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Reversible Severe Parkinsonism Caused by Extra-Pontine Myelinolysis in a patient with Primary Adrenal Failure.

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Abstract

PURPOSE: Extrapontine myelinolysis (EPM) is a highly uncommon, life-threatening disease, particularly in individuals who initially appear with severe clinical symptoms. Here, we describe a case of EPM caused by the rapid correction of hyponatremia that had severe clinical signs at first but parkinsonism symptoms were fully improved after treatment.

CASE REPORT: A 46-year-old female patient was admitted to the hospital due to impaired consciousness. Her medical history reveals that she has PAI, or primary adrenal insufficiency. Initial laboratory measurements showed that the serum's sodium (Na) concentration was 104 mEq/L, chloride (Cl) content was 70 mmol/L, potassium (K) content was 4.95 mEq/L, glucose was 42 mg/dL, hydrogen potential (Ph) was 7.12, and bicarbonate (HCO_3) concentration was 10 mmol/l. The adrenocorticotropic hormone (ACTH) level was 21 mg/ml, while the cortisol level was 1.2ug/dl. Her mental state was unclear, she had sluggish hypophonic speech, generalized akinesia/rigidity in both upper and lower extremities, trouble swallowing solid and liquid meals, and sialorrhea were all discovered after the Na level was corrected. Hyperintense lesions were visible in the bilateral putamen and caudate nuclei of the Magnetic Resonance Imaging (MRI) T2 and flair-weighted scans, which indicate EPM. EPM was treated with corticosteroids and dopamine agonists, and she was eventually released after complete recovery.

CONCLUSION: Even if there are severe clinical symptoms at first, prompt diagnosis and treatment, such as dopaminergic, corticosteroid, and palliative therapy, can save a patient's life.

CASE REPORT

Abbreviations:

ODS	- Osmotic demyelination syndrome
EPM	- Extra-pontine myelinolysis
CPM	- central-pontine myelinosis
PAI	- primary adrenal insufficiency
Na	- sodium
Cl	- chloride
K	- potassium
Ph	- the potential of hydrogen
HCO_3	- bicarbonate
ACTH	- adrenocorticotropic hormone
BBB	- blood-brain barrier
Flair -MRI	- flair-weighted Magnetic Resonance Imaging
DWI- MRI	- diffusion-weighted imaging MRI

INTRODUCTION

Osmotic demyelination syndrome (ODS) is a fairly uncommon, life-threatening, non-inflammatory myelin loss disorder that causes parkinsonism (Lv *et al.* 2021). Two ideas attempt to explain how ODS develops. Complement molecules are influx into the brain in the quick correction of hypo-osmolality due to the degeneration of the blood-brain barrier (BBB). According to animal research, they produce ODS by having a toxic impact, mainly on myelin cells in the brain (Baker *et al.* 2000). Another theory is that degradation of the osmotic equilibrium damages oligodendrocytes and leads to ODS (Garg *et al.* 2019). On MRI, ODS can be observed as solely central-pontine myelinosis (CPM) (30%), only EPM (20%), or both (10-50%) (Uchino *et al.* 2003). Patients with ODS have symptoms that include encephalopathy, seizures, pseudobulbar paralysis, extrapyramidal symptoms, and dysphonia. Patients with encephalopathy are said to have a somewhat better prognosis than those with other symptoms (Lv *et al.* 2021). Because ODS can be caused by a multitude of variables, in evaluating the disease's progress, the most essential aspect is determining the component that causes this condition in the patient and providing prompt treatment (Lv *et al.* 2021). The most common reason for ODS is the rapid correction of hyponatremia (Akyol *et al.* 2007).

We report a female patient with EPM without CPM who exhibited generalized akinesia/rigidity after quick correction of hyponatremia owing to primary adrenal insufficiency and recovered completely with adequate therapy, despite severe clinical findings at the onset.

CASE REPORT

For the past three days, a 46-year-old female patient had been complaining of loss of consciousness, nausea, and vomiting in the emergency department. Laboratory analysis of our patient was summarized in Table 1. In her medical history, PAI (Addison´s disease) was discovered. The patient's history of adrenal antibodies is unknown. The patient was treated with hypertonic saline solution for low serum Na level and corticosteroid for PAI. The exact correction rate is unknown since hyponatremia is treated in the emergency department. After the replacement, the patient was hospitalized and treated in the endocrine department. On T2-weighted and flair-weighted MRI, hyperintense lesions at the bilateral putamen and caudate nuclei, and diffusion-weighted imaging MRI (DWI- MRI) investigation diffusion restrictions were seen in the same regions (Figure 1). The cerebrospinal fluid investigations were normal. After 3-4 days of the correction of hyponatremia, her neurological evaluation, she had slow hypophonic speech, widespread akinesia/rigidity in bilateral upper and lower limbs, problems swallowing solid and liquid meals, and sialorrhea. There was no history of any movement disorder, in her family. 15 days following the first MRI, a control MRI was performed.

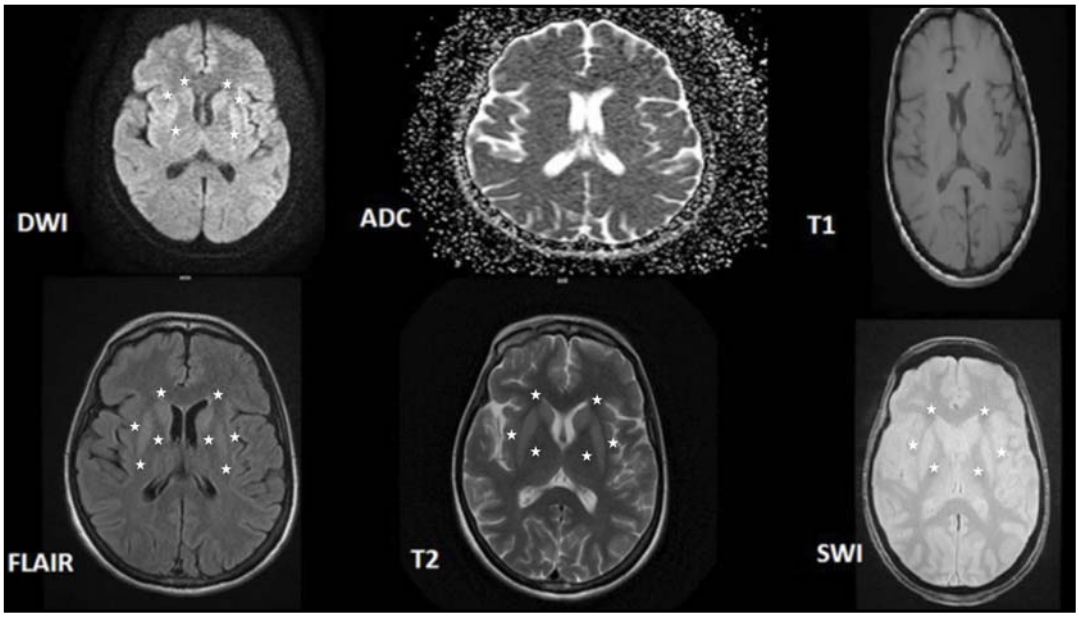

Fig. 1. On T2, Flair-weighted Magnetic Resonance Imaging (T2-Flair MRI), hyperintense lesions were seen in the bilateral putamen and caudate nuclei, indicating symmetric extra-pontine osmotic demyelination. Restriction of diffusion was also seen on diffusion-weighted imaging MRI (DWI- MRI) in the same areas. T1-weighted imaging MRI was also normal.

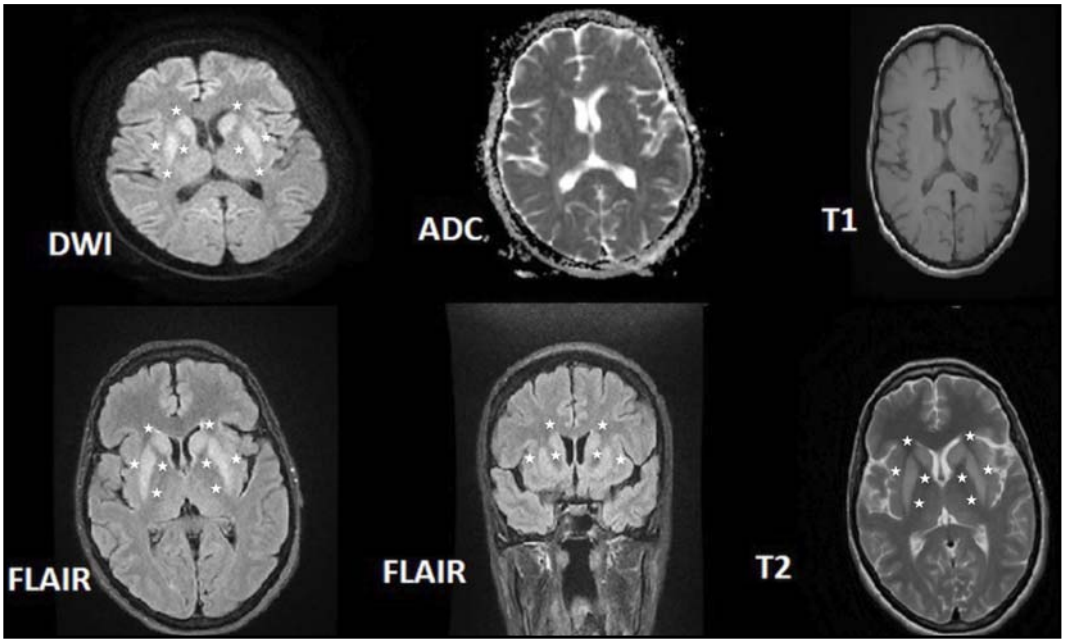

Fig. 2. After the initial Magnetic Resonance Imaging of approximately 15 days, On T2, Flair-weighted Magnetic Resonance Imaging (T2-Flair MRI), hyperintense lesions were seen in the bilateral putamen and caudate nuclei, indicating symmetric extra-pontine osmotic demyelination. Restriction of diffusion was also seen on diffusion-weighted imaging MRI (DWI- MRI) in the same areas. T1-weighted imaging MRI was also normal.

Tab. 1. Laboratory analysis of the patient

References	Na mEq/L (74-106)	Cl mmol/L (98-107)	K mEq/L (3,5-5,1)	Glucose mg/dL (74-106)	Ph (3-11,5)	HCO3 mmol/l (22-29)	Cortisol ug/dl (6,7-22,6)	ACTH mg/ml (7,2-63,3)
Patient's results	104 mEq/L	70 mmol/L	4.95 mEq/L	42 mg/dL	7.12	10 mmol/l	1.2 ug/dl	21 mg/ml

Hyperintensity was seen in the bilateral globus pallidus and caudate nucleus on T2-FLAIR weighted imaging, and diffusion restriction was on DWI-MRI (Figure 2). The patient was diagnosed with EPM and started 125 MG Madopar (Levodopa and benserazide hydrochloride) five times a day. Corticosteroid treatment was continued at 20mg/day. Due to swallowing difficulties, the feeding was continued via the nasogastric tube. In around three months, the dopaminergic therapy was decreased and stopped. The patient made a full recovery.

DISCUSSION

ODS is a relatively uncommon condition characterized by CPM and EPM (Uchino *et al.* 2003). Although the major reason for ODS is the rapid correction of hyponatremia, it has also been documented to occur in normal levels of Na, in hypernatremia, hypokalemia, and hyperglycemia (Akyol *et al.* 2007). Diseases presenting with hyponatremia should be examined first when considering the development of ODS. Among the causes of hyponatremia diuretic-associated hyponatremia, adrenal and pituitary insufficiency, inadequate antidiuresis syndrome, cerebral salt wasting, and exercise-related hyponatremia should be considered. The authors also take into account the pace at which hypertonic saline solution is administered in the hyponatremia treatment (raise 1–2 mmol/l/h until symptoms disappear, then lower to 8–12 mmol/l/day) (Lv *et al.* 2021). Other variables that encourage the establishment of ODS should be questioned in the absence of hyponatremia. Metabolic disturbances, liver disease (including those undergoing orthotopic liver transplantation), cancer, pregnancy/postpartum condition, malnutrition, the existence of severe illness/sepsis, alcoholism, and adrenal insufficiency syndrome are all risk factors for ODS in patients with and without chronic hyponatremia (Akyol *et al.* 2007). In patients with PM plus EPM, Garg *et al.* (Garg *et al.* 2019). reported that the presence of extrapontine diffusion signal abnormalities before pontine signal changes might be the first sign of ODS. Of course, this theory does not fully account for the development of only pontine myelinosis. Initially, in our case, restriction of diffusion was observed at the extra-pontin area but any signal changes were not seen at the pontin area later. The occurrence of symptoms other than encephalopathy in cases of ODS may be a hint of poor prognosis (Lv *et al.* 2021). Despite bilateral symmetrical diffuse akinetic/rigidity and a clinical picture that was too severe allowing oral dosing of drugs in our case, the patient recovered entirely with early and proper therapy, which is dopaminergic, corticosteroid, symptomatic, and supportive. Early diagnosis and treatment are far more significant in predicting prognosis than the presence of probable clinical signs.

CONCLUSION

Although EPM without CPM is exceedingly unusual, early detection and treatment can result in complete recovery, even in cases where the clinical indications are severe at the onset.

Written informed consent was obtained from the patient

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None.

Declaration of interest

None.

Financial source

None.

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