

Osmotic Demyelination Syndrome: A Clinical Disguise in a Patient with Head Injury and Alcohol

This article was published in the following Scient Open Access Journal:

SOA: Clinical Medical Cases, Reports & Reviews

Received August 07, 2017; Accepted August 28, 2017; Published September 04, 2017

Kumarasinghe N.R.^{1*} and Tennakoon A.B.²

¹Registrar in General Surgery, Pilgrim Hospital, Boston, United Lincolnshire NHS Trust, Lincolnshire, UK, England

²Consultant General Surgeon, Pilgrim Hospital, Boston, United Lincolnshire NHS Trust, Lincolnshire, UK, England

Introduction

A male fifty nine years of age was admitted to the emergency department with seizures and drowsiness. Clinical and biochemical evaluation revealed he was dehydrated with a low serum Sodium (Na) concentration of 113 mmol/l (normal range 135-146 mmol/l). He was known to have chronic alcohol related liver disease. The dehydration and hyponatremia which were thought to be the causative factors of the seizure was rapidly corrected and stabilised. A computed tomography (CT) scan of brain showed a 1millimetre thick subacute subdural hematoma (SDH) in left frontal region of the brain with no pressure symptoms. This was managed conservatively. His condition improved and was discharged from hospital on the third day.

Twelve days later he was readmitted with drowsiness and altered behaviour. Information gathered from his family members revealed he was drowsy, disorientated and not his normal self since coming home from hospital and had progressively worsened.

During the second admission he again developed generalised seizures and had a fluctuating Glasgow coma scale of between 10-14. A muscle power of grade three with generalised muscle hypotonia was evident on examination. The seizures were controlled with intravenous phenytoin followed by levetiracetam. Initial attention was given to the SDH as the causative factor. However repeated CT scans of brain failed to reveal an expanding haematoma, compressive effects or cerebral oedema. There were no derangement of blood investigations as before. However he did not improve despite anticonvulsive treatment, hydration, nutrition and treatment for alcohol withdrawal. Further clinical examination failed to identify focal neurological weakness, features of stroke or evidence of liver or renal failure. Hence, an Magnetic resonance imaging (MRI) of the brain was performed. The MRI finding was quite surprising. It showed of hyperintense areas in the pontine region on T2 images suggestive of demyelination. Hence was diagnosed of an entity known as central pontine myelinolysis.

The patient had a prolonged course of recovery. The principals of care were rehabilitation and supportive care based on providing nutrition (enteral nasogastric feeding and parenteral), water and electrolyte balance, anticonvulsive medication, thiamine infusions and general nursing care of an immobilised patient. Eventually he achieved an acceptable level of recovery and of being independent. On discharge from hospital he was prescribed long term prophylactic levitiracetam therapy.

Discussion

Osmotic demyelination syndrome belongs to an entity known as pontine myelinolysis. Of the numerous organic causes for seizures, pontine myelinolysis is one of the rarest. Data suggests it to be about 0.05% of all general hospital admissions and 0.4-0.5% admitted to neurological services [1,2]. Ambiguous nature of clinical presentations, low threshold of clinical suspicion and reporting, delayed appearance of radiological changes in imaging and need for advanced imaging modalities such as MRI and been an autopsy diagnosis may contribute for the reported incidence.

Pontine myelinolysis is characterised by loss of myelin sheaths in the pontine area. Been an autopsy based histological diagnosis it was first brought to attention by Adams et al, in 1959. He described four fatalities from neurological dysfunction whose brain autopsy showed demyelination in the pontine region of the brain stem. However he

*Corresponding author: Kumarasinghe N.R., Registrar in General Surgery, Pilgrim Hospital, Boston, United Lincolnshire NHS Trust, Lincolnshire, England, UK, Tel: 07849917510, Email: navam_k2000@yahoo.com

did not use the term 'demyelination' in order to differentiate it from other demyelination disorders [3]. It is now termed osmotic demyelination syndrome when it occurs in the presence of hyponatremia.

Rapid correction of chronic hyponatremia is thought to be the leading cause [4]. However conditions that predispose to hyponatremia such as primary polydipsia [5], peritoneal dialysis and causes unrelated to sodium correction such as liver and stem cell transplant [6,7] have been reported associated with pontine myelinolysis. Chronic alcohol use, severe malnutrition, type I and gestational diabetes, coeliac disease and Wilson's disease, Addisons disease and electrolyte imbalances like hypophosphatemia and hypernatremia have been reported as rare causes for pontine myelinolysis [8].

Interestingly in addition to rapid correction of hyponatremia in this patient, he was a chronic alcohol consumer with an inadequate nutritional intake. A combination of all these factors would have contributed to this condition.

Intracellular osmotic stress is linked to the pathogenesis of myelinolysis. Rapid correction of chronic hyponatremia leads to intracellular dehydration of brain cells. The resulting osmotic stresses cause leakage in the blood-brain barrier and results in release of inflammatory mediators. This in turn cause damage to oligodendrocytes which myelinate neurons in the central nervous system [9,10].

Classical clinical features in pontine myelinolysis are progressive quadriparesis and pseudobulbar palsy [11]. However a range of neuro psychiatric signs have been described in literature including seizures and features mimicking alcohol withdrawal. Retrospective analysis of clinical presentation demonstrate that this patient had an acute illness from which he temporally recovered, followed by a longer more debilitating period. Literature describes a characteristic two phase clinical presentation in pontine myelinolysis. The acute phase is related to the electrolyte imbalances. The second phase is associated with demyelination of pontine neurons [12,13].

Pontine myelinolysis is seldom suspected. This is due to its low incidence, diverse causative factors and ambiguous nature of clinical presentations. This is further confounded by the radiological challenges in diagnosis (Figure 1).

Magnetic resonance imaging (MRI) appears to be the choice of imaging in pontine myelinolysis.

The earliest feature seen within twenty four hours of the onset of quadriplegia is hyperintense regions in diffusion weighted imaging [14]. It is better seen as hyperintense areas in T2 images, hypointense areas in T1 images and FLAIR-hyperintensive [15]. Yet, these changes take up to 2 weeks to develop. This correlates with the timing of the MRI in this case presentation (Figure 3).

The CT brains commonly performed in patients with neurological deficits and head injury fail to show any notable features and can often can direct attention to a minor pathology. This was evident in this case history with initial attention focused on the very small SDH with no pressure effects (Figure 2).

Early reports reveal a poor prognosis from pontine myelinolysis with a high mortality and long term debility in survivors. Yet, this trend seems to be improving likely due to

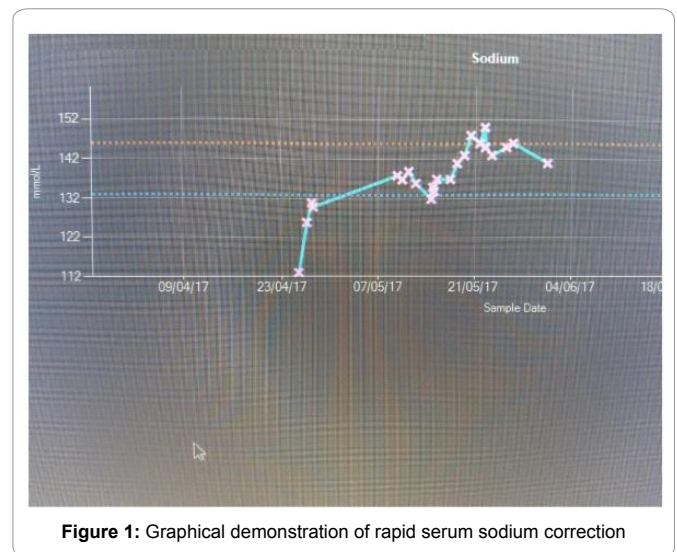


Figure 1: Graphical demonstration of rapid serum sodium correction

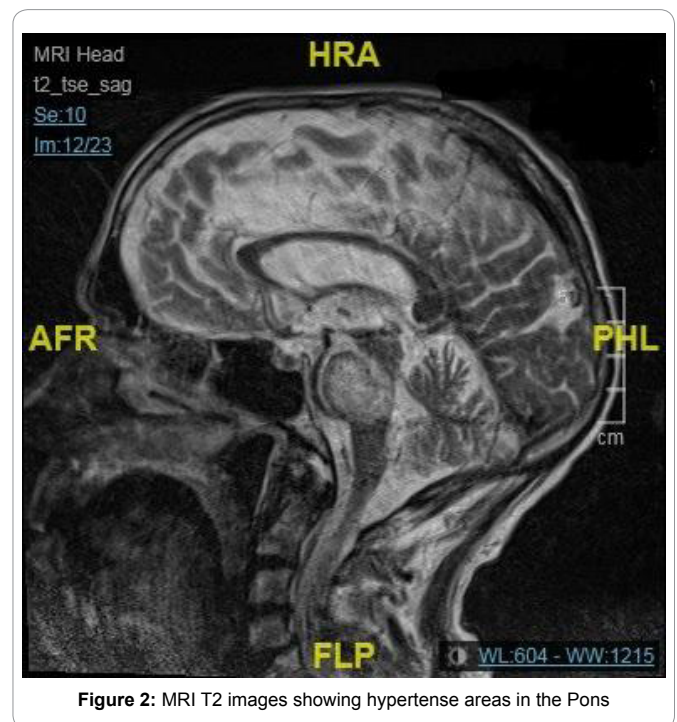


Figure 2: MRI T2 images showing hypertense areas in the Pons

better understanding of this condition and care [16,17]. This latter statement is exemplified by the recovery of this patient almost back to his normal. The cornerstone of care in these patients is supportive care and neuro-rehabilitation. This includes fluid and electrolyte balance, improving nutrition, vitamin supplements, managing seizures and alcohol withdrawal, minimising chest and catheter related infection risk and holistic nursing care. In established pontine myelinolysis several other methods have shown promising results. Administration of intravenous immunoglobulins to patients have shown to improve outcome [18]. Though unclear the mechanism is thought to be by detoxifying myelinolytic products and promoting remyelination [19]. Similarly plasmapheresis has also shown to be effective in this condition [20].

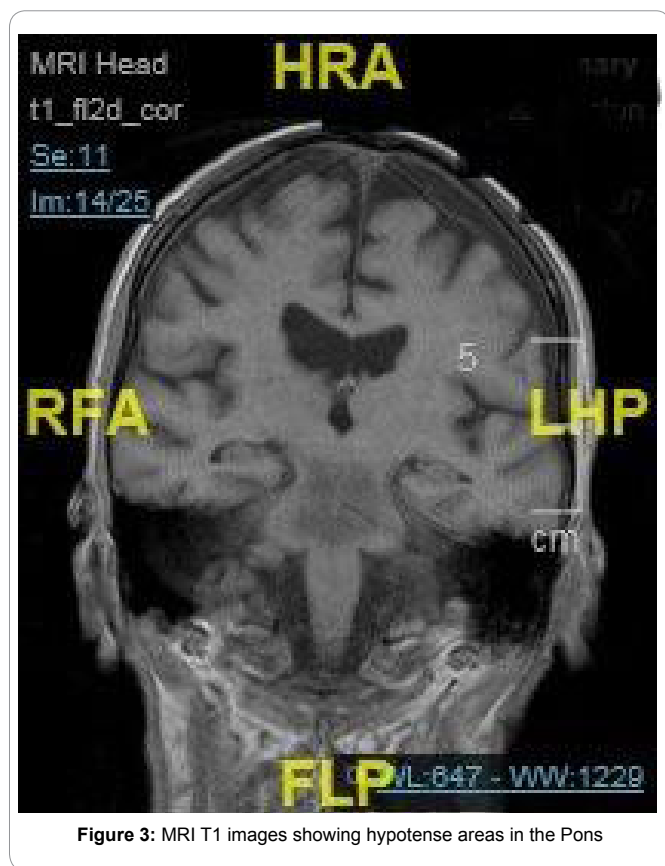


Figure 3: MRI T1 images showing hypotense areas in the Pons

Before embarking on treatment strategy it is important to identify patients with low serum sodium levels and prevent pontine myelinolysis by rapid correction. The accepted rate of sodium correction is 4-6 mmol/l in any given twenty four hour period [21].

References

1. de Souza A, Desai PK. More often striatal myelinolysis than pontine? A consecutive series of patients with osmotic demyelination syndrome. *Neural Res.* 2012;34(3):262-271.
2. Kallakatta RN, Radhakrishnan A, Fayaz RK, Unnikrishnan JP, Kesavadas C, Sarma SP. Clinical and functional outcome and factors predicting prognosis in osmotic demyelination syndrome (central pontine and/or extrapontine myelinolysis) in 25 patients. *J Neurol Neurosurg Psychiatry.* 2011;82(3): 326-331.
3. Bose P, Kunnacherry A, Maliakal, P. Central pontine myelinolysis without hyponatraemia. *J R Coll Physicians Edinb.* 2011;41(3):211-214.
4. Babar S. SIADH Associated With Ciprofloxacin. *Ann Pharmacother.* 2013;47(10):1359-1363.
5. Lim Leslie, Krystal Andrew. Psychotic disorder in a patient with central and extrapontine myelinolysis. *Psychiatry Clin Neurosci.* 2007;61(3):320-322.
6. Lim KH, Kim S, Lee YS, et al. Central pontine myelinolysis in a patient with acute lymphoblastic leukemia after hematopoietic stem cell transplantation: a case report. *J Korean Med Sci.* 2008;23(2): 324-337.
7. Singh N, Yu VL, Gayowski T. Central nervous system lesions in adult liver transplant recipients: clinical review with implications for management. *Medicine (Baltimore).* 1994;73 (2):10-18.
8. Gautam Dagur and Sardar A. Khan. Current Concepts in Pontine Myelinolysis: Review of Literature. *Transl Biomed.* 2015;6(4).
9. Seah AB, Chan LL, Wong MC, Tan EK. Evolving spectrum of movement disorders in extrapontine and central pontine myelinolysis. *Parkinsonism Relat Disord.* 2002;9(2):117-119.
10. Martin RJ. 0 Central pontine and extrapontine myelinolysis: the osmotic demyelination syndromes. *J Neurol Neurosurg Psychiatry.* 2004;75(Suppl 3):22-28.
11. Lauren R, Karp BI. Myelinolysis after correction of hyponatremia. *Ann Intern Med.* 1997;126(1):57-62.
12. Musana AK, Yale SH. Central pontine myelinolysis: case series and review. *WMJ.* 2005;104(6): 56-60.
13. Odier C, Nguyen DK, Panisset M. Central pontine and extrapontine myelinolysis: from epileptic and other manifestations to cognitive prognosis. *J. Neurol.* 2010;257 (7):1176-1180.
14. Ruzek KA, Campeau NG, Miller GM. Early diagnosis of central pontine myelinolysis with diffusion-weighted imaging. *AJNR Am J Neuroradiol.* 2004;25(2):210-213.
15. Kallakatta RN, Radhakrishnan A, Fayaz RK, Unnikrishnan JP, Kesavadas C, Sarma SP. Clinical and functional outcome and factors predicting prognosis in osmotic demyelination syndrome (central pontine and/or extrapontine myelinolysis) in 25 patients. *J Neurol Neurosurg Psychiatry.* 2011;82(3): 326-331.
16. Louis G, Megarbane B, Lavoué S, et al. Long-term outcome of patients hospitalized in intensive care units with central or extrapontine myelinolysis. *Crit Care Med.* 2012;40(3):970-982.
17. Luzzio, Christopher . Central Pontine Myelinolysis. *Medscape.* 2017.
18. Murthy SB, Izadyar S, Dhamne M, Kass JS, Goldsmith CE. Osmotic demyelination syndrome: variable clinical and radiologic response to intravenous immunoglobulin therapy. *Neural Sci.* 2013;34(4):581-594.
19. Rodriguez M, Miller DJ, Lennon VA. Immunoglobulins reactive with myelin basic protein promote CNS remyelination. *Neurology.* 1996;46(2):538-545.
20. Saner FH, Koeppen S, Meyer M, et al. Treatment of central pontine myelinolysis with plasmapheresis and immunoglobulins in liver transplant patient. *Transpl Int.* 2008;21(4):390-391.
21. Verbalis JG, Goldsmith SR, Greenberg A, et al. Diagnosis, evaluation, and treatment of hyponatremia: expert panel recommendations. *Am J Med.* 2013;126 (10 Suppl 1):S1-42.