospital one month following his initial presentation. Neurological examination revealed an awake patient who could follow three step commands. He had a pseudobulbar affect with marked dysarthria and dysphagia. Extraocular movements were full. Sensation was intact. There was a symmetric spastic quadriplegia. He was diffusely hyperreflexic with bilateral extensor planar responses. Over the next 4 months, the patient's deficits largely resolved. However, as the patient's gait and hand function improved, he slowly developed a painful action dystonia primarily involving the left upper extremity with hyperextension of the proximal phalanges, abduction and hyperextension of the thumb and external rotation of the arm.

Palpable co-contraction of the flexors and extensors of the forearm was present. The dystonic movement was most prominent during walking or active use of either arm. When the patient was at complete rest, there was no increased tone or spasticity in any limb; however, he remained symmetrically hyperreflexic. Plantar responses became normal. Repeat MRI scans 5 and 7 months after his initial deterioration revealed no evidence of any extrapontine lesion. The large pontine lesion remained unchanged.

The patient was treated empirically with benztropine (1 mg t.i.d.) with an objective decrease in the severity of the dystonia. Therapy was discontinued owing to urinary retention. Subsequent patient trials of carbidopa/levodopa and diphenhydramine had no effect and trihexyphenidyl only minimal effect on the dystonia.

Central pontine myelinolysis was originally described by Adams, et al. Our patient exhibited the typical finding of rapidly evolving quadriplegia with prominent pseudobulbar symptoms following rapid correction of severe sodium depletion. Initially CPM was felt to be almost uniformly fatal. Our case and several other recent reports demonstrate that a significant degree of recovery from CPM is possible. This apparent change in the prognosis is most likely related both to increased recognition of the syndrome and to the development of improved imaging techniques which have allowed premorbid diagnosis. As in several other cases, CT scanning failed to demonstrate the brainstem lesion which was readily detected with MRI.

Symptomatic dystonia has not previously been fully reported as a sequelae of central pontine myelinolysis. Examination of our patient following recovery from the quadriplegic state of acute CPM revealed a movement disorder consistent with focal dystonia. There was no evidence in the history to suggest that the CPM was amplifying an already present action dystonia, nor evidence to suggest the presence of any other drug induced or familial dystonia. Although a definite causal relationship cannot be made, the temporal sequence between recovery from CPM and subsequent development of a profound dystonia in this patient is striking. As in other cases of focal dystonia, the onset of symptoms occurred with a significant delay after the initial nervous system insult, in this case 4 months.

Focal dystonia has been reported as a complication of a variety of structural lesions. CT, neuropathological studies, and a limited number of MRI investigations have demonstrated that, most commonly, lesions involve the caudate nucleus (particularly the head of the caudate), the putamen, or the thalamus. Extrapontine myelinolysis occurring with CPM is a well known post-mortem finding and has been demonstrated by premortem MRI. Few of these patients have developed movement disorders. Of the reported MRI studies performed to date on patients with CPM only one case had an action dystonia and this MRI demonstrated the typical abnormalities within the basis pontis and only "subtle changes within the basal ganglia". Despite the relative sensitivity of MRI, we were unable to demonstrate with serial studies any abnormality other than the single lesion within the pons. No prior reports of dystonia occurring in patients with lesions limited to the brainstem exist. The anatomic basis of the dystonia seen in our patient as well as the movement abnormalities in other cases of CPM remains uncertain. The findings here suggest that dystonia might also be induced by limited brainstem injury. Further neuropathological and MRI correlations are needed.

This work was presented in part at the thirty-ninth annual meeting of the American Academy of Neurology, New York, NY, April, 1987 and was supported in part by Veterans Administration medical research funds.

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Fig. Mid-sagittal section of a T1-weighted magnetic resonance image generated with a 0-5 Tesla magnet demonstrating an ovoid lesion in the basis pontis. Image obtained 16 days after correction of hyponatraemia.
Reversible intracranial circulatory arrest in acute subarachnoid haemorrhage

Sir: Recently there was a report in this journal on cerebral blood flow velocities measured in the middle cerebral artery (MCA) using transcranial Doppler sonography (TCD) in 17 children developing brain stem death due to various causes. The authors stated that with mean flow velocities of less than 10 cm/s and/or substantial diastolic reverse flow observed over a 30 minute period, recovery of brain stem reflexes or holodiastolic forward flow was never encountered in their series. TCD is indeed a valuable monitoring method in such cases of intracranial hypertension and intracranial circulatory arrest. So-called oscillating Doppler profiles of the MCA, that is, biphasic patterns with equivalent inflow and outflow components, correctly predicted angiographical non-filling of the anterior intracranial circulation in a series of eight patients. Extracranial obstruction of contrast was demonstrated in other cases even up to slightly positive net time averaged mean velocities of 6 cm/s in the basal MCA.

Biphasic Doppler spectra are due to back and forth movement of the arterial blood column in the presence of distal outflow obstruction. However, as observation periods in clinical TCD application tend to be shorter than 30 minutes, attention must be drawn to transient elevations of intracranial pressure (ICP) that can produce a reversible cerebral circulatory arrest and an biphasic TCD pattern with subsequent survival of the patient and return of normal TCD flow after a few minutes. This was encountered by the authors in three patients during acute aneurysmal subarachnoid haemorrhage in whom a transient circulatory arrest in the MCA could be documented by simultaneous TCD recording (fig). Hornes et al have reported on their observations of increasing ICP and blood flow approaching zero in the extracranial internal carotid artery in patients with current subarachnoid haemorrhage. It was assumed that...