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Plasmapheresis improves the outcome of central pontine myelinolysis

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Sirs: Central pontine myelinolysis (CPM) is a demyelinating disease of the pons rarely involving other areas of the central nervous system. The etiology and the pathogenesis of CPM remain unclear, though aggressive osmolar correction and in particular rapid correction of hyponatremia are the main factors in treating patients at risk. There is no specific therapy of choice. Several uncontrolled therapeutic trials have led to a substantial improvement of the prognosis which had always been deemed uniformly bad [1]. Bibl et al. successfully treated three young female patients with extensive therapeutic plasmapheresis soon after the diagnostic confirmation of CPM by magnetic resonance imaging [2]. All patients had undergone correction of severe hyponatremia three to five days before the onset of neurological symptoms comprising a rapidly evolving flaccid quadriplegia with dysphagia and dysarthria. Significant clinical improvement was obtained one month after plasmapheresis and neurological examination one year later disclosed partial recovery up to total remission in one patient. The assumption is that undefined myelin toxic compounds released by the osmotic stress contribute to the irreversible demyelinating process in CPM and that therapeutic plasmapheresis may reduce high-molecular myelin toxic substances leading to clinical improvement. We report another case of CPM dramatically improved by plasmapheresis started a week after the onset of neurological symptoms.

A 59-year-old woman with a history of mild alcohol abuse and mild hypertension treated by a thiazid diuretic gradually developed general malaise followed by motor weakness and mental slowness. Within two weeks the patient became lethargic and was admitted to hospital. Blood tests revealed severe hyponatremia (113 mmol/L) that was rapidly adjusted over three days to normal values by infusing hypertonic saline, but remaining within a target volume not above 10 Meq/L/24 hours. Correction of hyponatremia resulted in a complete recovery of mental status and motor function. Three days after recovering, the patient's consciousness rapidly deteriorated again. In a few days she developed slurred speech, dysphagia for liquid foods and legs weakness. A week later neurological examination disclosed pseudobulbar palsy with flaccid tetraplegia associated with deep-tendon hyperreflexia, pathological crying, impairment of ocular movements, anarthria and dysphagia. Magnetic resonance imaging (MRI) performed on admission showed an area of hyperintensity in the central pons on T2-weighted images suggestive of CPM (Fig. 1). Ten consecutive therapeutic plasmapheresis sessions were started with two to three sessions a week for one month with a total of 37300 mL plasma exchanged with albumin 3 % and crystalloids. Significant clinical improvement began ten days after plasmapheresis (Fig. 2)

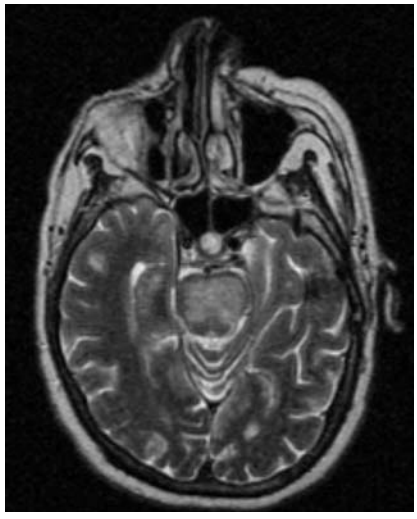

Fig. 1 Axial T2-Weighted MRI image showing the central hyperintense pontine lesion on admission, before treatment

continuing until intensive rehabilitation was possible. Eight months later, a mild tetraparesis with the ability to walk unaided, slight dysphonia and dysarthria were detected on neurological examination. Despite clinical improvement, the pontine lesion and signal hyperintensity on MRI T2-weighted images appeared to be unmodified. The positive outcome in our patient and the strong temporal link between treatment onset and clinical improvement, support the hypothesis of plasmapheresis as an effective treatment for CPM. However, the possibility of a spontaneous good outcome cannot be excluded [3].

Because of the severity of the neurological deficits especially in the acute phase and the possibility of a poor prognosis up to death, prompt, safe and apparently effective treatment with plasmapheresis should be considered once CPM is diagnosed.

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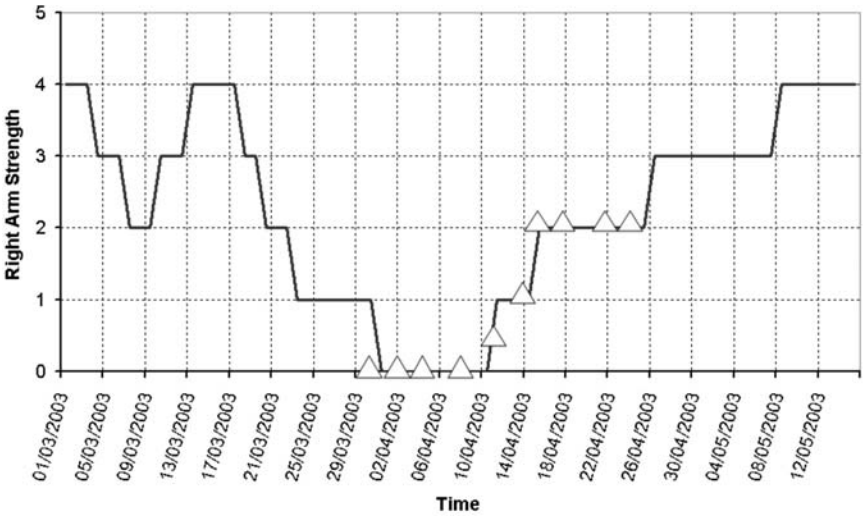

Fig. 2 Relation between improved right arm strength and treatment with plasma exchange. Note the change in disease course ten days after plasmapheresis

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