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CASE REPORT

Severe course of osmotic demyelination syndrome complicated by recurrent respiratory failure – Case Report

Kacper Janowski[1], Klaudia Michalak[1], Mateusz Wilkowski[2], Ireneusz Jurczak[3]

[1] *Department of Internal Medicine, Specialist Regional Hospital, Ciechanów, Poland*
[2] *Department of Neurology, Specialist Regional Hospital, Ciechanów, Poland*
[3] *Department of Diagnostic Imaging, Specialist Regional Hospital, Ciechanów, Poland*
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Abstract

Osmotic demyelination syndrome (ODS) is a non-inflammatory disorder of the central nervous system that arises from osmotic disturbances, particularly related to the rapid correction of chronic ionic imbalances, especially concerning sodium levels. ODS has been shown as an interdisciplinary problem which requires the collaboration of many specialists to achieve successful goals. This case study describes a 46-year-old male patient with a history of chronic alcohol abuse and other underlying health issues, including sodium imbalances. The patient required extensive care, spanning multiple departments – emergency, neurology, psychiatry, and detoxification, over a prolonged hospitalization of 80 days. The patient presented multiple symptoms, including recurrent respiratory disorders requiring intubation. Even with increasing improvements in management, osmotic demyelination syndrome still presents significant therapeutic challenges. Standardized treatment guidelines remain lacking.

Key words

central nervous system, hyponatraemia, sodium, osmosis, demyelination syndrome, osmotic demyelination syndrome

INTRODUCTION

Osmotic demyelination syndrome (ODS) is non-inflammatory demyelinating disorder of the central nervous system. It includes central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM), and particularly affects the white matter tracts of the pontine region. In clinical practice is often underdiagnosed and may lead to serious complications [1]. It was described by Adams in 1959 as central pontine myelinolysis, secondary to alcoholism and malnutrition [2]. In 1962, it was been shown the syndrome occurs outside the pons and referred to as extrapontine myelinolysis [3]. The most common form of the syndrome is central pontine myelinolysis, which can occur alone or in combination with extrapontine myelinolysis. ODS can present a variety of symptoms and neurological features – from asymptomatic to life-threatening, including death, and depends on the location of the demyelinating lesions in the central nervous system [4, 5].

Myelinolysis is most commonly iatrogenic and is often caused by overly rapid correction of serum sodium deficiency, primarily in cases of hyponatraemia, defined as a serum sodium level lower than 135 mEq/L, and is the most common electrolyte disorder which affects approximately 5% of adults and 35% of hospitalized patients. It is most often caused by water retention. Even mild hyponatraemia influences increased hospital stay and mortality [6]. In ODS, damage to the myelin sheath of brain cells occurs as a result of rapid plasma osmotic shifts, and has been reported to be commonly associated with serum sodium correction greater than 12 mmol/l in 24h [7, 8]. The significant risk factor for demyelination is intense osmotic stress on brain cells which can result in the loss of astrocytes and oligodendrocytes, trigger the activation of microglia, and facilitate the infiltration of macrophages that degrade myelin. Additionally, it disrupts the blood-brain barrier (BBB), ultimately leading to myelin damage [7]. The pathophysiology of osmotic demyelination syndrome is based on disturbances in the osmotic balance of the brain. In chronic hyponatraemia, intracellular concentrations of osmotically-active substances decrease as compensatory mechanisms protecting against oedema. Rapid correction of hyponatraemia and the subsequent increase in extracellular fluid osmolality lead to the movement of water out of cells, interstitial brain oedema, and ultimately demyelination, particularly affecting cells around the central canal of the pons and, to a lesser extent, extrapontine structures.

The exact mechanism of demyelination and the specific susceptibility of pontine cells remain unclear. Additional predisposing factors include chronic alcoholism, malnutrition, liver cirrhosis, and other water-electrolyte disturbances, such as hypokalaemia and hypophosphataemia. CDS cases were observed in association with diabetes mellitus, renal failure, haemodialysis, hyperemesis gravidarum, anorexia nervosa, Wilson disease, severe burns, systemic lupus erythematosus and liver transplant recipients [5, 9].

The clinical symptoms of demyelination syndrome are variable, from asymptomatic cases detected only in imaging studies to severe neurological deficits, coma or death. The syndrome often follows a biphasic course – the first phase

✉ Address for correspondence: Kacper Janowski, Department of Internal Medicine, Specialist Regional Hospital in Ciechanów, Powstanców Wielkopolskich 2, 06-400, Ciechanów, Poland
E-mail: Kacper.j90@gmail.com

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(associated with severe electrolyte disturbances) presents encephalopathy symptoms, including seizures and delirium. After electrolyte correction, a transient improvement is observed (days 1–7). However, approximately 2–3 days later, the second phase emerges, typically presenting with dysarthria, dysphagia, oculomotor disturbances, quadriplegia (initially flaccid, later spastic), behavioural disorders, psychosis, bulbar and pseudobulbar palsy, seizures and altered consciousness, including coma. Cases of locked-in syndrome have been described. In extrapontine myelinolysis, ataxia, parkinsonism, dystonia, choreoathetosis, and paroxysmal movement disorders triggered by motion are also common. The disease usually progresses rapidly, with death occurring within several days to weeks. Magnetic resonance imaging (MRI) in ODS is preferred for the diagnosis and subsequent follow-up of brain parenchymal abnormalities. MRI shows T2-weighted hyperintensity and fluid-attenuated inversion recovery (FLAIR) sequences and hypo-intensity in T1-weighted sequences in the central pons – usually symmetrically, with a characteristic trident-shaped, bat-winged or piglet appearance with sparing of the tegmentum and ventrolateral pons [9].

CASE REPORT

A 46-year-old male with a history of chronic alcohol abuse, hypertension, peptic ulcer disease, and oesophagitis was admitted to the emergency department due to worsening consciousness disturbances, developing delirium preceded by a generalized seizure. Laboratory tests revealed severe hyponatraemia (107 mmol/L; normal range 136–145 mmol/L), hypokalaemia (2.6 mmol/L; normal range 3.5–5.1 mmol/L), hypophosphataemia (1.7 mg/dL; normal range 2.5–4.5 mg/dL), elevated liver enzymes, thrombocytopenia, and mildly elevated inflammatory markers. A computed tomography (CT) scan of the brain showed mild cortical atrophy and small calcifications in the anterior part of the semi-oval centre of the left hemisphere of the brain. A few days before admission, the patient completed a prolonged period of heavy alcohol consumption and reported generalized weakness. Neurological examination revealed no meningeal signs, generalized muscle hypotonia and disorientation without other significant abnormalities. He was admitted to the Internal Medicine Department for electrolyte correction and further diagnostics. The patient received symptomatic treatment, including fluid therapy, sodium supplementation, and empirical antibiotic therapy. Due to persistent behavioural disturbances, diazepam and haloperidol were administered. Laboratory tests showed gradual electrolyte correction (sodium: 114 mmol/L on day 2, 131 mmol/L on day 3, and 135 mmol/L thereafter).

Despite the treatment, alcohol withdrawal delirium worsened, necessitating physical restraint. After psychiatric consultation, on day 4 the patient was transferred to a detoxification ward. However, progressive consciousness disturbances and respiratory insufficiency with desaturation episodes developed which required intubation and transfer to the Intensive Care Unit (ICU). Short-term mechanical ventilation improved his respiratory and general status. Follow-up CT scan showed no new findings compared to the previous CT scan. The patient was extubated on day 6 and later transferred to the Neurology Department for further management. Changes in serum sodium levels observed throughout the patient's hospital stay are shown in Figure 1.

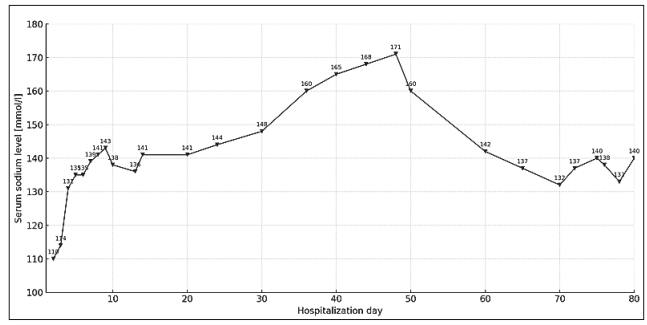

Figure 1. Serum sodium levels during hospitalization

On admission to the Neurology Department on day 11, the patient was haemodynamically stable but somnolent, with psychomotor slowing and spontaneous limb movements. Neurological examination revealed moderate dysarthria, global muscle hypotonia, diminished deep tendon reflexes, and moderate flaccid quadriparesis, rated 3/4 in all limbs on the Medical Research Council scale (MRC), without meningeal and pathological signs. Brain MRI showed symmetrically-distributed hyperintense areas in T2W1 imaging and hypointense areas in T1W1 imaging localized in the brainstem and in the cerebral hemispheres, especially in the basal ganglia (MR imaging suggested central pontine myelinolysis and extrapontine myelinolysis), small, symmetric T1W1 and T2W2 hyperintensities in the lentiform nuclei, suggesting prior microhemorrhages (Fig. 2, Fig. 3). After temporary improvement, the patient's condition deteriorated again, with worsening verbal-logic contact, consciousness, and paresis to flaccid quadriplegia. Laboratory tests showed severe hypernatraemia (171 mmol/L), transient renal impairment, and increased inflammatory markers. Staphylococcal pneumonia was diagnosed. Due to respiratory failure, the patient required re-intubation, mechanical ventilation, tracheostomy, and percutaneous endoscopic gastrostomy (PEG) placement due to deepen dysphagia. Gradual improvement was achieved thanks to intensive therapy, including targeted antibiotic therapy, fluid resuscitation and symptomatic treatment. Consciousness, respiratory function, laboratory test results, swallowing were normalized allowing removal of PEG and tracheostomy. The patient received speech and psychological therapy.

Over the following weeks, his verbal and cognitive abilities improved. By discharge on day 80, the patient presented mild memory deficits, mainly concerning the acute period of the disease; he retrieved full orientation (auto- and allopsychic) and improved mobility. The patient was able to move with the support of a physiotherapist, and took a few steps independently with the help of a railing. Neurological examination showed residual quadriparesis (MRC 4), increased muscle tension of mixed spastic-extrapyramidal character, hyperreflexia (deep reflexes) without lateralization, postural-kinetic tremor, lowering in speech volume, and mild dysarthria. He was referred for further neurological rehabilitation.

DISCUSSION AND CONCLUSIONS

The presented case illustrates the diagnostic and therapeutic challenges of osmotic demyelination syndrome, an interdisciplinary condition requiring collaboration between multiple medical specialties – physiotherapy, psychology

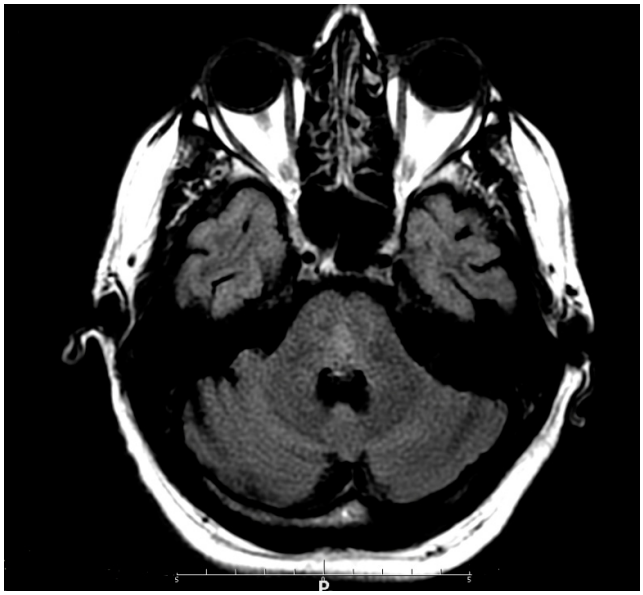

Figure 2. FLAIR MRI of the head – transverse section of the brain showing symmetrically distributed hyperintense areas located in the brainstem

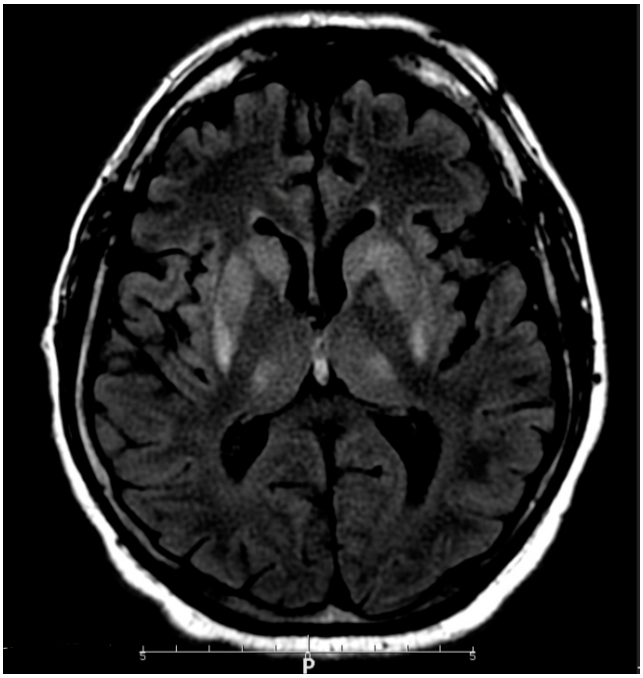

Figure 3. FLAIR MRI of the head – transverse section of the brain showing symmetrically distributed hyperintense areas located in the subcortical nuclei

and speech therapy [11]. The case involved both pontine and extrapontine myelinolysis, initially manifesting as encephalopathy with seizures and delirium, followed by fluctuating consciousness disturbances, orientation and memory deficits, dysarthria, dysphagia, episodic quadriplegia, and recurrent respiratory failure with electrolyte imbalances, necessitating intensive care, including mechanical ventilation. Despite the severe course and prolonged hospitalization of 80 days, significant functional recovery was achieved.

Due to the rarity of osmotic demyelination syndrome, standardized treatment guidelines remain lacking. Once the demyelination process has begun in the pons, there is no way to stop it or provide specific treatment. ODS is often underdiagnosed which can lead to disastrous consequences. Some reports suggest potential benefits of immunotherapy or vasopressin receptor antagonists (vaptans) in severe cases [10, 12]. However, management primarily focuses on supportive care and prevention, particularly cautious correction of electrolyte imbalances, especially in high-risk groups, such as malnourished or alcohol-abusing people with chronic hyponatraemia. Correction rate depends on general health status of the patient, comorbidities, and serum natremy level. The correction of 8–10 mmol/L per day (0.5 mmol/L per hour) is mostly considered as safe [13, 14]. The amount and rate of sodium compensation has been part of the debate for decades.

Since hypokalaemia may contribute to demyelination, potassium correction is recommended before sodium correction. The prognosis in ODS depends on comorbidities such as hypokalaemia, liver transplantation history, and initial neurological severity. Some improvement in extrapontine myelinolysis-associated movement disorders has been noted with dopaminergic therapy.

Despite increased awareness and improved management, osmotic demyelination syndrome remains a diagnostic and therapeutic challenge, not infrequently resulting in severe long-term disability, or even in death.

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