

# Isolated Extrapontine Myelinolysis Presenting as Acute-onset Reversible Parkinsonism in a Boy with Adrenocortical Insufficiency

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## Abstract

Isolated extrapontine myelinolysis is an uncommon form of osmotic demyelination syndrome (ODS) in paediatric age group. We describe a child with primary adrenocortical insufficiency manifesting as cyclical vomiting syndrome, which in turn has complicated in the form of extrapontine myelinolysis manifesting as reversible acute parkinsonism.

**Keywords:** Acute Parkinsonism; Extra pontine myelinolysis; Osmotic demyelination; Cyclical vomiting; Adrenocortical insufficiency

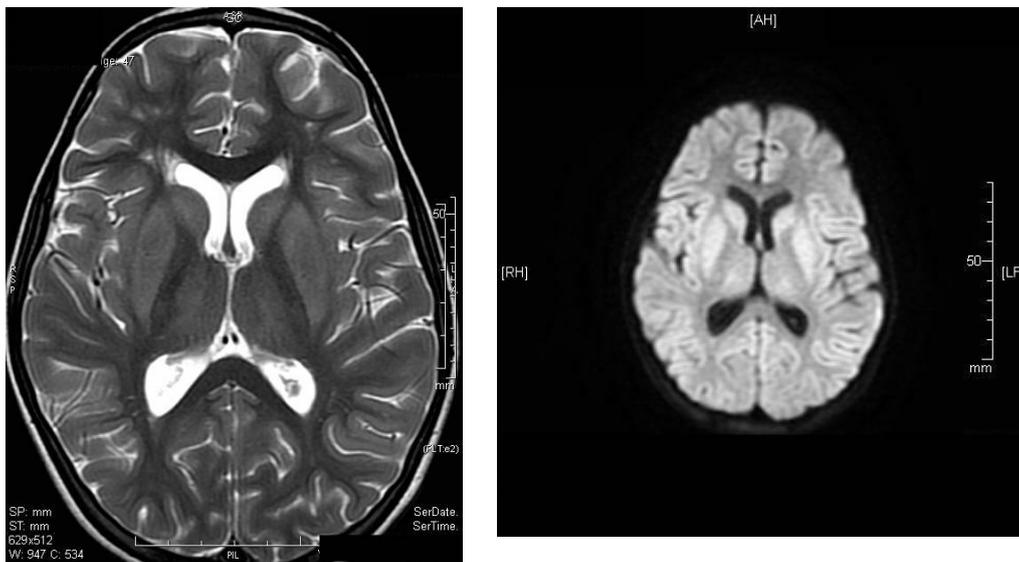
## Introduction

Osmotic demyelination syndrome involving pons and extrapontine structures is an uncommon neurological emergency occurring in the setting of rapid correction of hyponatremia. There are very few paediatric reports of isolated extrapontine myelinolysis presenting as acute-onset parkinsonism with complete neurological recovery.

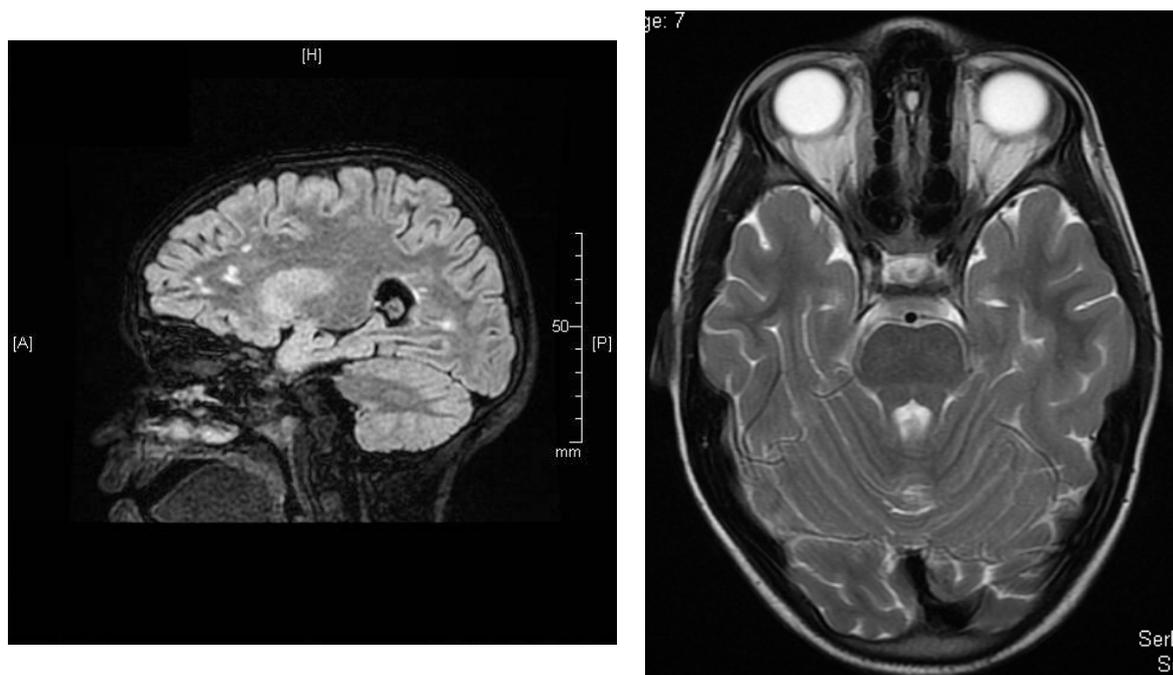
## Case Report

A 9 year 3 months-old, previously healthy boy presented with episodic vomiting for the preceding four months. He had three previous episodes at intervals of 3-4 weeks, with non-projectile, non-bilious vomiting lasting 4-5 days and improved with symptomatic treatment (intravenous fluids and anti-emetics). In between episodes, he was healthy. He was brought to our centre with 9-10 vomiting's the previous day. He was lethargic, had feeble peripheral pulses and hypotension. His investigations revealed: serum sodium 110 mmol/L, potassium 4 mmol/L, chloride 83 mmol/L, calcium 6.7 mg/dL, magnesium 1.8 mmol/L, serum osmolarity 213, urine osmolarity 263, urine sodium 68 mEq/L; arterial blood gas: pH 7.30, bicarbonate 12 mmol/l, and base deficit 18 mmol/l suggesting severe hyponatremia and hypochloremic metabolic acidosis. His renal and liver functions and ammonia were normal. In view of severe hyponatremia, serum cortisol was obtained which was 4.5 mcg/dl (normal 5-25). He was treated with intravenous fluids, inotropes and antibiotics. His serum sodium was gradually corrected over 72 hours (118 by 24 hours, 126 by 48 hours and 135 by 72 hours). By day 3 his sensorium also improved; he was able to sit on his own, started speaking and started oral intake. The CT scan of brain, (day-2 of admission), was normal. There was family history of migraine without aura in mother. In view of episodic cyclical vomiting and other investigations being normal, a provisional diagnosis of cyclical vomiting syndrome was made and discharged after seven days on Coenzyme Q, Pantoprazole, Domperidone and Amitriptyline. Five days later, he was brought back in non-ambulatory state, with complaints of tremulousness of face and both hands (left more than right) exacerbated by emotions and on attempt to reach for objects. He had intermittent twitching of left eye lasting for 10 seconds to 2 minutes along with slurred, unclear speech (hypokinetic speech). Two hours prior to admission, he had abnormal, clonic movements of left hand and persisting staring to left lasting for 2 hours. The movements subsided after giving intravenous Lorazepam and intravenous Levetiracetam. His vitals, general physical examination, respiratory and cardiac functions were unremarkable. He was conscious, oriented to time, place and person, with main mode of communication being gestures especially 'yes' and 'no' using head nod and eye blinks. He was able to follow simple commands ('show me your tongue', 'lift your hand up'). He was emotionally labile with intermittent crying and occasional laughing without any reason. He had bradykinesia, with minimal facial expressions and minimal spontaneous movements of limbs. Even at rest, he had tremors involving face and hands. All the limbs were rigid with preserved antigravity movements and deep tendon reflexes. His investigations revealed serum sodium of 125

mmol/l, potassium 4.9 mmol/l, chloride 96 mmol/l, serum cortisol 2.27 mcg/dl (normal 5-25 mcg/dl), normal complete blood counts, liver and renal function tests. The CSF protein was 20 mg/dl, sugar 59 mg/dl (blood sugar 90 mg/dl), 5 lymphocytes and lactate 1.5 mmol/L (arterial lactate 1.1 mmol/l). The plasma amino acids, ammonia and urine organic acids were normal. The electroencephalogram (performed after IV Lorazepam and Levetiracetam) was unremarkable. The MRI of brain revealed T2W and FLAIR hyperintense signal changes with restricted diffusion involving bilateral caudate nuclei and putamina with sparing of globi pallidi (Figure 1A, B) and patchy signal changes involving frontal white matter with no restricted diffusion (Figure 2A). The posterior fossa structures including pons and cerebellum were unremarkable (Figure 2B). Based on these, he was diagnosed as a case of primary adrenocortical insufficiency presenting with cyclical vomiting, and acute parkinsonism possibly due to extra pontine myelinolysis. He was treated with hydrocortisone (initially intravenous and later oral), levetiracetam, trihexyphenidyl, clonazepam, and propranolol. But, dramatic improvement in his parkinsonian symptoms was noted after addition of carbidopa and levodopa: 1 mg/kg/day of levodopa initially, later increasing up to 3 mg/kg/day. By day 7, he started walking independently with mild unsteadiness of gait, started taking orally and able to speak in clear sentences. His facial expressions were normal and there was no emotional lability. By 3 weeks, he was neurologically normal. The Levodopa and Levetiracetam were completely stopped after 4 weeks. A final diagnosis of primary adrenocortical insufficiency presenting as cyclical vomiting with resultant hyponatremia was made.



**Figure 1:** T2-weighted axial MRI showing subtle hyperintense signal changes involving bilateral caudate nuclei and putamina with sparing of globi pallidii (white arrows in A). Axial diffusion weighted image shows restricted diffusion involving bilateral caudate nuclei and putamina (black arrows in B)



**Figure 2:** Sagittal T2-weighted MRI showing hyperintensities involving frontal white matter (white arrows in A). Axial T2-weighted image showing unremarkable pons and cerebellum (B)

## Discussion

Osmotic demyelination is a neurological emergency due to rapid-onset demyelination involving pons (central pontine myelinolysis) and/or extrapontine structures (extrapontine myelinolysis). Initially described as a distinct entity in alcoholics and malnourished persons, it is commonly seen following rapid correction of hyponatremia [2]. The syndrome of osmotic demyelination typically has a biphasic course with initial hyponatremic seizures and encephalopathy which resolves with sodium correction, and reappearance of characteristic neurological symptoms and signs such as dysarthria, dysphagia, flaccid quadriparesis, encephalopathy and, oculomotor palsies due to pontine involvement [3]. Although pathological studies reported isolated extrapontine involvement in up to two-fifth cases, symptomatic cases are much less especially in children [4,5]. The manifestations include extrapyramidal symptoms like parkinsonism, dystonias, myoclonic jerks and catatonia [4,6]. The reversible parkinsonism has been reported in very few cases [5,7,8]. These are attributed to decreased Dopamine receptor binding in striatal neurons as reflected by elevated CSF homovanillic acid and rapid recovery following L-dopa administration [5,9]. With high index of suspicion and early identification, prognosis is good, especially if underlying cause is identified. A recent review has delineated two types of evolution and their varied response to treatment [10]. The acute akinetic-rigid syndrome said to have better response to dopaminergic therapy, while delayed presentation often refractory to medical treatment. In our case, primary adrenal insufficiency and malnutrition have contributed to extrapontine myelinolysis despite slow correction of hyponatremia, but recovered promptly with hydrocortisone supplementation and L-dopa. The usual period of recovery is around 3-6 weeks, by which time both clinical and radiological resolution can be expected in majority of cases. The levodopa supplementation should be continued till satisfactory symptomatic improvement is observed.

Many metabolic and endocrinological disorders (Addison disease, organic acidemias, urea cycle defects, fatty acid oxidation defects and mitochondrial cytopathies) can present as cyclical vomiting in children [1]. In our case, adrenocortical insufficiency manifested as cyclical vomiting and complicated as osmotic demyelination in the form of reversible parkinsonism. Hence, idiopathic cyclical vomiting syndrome should only be diagnosed after exclusion of all these disorders, apart from common gastrointestinal disorders.

This young boy with recent-onset cyclical vomiting had hyponatremia during first admission with new onset neurological symptoms five days after discharge. He had features suggestive of acute onset parkinsonism in the form of bradykinesia, rigidity and resting tremors with preserved consciousness. This suggests predominant involvement of basal ganglia especially corpus striatum (caudate nuclei and putamina). The causes for new-onset extrapyramidal syndrome in this setting could be osmotic demyelination with extrapontine myelinolysis or mitochondrial cytopathy associated with cyclical vomiting syndrome. Other differentials to be considered are: Addison's disease, organic acidemias like methylmalonic acidemia and propionic acidemia, urea cycle defects (known associations with cyclical vomiting) and drug-induced dyskinesias (domperidone). MRI of brain, arterial and cerebrospinal fluid (CSF) lactate, plasma ammonia, plasma amino acids, urine organic acids, serum cortisol and serum electrolytes will be helpful for further narrowing the differentials.

## Author contributions

RK and LL managed the case, did the work up, and drafted initial manuscript; RVD gave radiological inputs and critically reviewed the manuscript.

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