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A Case of Intra- and Extra-Pontic Myelinolysis Complicating the Rapid Correction of Severe Hyponatremia

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Abstract **Case Report**

Centro and extrapontine myelinolysis (MCP-MEP) or osmotic demyelination syndrome, is a demyelination of the central part of the protuberance and other extraprotuberance territories, the main predisposing factor being too rapid correction of severe hyponatremia. We report a case of osmotic demyelination syndrome in a 37-year-old patient, followed for acute adrenal insufficiency, complicated by three cardiorespiratory arrests, with recoveries after resuscitation measures, the evolution was marked by a neurological aggravation with confusion and generalised hypotonia and myoclonic seizures. The diagnosis of centropontine and extrapontine myelinolysis was confirmed by brain magnetic resonance imaging performed seven days after an initial scan, which showed no specific abnormalities. Rapid correction of hyponatremia was the main cause of this syndrome. The course of centropontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) is variable. Treatment is mainly preventive, based on careful correction of severe hyponatremia and the factors contributing to it.

Keywords: Osmotic Demyelination Syndrome, Centropontine Myelinolysis, Extrapontine Myelinolysis, Hypnatremia, MRI.

INTRODUCTION

Centropontine and extrapontine myelinolysis (CPM, EPM) or osmotic demyelination syndrome (ODS) is a rare disorder characterised by destruction of the myelin sheaths [1]. The precise pathophysiology of this syndrome is still poorly understood. Centropontine myelinolysis was first described in 1959 by Adams and colleagues as a condition affecting alcohol-dependent and malnourished patients [2]. The association of MCP and MEP lesions is frequent and may be as high as 30% [3]. Overly rapid correction of hyponatremia remains the most common cause of this syndrome [4]. In the literature, the development of RDS can be linked to numerous predisposing factors such as inappropriate secretion of antidiuretic hormone, chronic alcoholism, malnutrition, psychogenic polydipsia, liver transplantation, dialysis, etc. [5]. Other factors such as hypokalaemia also seem to play a favourable role [4]. We report an observation of MCP and MEP occurring after rapid correction of severe hyponatremia in a patient followed for acute adrenal insufficiency.

OBSERVATION

We report the case of a 37-year-old patient, followed for acute adrenal insufficiency, complicated by three cardiorespiratory arrests, with recoveries after resuscitation measures, the evolution was marked by confusion, generalized hypotonia and convulsive seizures. An initial MRI scan of the brain revealed no significant abnormalities. Biological tests showed severe hyponatremia at 110 mmol/l. After rapid correction in the intensive care unit with isotonic saline at a rate of more than 1 mmol/l/h, natraemia rose to 136 mmol/l after 24 hours in hospital (an increase of 25 mmol/l in 24 hours). 48 hours after this rapid correction of hyponatraemia, the patient was still suffering from neurological deterioration, with a control natraemia of between 132 and 134 mmol/l. A brain MRI (Figure 1, Figure 2) was repeated.

Seven days after his first imaging examination, he developed bi-thalamic nodular signal anomalies with T1-isolated signal, T2-hyper-signal, T2-flare signal, diffusion hypersignal with high ADC and associated centro-pontine signal anomalies with T1-isolated signal, T2-hyper-signal, diffusion hypersignal with discrete ADC restriction. The diagnosis of MCP and MEP was

accepted. Management consisted of daily monitoring of his blood ionogram, daily respiratory physiotherapy, enteral feeding, and gastric and thromboembolic protection. The outcome was unfavourable, with the patient dying after two weeks in hospital following pulmonary-onset septic shock.

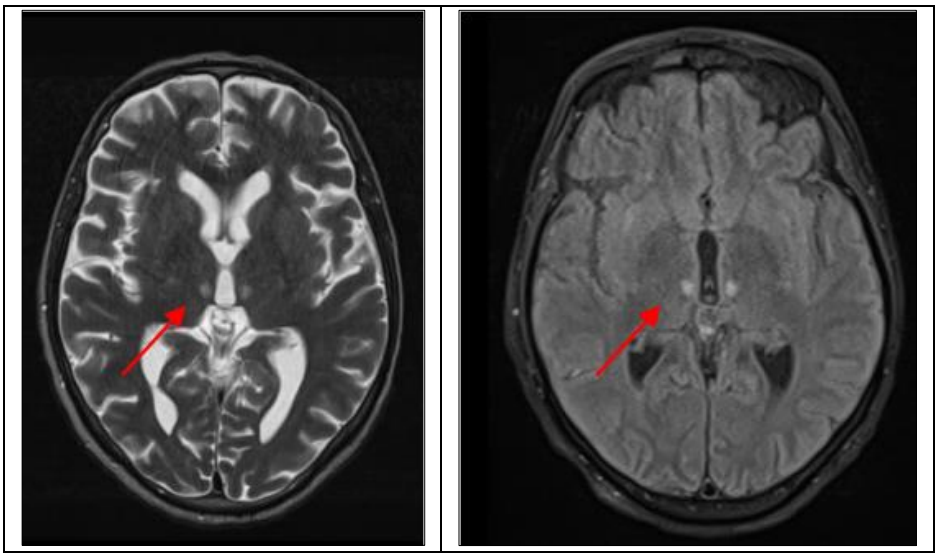

Figure 1: Cerebral MRI in axial section in T2 and T2 Flair sequences: showing a nodular and symmetrical bi-thalamic (arrow) hypersignal in favour of extrapontine myelinolysis.

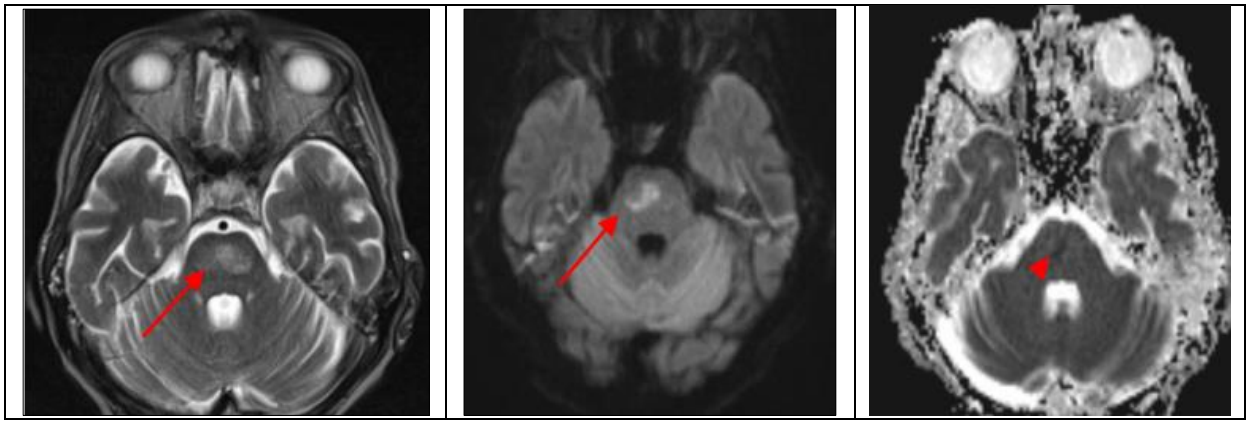

Figure 2: Cerebral MRI in axial section in T2 and diffusion sequences: centro-pontine signal anomalies in T2 hypersignal, diffusion hypersignal (arrow) with discrete ADC restriction (arrowhead) in favour of intrapontine myelinolysis.

DISCUSSION

Centropontine and extrapontine myelinolysis are grouped together under the term osmotic demyelination syndromes (ODS) [6]. To date, the true incidence of this condition is unknown [7]. Microscopically, it involves symmetrical destruction of myelin in all nerve fascicles associated with a loss of oligodendrocytes [2]. In MCP, one study showed the presence of significant axonal lesions associated with an inflammatory infiltrate [8]. Rapid correction of a chronic osmolar disorder in cases of organic osmolyte deficiency exposes brain cells, particularly oligodendrocytes, to the risk of shrinkage and consequent demyelination [9]. In a study of 22 cases with hyponatremia, hypokalemia was found to be a predisposing factor in 7 cases of MCP [10]. In cases of hypokalaemia, the decrease in NaK-ATPase concentration in the endothelial cell membrane may predispose the cell to damage by the osmotic stress associated with the rapid increase in natraemia [11].

Diagnosis of MCP and MEP is essentially clinical, with symptoms generally following a biphasic course. Initially, the patient may present with encephalopathy, which may recover rapidly after restoration of natraemia. Several days later, the second phase is characterised by the appearance of dysarthria and dysphagia secondary to cortico-bulbar damage, and flaccid quadriparesis which then becomes spastic secondary to cortico-spinal damage [6].

Cerebral CT is not very sensitive for diagnosis [12]. Cerebral MRI is the radiological examination of choice, showing central demyelinating lesions intra-pontically and symmetrically extra-pontically with T1-weighted hyposignal and hypersignal on T2-weighted

and FLAIR sequences without enhancement after injection of Gadolinium [13].

The size of radiological lesions does not appear to correlate with the severity of the initial neurological damage, nor is there any correlation between the evolution of radiological abnormalities and the persistence of symptoms. The appearance of these abnormalities is usually delayed, taking around 10 to 15 days, and an initially normal MRI scan does not rule out the diagnosis [14]. CSF studies are usually normal, but hyperproteinorachia may be present [15, 16]. The EEG, which is non-specific, usually shows a diffuse slowing of brain activity [15]. Data in the literature vary considerably in terms of mortality, ranging from 6% to 90% [3-17]. Treatment is essentially preventive, based on reasoned and progressive correction of all hyponatremia, without exceeding a correction rate of 0.5 mmol/l/h [18]. There are other recent correction formulas, in particular that of d'Adrogué and Madias, which is a considerable help as a therapeutic means in all situations of dysnatremia [19]. To date, no curative treatment has been codified.

CONCLUSION

The course of MCP and MEP is variable and often unfavourable. Treatment is mainly preventive, based on simultaneous and careful correction of severe hyponatremia and other contributing factors, including hypokalaemia. Current research is focusing on studying the efficacy of certain therapeutic options, in particular plasma exchange and intravenous immunoglobulins, which have been shown to be effective in certain isolated cases.

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