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Short communication

"Man-in-the-barrel" syndrome as delayed manifestation of extrapontine and central pontine myelinolysis: Beneficial effect of intravenous immunoglobulin

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Abstract

"Man-in-the-barrel" syndrome has been rarely described following osmotic myelinolysis. We report a case of a 45-year-old woman admitted with septicemia and severe hyponatremia. She presented with a "man-in-the-barrel" syndrome which developed more than 10 days after rapid correction of the hyponatremia. There was radiological evidence of central pontine and extrapontine myelinolysis. Three days after completing a course of intravenous immunoglobulin therapy (0.4 g/kg body weight/day for 5 days) there was considerable improvement (Expanded Disability Status Scale score improved 30%). This case, reported for its peculiar mode of development, unusual presentation and challenging therapeutic response to intravenous immunoglobulin, highlights the enigmatic and unpredictable aspects of osmotic myelinolysis.

Keywords: Central pontine myelinolysis; Magnetic resonance imaging; Extrapontine myelinolysis; Hyponatremia; Intravenous immunoglobulin; Man-in-the-barrel syndrome

1. Introduction

Osmotic demyelination disorders remain esoteric but fascinating to the clinician. The most eminent central pontine myelinolysis (CPM) has classically been reported following rapid fluxes in the patient's serum sodium concentration secondary to the rapid correction of hyponatremia of any cause (including diuretic therapy) and more recently following orthotopic liver transplantation [1,2]. However osmotic demyelination may also occur when serum sodium levels are normal or elevated, or when hyponatremia is corrected within limits considered safe. In addition, it also rarely occurs in conditions with extreme serum hyperosmolality (e.g. mannitol infusion, diabetic ketoacidosis) or debilitating neurological disorders such as motor neuron disease [1]. Although two clinical distinct variants of osmotic demyelination have been described; CPM and extrapontine myelinolysis (EPM), findings seem to indicate that in over 75% of cases with CPM there is association with EPM, affecting the basal ganglia, cerebellum, and the lateral geniculate nucleus [3,4]. CPM has a variable and wide clinical spectrum, presented in its mild form with confusion, mutism, dysarthria, and in its severe form with rapidly evolving flaccid quadriplegia, pseudobulbar palsy with inability to swallow, and occasionally lock-in syndrome or coma [1].

"Man-in-the-barrel" syndrome refers to a clinical syndrome of brachial diplegia. Classically, it has been associated with cerebral hypoperfusion resulting from watershed infarctions between the vascular territories of

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the middle and anterior cerebral arteries [5]. We report a patient who developed a "man-in-the-barrel" syndrome as a delayed complication of CPM and EPM, and with radiological evidence of osmotic pontine and EPM. This in addition to a beneficial response to intravenous immunoglobulin makes this case a unique observation.

2. Case report

A 45-year-old Sri Lankan woman with a past history of dyslipidemia and chronic hypertension treated for the last 10 years with indapamide and atenolol was admitted through the Accident and Emergency Department presenting with subacute onset brachial diplegia. There was no past history of chronic alcohol abuse, exposure to neuroleptic drugs or toxic chemicals.

She was discharged 5 days before in an apparently healthy condition. At her initial admission, 2 weeks before and which lasted 9 days, she presented with a 1-week history of fatigue, anorexia, vomiting and fever. At that time, laboratory results showed severe hyponatremia (serum sodium 105 mmol/L), hypokalemia (serum potassium 2.4 mmol/L), hypocalcemia (2.06 mmol/L), hypophosphatemia (0.17 mmol/L) and serum hyposmolarity (212 mosM/L). Her cell blood count showed a white cell count of 17.4×10^9/L with 93% neutrophils, hematocrit of 36.4% and normal hemoglobin and platelet count. Blood cultures were positive for *Escherichia coli*, indicating septicemia, which was treated with intravenous fluid and electrolyte replacement (see below), antibiotics (ceftriaxone) and steroid replacement therapy (hydrocortisone and fludrocortisone). The hyponatremia was rapidly corrected over 36 h with 3% hypertonic saline (correction rate of 15 mmol/L over 24 h) to 128 mmol/L (Fig. 1). Subsequently, hypertonic saline was substituted for normal saline and serum sodium level reached normal values over the next 2 days. Although she was confused and disoriented at first admission, physical and neurological examinations over the following days did not identify any abnormalities to suggest CPM. Hence she was discharged in an apparently good health. Five days later she was readmitted because of subacute onset dysarthria, and bilateral brachial weakness. She was found to be alert, attentive with preserved comprehension but appeared to be rather indifferent towards her clinical problem, with bursts of emotional incontinence. Although she seemed to be aphasic, prompting revealed a severe pseudobulbar speech. Fundoscopy, pupil reflexes and eye movements were intact. Gag reflex was weak. There was generalized hyperreflexia, including the jaw jerk, with equivocal plantar responses. Bilateral proximal and distal paralysis in the upper limbs (1–2/5), while strength in the lower limbs was preserved. Sensation on pinprick was intact, as she withdrew appropriately in all parts of the body. She could stand and walk. She was incontinent. There was no neck stiffness and general examination was unremarkable. Serum sodium levels and other biochemical parameters were normal. A magnetic resonance imaging (MRI) of the brain revealed well-defined crescent-shaped high intensity lesions on T2-weighted imaging symmetrically involving caudate and putamen, thalami, adjacent external capsule, as well as the central pons (Fig. 2). These neuroimaging findings were consistent with CPM and EPM. The T1-weighted MRI images of the brain were normal. Cerebrospinal fluid analysis was normal. Electroencephalogram showed generalized slowing of the background activity. She was treated supportively with nursing care. One week after admission, she was started on intravenous immunoglobulin therapy (0.4 g/kg for 5 days). Three days after completing the course the weakness in the arms started to improve

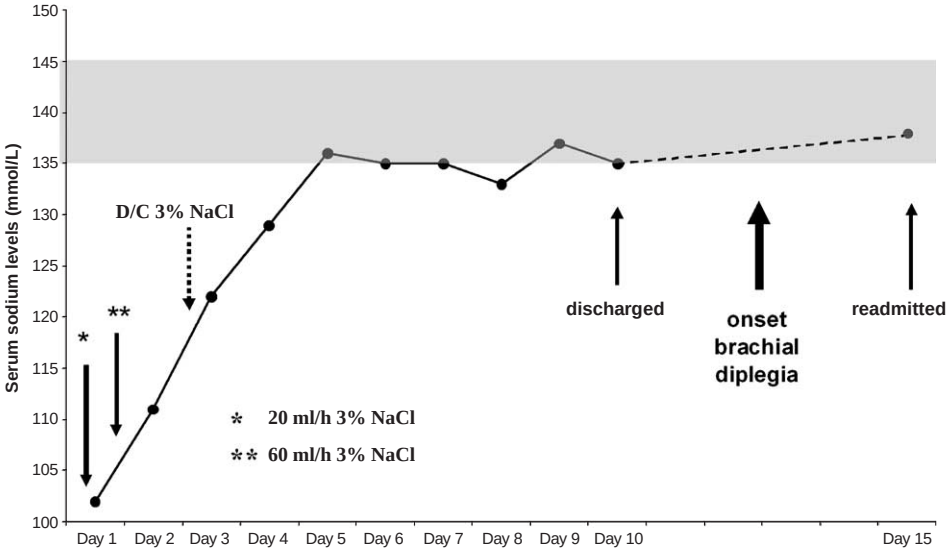

Fig. 1. Serum sodium levels and the rate of correction of the hyponatremia. Normal serum sodium range indicated by the shaded area (135–145 mmol/L). Neurological deterioration occurred around day 13. D/C, discontinuation.