SHORT REPORT

# Parkinsonism and dystonia in central pontine and extrapontine myelinolysis

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**Abstract**

**Parkinsonism as well as dystonic signs are rarely seen in central pontine myelinolysis and extrapontine myelinolysis. A 51 year old woman developed central pontine myelinolysis and extrapontine myelinolysis with parkinsonism after severe vomiting which followed alcohol and drug intake, even though marked hyponatraemia had been corrected gradually over six days. Parkinsonism resolved four months after onset, but she then exhibited persistent retrocollis, spasmodic dysphonia, and focal dystonia of her left hand. Although the medical literature documents three similar patients, this patient is different as dystonic symptoms only developed four months after parkinsonian signs had resolved.**

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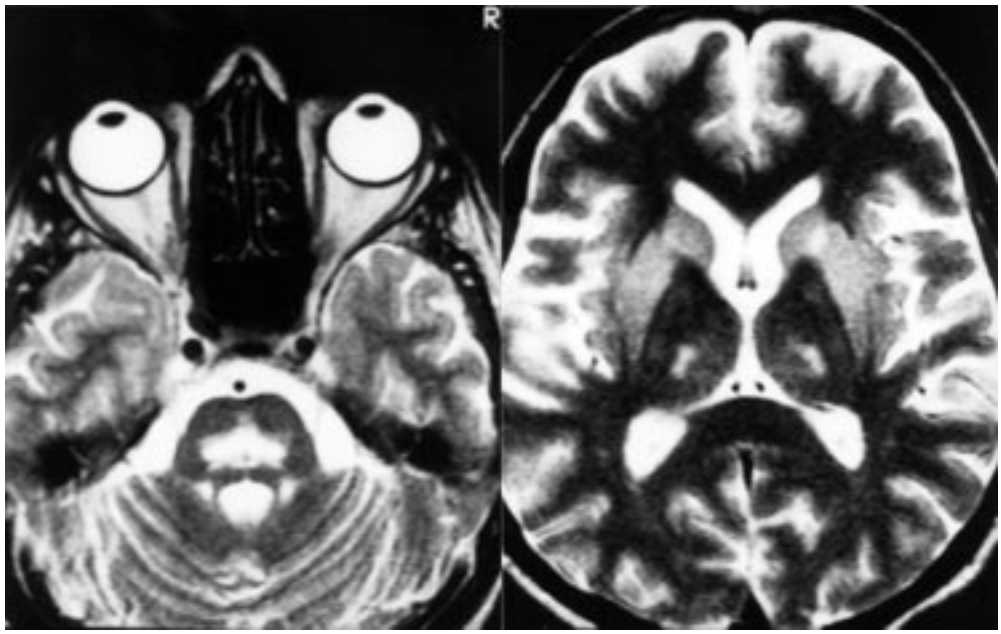
Keywords: central pontine myelinolysis; extrapontine myelinolysis; parkinsonism; dystonia

Central pontine myelinolysis is a well defined syndrome characterised by various degrees of tetraparesis and brain stem symptoms as a sequel of rapid correction of electrolyte disturbances, hyponatraemia in particular. In more severe cases, additional demyelination may occur in extrapontine locations, giving rise to parkinsonian signs and symptoms. Tomita *et al* recently reported on a patient with extrapontine myelinolysis, exhibiting symptoms of parkinsonism and dystonia in his fingers.[1] Here we report a case in which parkinsonism and dystonia occurred sequentially within a period of four months and speculate that preclinical damage due to regular alcohol intake might have predisposed our patient to develop dystonic signs only after parkinsonism had subsided.

## Case report

A 51 year old female patient with a history of repeated alcohol and drug misuse was admitted with severe and persisting vomiting after alcohol and drug intake. She became increasingly weak and drowsy and was only able to take a few assisted steps. Gastroscopy showed haemorrhagic gastritis and duodenitis. The admission values for serum sodium, chloride, and potassium were 93 mmol/l, 58 mmol/l, and




*T2 weighted images show a symmetric signal increase in the central pons region, putamen, caput nuclei caudati, and lateral thalamus.*

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*Cases of parkinsonian, dystonia, and both after extrapontine myelinolysis*

| *Reference* | *n* | *Prominent clinical signs* |
|---|---|---|
| Parkinsonian: | | |
| Stam *et al*[8] | 1 | Tremor, mask-like facial expression |
| Dickoff *et al*[5] | 1 | Rest tremor, cogwheel rigidity facial hypomimia, dysphagia, bradykinesia, retropulsion |
| Kurlan *et al*[6] | 1 | Akinetic-rigid features, dysarthria dysphagia |
| Tinker *et al*[9] | 1 | Impassive face, bradykinesia, rest tremor, cogwheel rigidity, parkinsonian gait |
| Maraganore *et al*[7] | 1 | Slow resting tremor, parkinsonian gait |
| Sadeh *et al*[4] | 1 | Facial hypomimia, dysarthria, hypokinesia and bradykinesia, resting tremor, cogwheel rigidity |
| Dystonia: | | |
| Grafton *et al*[10] | 1 | Focal action dystonia, dysarthria |
| Kurlan *et al*[6] | 1 | Dystonia (limb, trunk, orolingual) |
| Thompson *et al*[11] | 1 | Mobile dystonic posturing, wide based gait, dysarthria (2a) |
| Tison *et al*[12] | 1 | Generalised dystonia, choreoathetosis, dystonic posturing |
| Maraganore *et al*[7] | 2 | Generalised action dystonia, athetoid movements |
| Parkinsonian and dystonia: | | |
| Kurlan *et al*[6] | 1 | Dystonia, akinetic-rigid features |
| Niwa *et al*[13] | 1 | Dystonia and rigidity |
| Tomita *et al*[1] | 1 | Mask like face, bradykinesia, difficulties protruding the tongue, dysarthria, parkinsonian posture finger dystonia |

2.9 mmol/l, respectively. The electrolytic indices were gradually corrected over the subsequent six days using intravenous physiological saline solution under permanent cardiac monitoring. She then became markedly slowed but was fully oriented and of normal intelligence and general knowledge. However, recent memory and ability to learn were impaired. She exhibited cogwheel rigidity of all four limbs, bradykinesia, facial hypomimia, monotonous speech, and parkinsonian gait with associated retropulsion. Intermittent action myoclonus was seen. The deep tendon reflexes were brisk with bilateral extensor plantar responses. The grasp reflex was elicited bilaterally but there was no sensorimotor weakness, only slight dysarthria.

A low dose levodopa/benserazide therapy was started. The parkinsonian signs disappeared. However, an examination four months after admission showed that she had developed marked retrocollis, an oromandibular dystonia with difficulties in opening her mouth and protruding her tongue, and severe dysphagia which required transient feeding through a nasogastric tube, as well as spasmodic dysphonia and focal dystonia of her left arm with severe functional impairment. Subsequently, the dysphagia, retrocollis, and oromandibular dystonia improved. However, the focal dystonia of the right arm and the spasmodic dysphonia were still present during a follow up period of 20 months, despite treatment with tiaprid and perphenazine.

Brain MRI showed a signal increase in the central pons on T2 weighted images but also bilateral hyperintense areas within the putamen, caput nuclei caudati, and lateral thalamus (figure). Subsequent control images made up to six months after the onset of the condition showed a marked decrease of these signal intensities. An EEG disclosed diffuse slow background activity and bilateral theta and delta activity which improved gradually during the subsequent months. Brain stem auditory evoked potentials measured eight months after onset of disease were normal, as were CSF and a β-CIT SPECT examination of the brain.

## Discussion

Central pontine myelinolysis results from rapid correction of hyponatraemia and generally presents with tetraparesis and various degrees of brain stem dysfunction such as pontine dysfunction, pseudobulbar palsy, and, occasionally, locked in syndrome.[2] Extrapontine myelinolysis occurs in about 10% of patients with central pontine myelinolysis.[3] Extrapyramidal symptoms are, however, rarely seen, as they are often masked by involvement of the pyramidal tract and brain stem.[4] To date, six such patients with parkinsonism have been described. The table shows the symptoms.[4–9] Most patients responded to dopaminergic therapy. Brain MRI disclosed hyperintense lesions in the striatum, especially in the putamen and the caput nuclei caudati. Dystonia was reported in six patients (table).[6–7 10–12] The onset was delayed in most patients and response to treatment was variable. In no patients did the pathological changes visualised on MRI fully explain the dystonic symptoms. Three further patients developed a combination of simultaneous parkinsonian and dystonic symptoms (table).[1 6 13] However, our patient is the first to develop dystonic symptoms within four months and only after complete regression of parkinsonism had occurred. In addition, it is remarkable that central pontine myelinolysis developed despite the relatively slow and delayed correction of hyponatraemia. It seems feasible that the rather low initial serum electrolyte concentrations, as well as pre-existing alcohol and drug misuse increased the risk of myelin damage.[14]

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