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A Rare Case of Acute Psychosis as an Isolated Manifestation of Extrapontine Myelinolysis

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Authors' contributions

This work was carried out in collaboration between all authors. Author WAWS conceived the work and wrote the first draft of the manuscript. All authors equally participated in the preparation of the manuscript, read and approved the final manuscript.

Case Study

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ABSTRACT

Aims and Background: Central pontine myelinolysis (CPM) and extrapontine myelinolysis (EPM) are recognized as osmotic demyelination syndrome (ODS). ODS is pathologically characterized by non-inflammatory demyelination of several brain structures with sparing of axons. This condition is usually associated with overzealous correction of hyponatraemia. Acute psychosis as the sole clinical manifestation is extremely rare.
Presentation of Case: Hence, we report an interesting case of a middle-aged man who was diagnosed with EPM, following rapid correction of hyponatraemia and subsequently developed acute psychosis. He made a good recovery with supportive treatment alone.
Discussion and Conclusion: The possibility of psychosis as a manifestation of ODS, particularly in patients with recent correction of hyponatraemia. The rate of correction of plasma sodium level is the key point for preventing ODS and its complications.

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Keywords: Osmotic demyelination syndrome; extrapontine myelinolysis; hyponatraemia; psychosis.

1. INTRODUCTION

Extrapontine myelinolysis (EPM) is an uncommon type of osmotic demyelination syndrome. The demyelinating lesions in EPM are found to involve extra-pontine structures, and may co-exist with central pontine myelinolysis (CPM). However, studies show that the numbers of cases of isolated extrapontine lesions were almost similar to cases of both CPM and EPM [1].

Osmotic demyelination syndrome (ODS) was first described in alcoholic and malnourished patients [2]. A decade later, a correlation between abrupt correction of hyponatraemia and osmotic demyelination was observed [3]. Currently, rapid correction of hyponatraemia is the second most frequent cause of CPM, representing 21.5% of cases. Chronic alcoholism comes as the commonest, with 39% of reported cases whereas the other causes include pulmonary infections, malignant tumour and disease of central nervous system [4].

2. PRESENTATION OF CASE

A 58-year-old man was brought to the emergency department due to sudden behavioral change with hallucination. The patient believed that he was God's messenger and could communicate with angels. The patient had the suspicion that his family wanted to harm him and he was irritable and aggressive He had no previous history of psychiatric illness, alcohol or substance abuse. His premorbid personality was an easy-going, sociable and religious person. The patient had medical comorbidities of hypothyroidism, hypertension and type 2 diabetes mellitus, and was treated with levothyroxine, felodipine and metformin, respectively. The patient also had a prior history of hypoadrenalism 6 years ago but had not been on replacement therapy since 3 years ago.

Mental state examination revealed symptoms of grandiosity, persecutory delusions, visual and auditory hallucinations. However, the patient was fully conscious and orientated with clear sensorium. There were no neurological deficits, pyramidal or extra pyramidal signs. The routine blood investigations, such as electrolytes and sepsis parameters were within the normal range.

The patient was recently discharged from the medical ward a week earlier. The patient initially presented with episodes of vomiting and dehydration and subsequently developed generalized tonic-clonic seizures. Blood investigations revealed severe hyponatraemia (100mmol/L), hypokalaemia (2.6mmol/L) and normal thyroid function. Lumbar puncture for CSF examination was not performed, as the patient and his family declined consent.

The patient was treated for acute adrenal crisis and severe symptomatic hyponatraemia, precipitated by acute gastroenteritis, and was given intravenous infusion of 3% hypertonic saline, intravenous potassium chloride and hydrocortisone 100mg TDS. A careful and gradual sodium infusion was initiated, with frequent electrolyte monitoring. The sodium correction rate aimed at 8mmol/l over 24 hours. The hyponatraemia improved to 125mmol/l over 3 days, with marked clinical recovery and cessation of seizures. Over the next week, the patient was noted to be mildly confused. However, neither psychosis nor neurological deficits were present during this initial period. Intravenous hydrocortisone was then tapered to oral hydrocortisone 40mg and later to 20mg twice a day upon discharge.

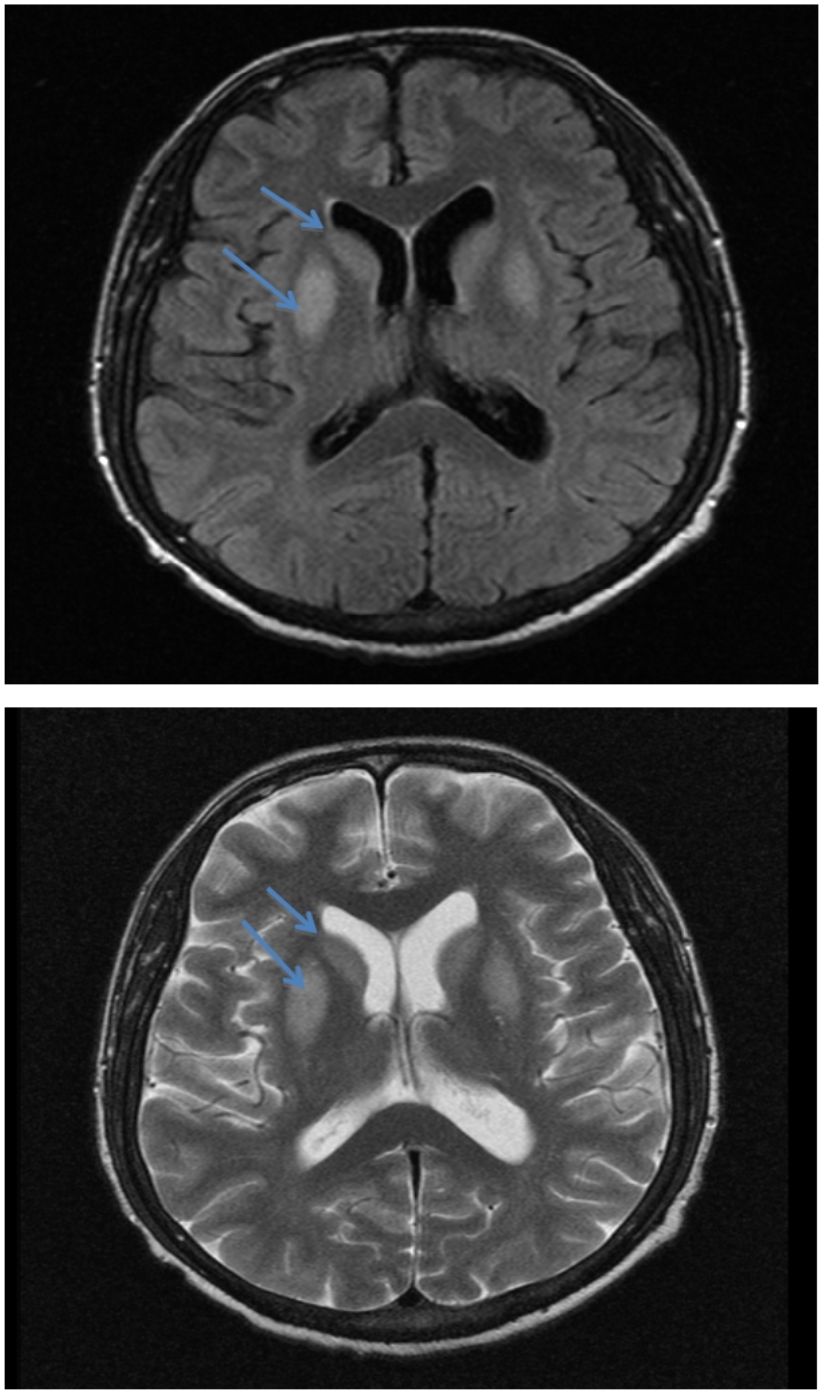

Fig. 1. Bilateral basal ganglia (caudate and putamen) hyperintensities on FLAIR and T2W1 sequences indicative of extrapontine myelinolysis

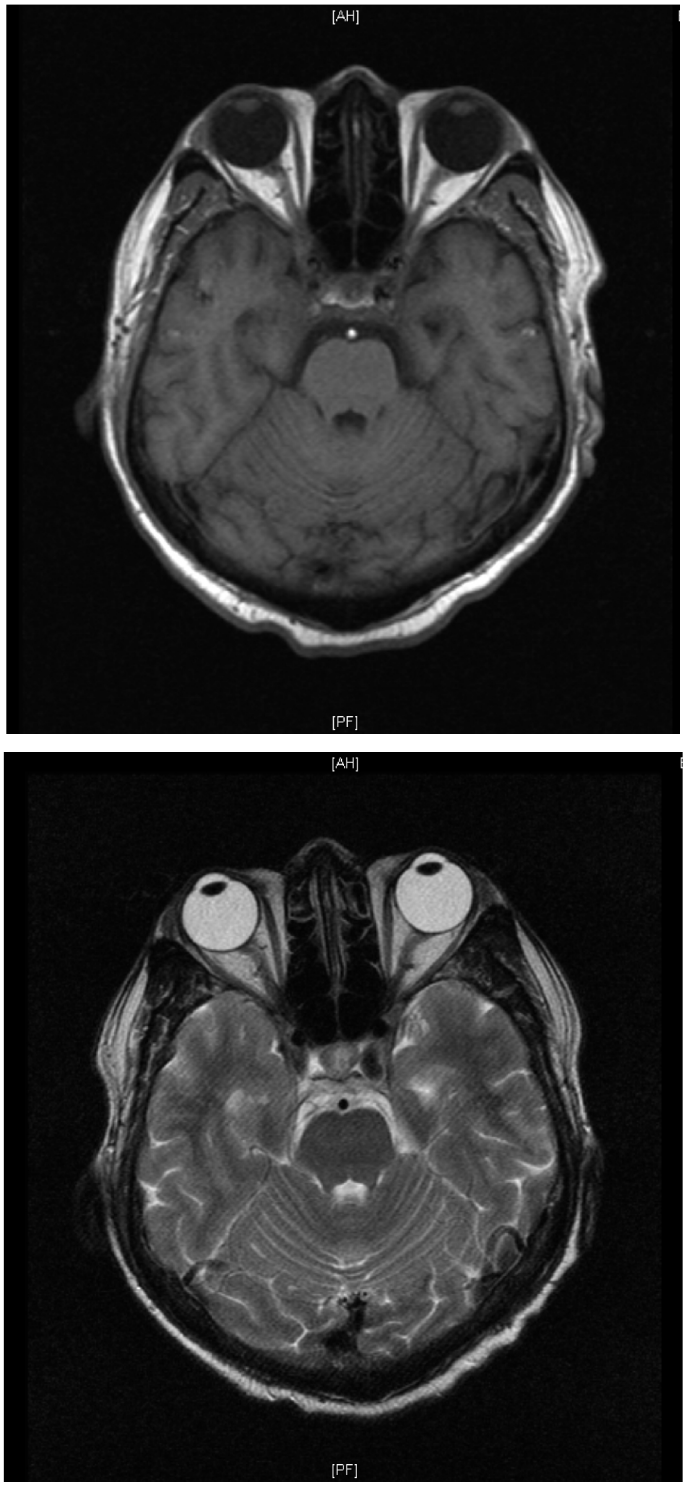

Fig. 2. No focal lesion or abnormal signal intensity noted in the pons on FLAIR and T2W1 sequences

A magnetic resonance imaging (MRI) brain scan found high-intensity lesions with bilateral involvement of basal ganglia affecting caudate and putamen on T2 weighted and FLAIR images (Fig. 1). The pons showed no sign of demyelination, thus the diagnosis of isolated extra pontine myelinolysis was made (Fig. 2). During the second admission, the patient was started on risperidone tablets 1 mg twice a day for his psychosis and sodium valproate tablets 200 mg twice a day as a mood stabilizer. He was discharged home with improvement of his psychotic symptoms. After 3 months the patient was completely recovered from his psychotic symptoms.

3. DISCUSSION

The sequelae of rapid correction of hyponatraemia usually manifest only several days after rapid sodium correction. The clinical deterioration is usually due to the development of ODS, that consist of CPM with or without EPM, or isolated EPM [5]. This case classically portrayed the stereotypical biphasic clinical course of ODS, with the initial clinical improvement of the patient after hyponatraemia correction, followed by the sudden deterioration in his mental state [6]. However, there was no associated neurological deficit. His acute pychosis was the only clinical manifestation of ODS, a rare complication, and to the best of our knowledge, this is the only reported case of psychosis in isolated EPM. Another recent case of acute psychosis with central pontine/extrapontine (CPEM) reported in India also had a similar favorable outcome [7]. There were 6 cases found in the literature linking isolated EPM with adrenal insufficiency, most likely due to rapid correction of hyponatraemia [8].

Clinical symptoms of CPM usually manifest as neurological deficits such as acute progressive quadriplegia, dysarthria, dysphagia and loss of consciousness. However, clinical presentations in isolated EPM are different compared to CPM. Since pathological lesions in EPM are extrapontine, the reported manifestations are more towards behavioral and neuropsychiatric changes, such as parkinsonism, mutism, dystonia or catatonia [9]. The best radiological modality to diagnose EPM is MRI brain. Hyperintense lesions on T2-weighted and FLAIR sequences are the characteristic features, which commonly occurs in the basal ganglia, mainly the caudate and putamen, with a typical sparing of the pallidum [10]. Other sites include the involvement of cerebellum, subcortical white matter and thalamus [3]. However, the findings should be correlated on clinical grounds as these radiological features alone are not specific for ODS.

In this case, we reported a rare occurrence of acute psychosis as the only manifestation of isolated EPM. The diagnosis of EPM was established prior to the psychotic episodes by radiological imaging. Thus, the psychotic symptoms that appeared a week later were attributed to organic psychosis secondary to EPM. Corticosteroid-induced psychosis was less likely since the stress dose of hydrocortisone was already tapered to physiological doses about a week before the onset of psychosis [11]. Moreover, the MRI findings were in favor of the diagnosis of EPM. Acute delirium was also unlikely due to the patient's preserved sensorium with normal electrolytes and septic parameters.

The most probable explanation for the acute psychosis in this patient was the demyelination lesions found exclusively in the bilateral caudate and putamen. There was a similar case reported, describing schizophrenia-like symptoms in CPEM with presence of lesions in the caudate and putamen on the MRI findings [12]. Abnormalities of the basal ganglia or muscarinic cholinergic system in the caudate and putamen have been associated with schizophrenia, based on a two different studies, which could further explain the psychotic symptoms in CPEM [13,14].

There have been no studies done on any specific treatments and at present, supportive treatments such as atypical antipsychotics are the mainstay of therapy [9]. This makes prevention to be the most important measure. A wide range of guidelines on gradual correction of hyponatraemia to avoid CPEM, have been suggested. The rate of correction should always be guided by the nature of chronicity and neurological manifestation of the patient.

Recently, expert panels have published a consensus on hyponatraemia correction [6]. Current recommendation for correction of acute symptomatic hyponatraemia is urgent correction by 4-6mmol/L. For severe symptoms, this can be achieved with a regime of 100ml bolus of 3% saline infused over 10 minutes that may be repeated up to 3 times if required. For mild to moderate symptoms, infusion of hypertonic 3% saline can be given at a rate of 0.5-2ml/kg/hour. If the chronicity of hyponatramia is uncertain, correction must be based on chronic hyponatraemia guidelines. For asymptomatic and chronic hyponatraemia, there are various recommendations. However, in the recent panel expert review, the suggested rate of correction must be based on the risk of ODS. Patients at risk of ODS include severe hyponatraemia with serum sodium <120mmol/L for more than 48-hour duration. For these high-risk patients, correction rate is lower at 4-6mmol/L per day and should not exceed 8mmol/L per day. For normal risk patients, the correction rate is higher, 4-8mmol/L per day, and maximum correction should not be more than 10-12mmol/L over 48 hours [6].

Even with cautious hyponatremia correction according to guidelines, patients are still susceptible to develop ODS. The exact aetiopathogenic mechanism on how correction of hyponatraemia leads to ODS despite careful correction is still unknown. Two significant predisposing factors seem to be the chronic nature of the hyponatraemia and the acute nature of its improvement [4]. Patients with cortisol deficiency, particularly, were found to have high risk of developing EPM [8,15]. Other risk factors include hypokalemia, hypoxia, burn injury, malnutrition, and chronic alcoholism [16].

4. CONCLUSION

This case highlights the possibility of psychosis as a manifestation of ODS, particularly in patient with recent correction of hyponatraemia. A careful correction of hyponatraemia is the most important measure in order to prevent ODS and its complications.

CONSENT

All authors declare that ‘written informed consent was obtained from the patient (or other approved parties) for publication of this case report and accompanying images.

ETHICAL APPROVAL

Not applicable.

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COMPETING INTERESTS

Authors have declared that no competing interests exist.

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