Parkinsonism and dystonia in central pontine and extrapontine myelinolysis

A Seiser, S Schwarz, M M Aichinger-Steiner, G Funk, P Schnider, M Brainin

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. (J Neurol Neurosurg Psychiatry 1998;65:119–121)

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

<table>
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<th>Reference</th>
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<th>Prominent clinical signs</th>
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<tr>
<td>Parkinsonian:</td>
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<td>Tremor, mask-like facial expression</td>
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<td>Dickoff et al 4</td>
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<td>Rest tremor, cogwheel rigidity facial hypomimia, dysphagia, bradykinesia, retropulsion</td>
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<td>Kurlan et al 6</td>
<td>1</td>
<td>Akinetiform features, dystartheria dystaphy</td>
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<td>Tinker et al 6</td>
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<td>Impassive face, bradykinesia, rest tremor, cogwheel rigidity, parkinsonian gait</td>
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<tr>
<td>Maraganore et al 17</td>
<td>1</td>
<td>Slow resting tremor, parkinsonian gait</td>
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<tr>
<td>Sadeh et al 5</td>
<td>1</td>
<td>Facial hypomimia, dystartheria, hypokiniesia and bradykinesia, resting tremor, cogwheel rigidity</td>
</tr>
</tbody>
</table>

Dystonia:

| Grafom et al 11 | 1 | Focal action dystonia, dystartheria |
| Kurlan et al 6 | 1 | Dystonia (limb, trunk, orolingual) |
| Thompson et al 33 | 1 | Mobile dystonic posturing, wide based gait, dystartheria (2a) |
| Tison et al 33 | 1 | Generalised dystonia, choreoathetosis, dystonic posturing |
| Maraganore et al 17 | 2 | Generalised action dystonia, athetoid movements |

Parkinsonian and dystonia:

| Kurlan et al 6 | 1 | Dystonia, akinetic-rigid features |
| Niwa et al 17 | 1 | Dystonia and rigidity |
| Tomita et al 17 | 1 | Mask like face, bradykinesia, difficulties protruding the tongue, dystartheria, parkinsonian posture, finger dystonia |

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. Extrapyramidal symptoms are, however, rarely seen, as they are often masked by involvement of the pyramidal tract and brain stem. To date, six such patients with parkinsonism have been described. The table shows the symptoms. 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). 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). 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.

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