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The Trident in the Brain- A Case of Osmotic Demyelination Syndrome Secondary to Alcohol Withdrawal

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ABSTRACT

Osmotic Demyelination Syndrome (ODS) is characterised by demyelination of pons and is also called as central pontine myelinolysis. It has been observed in patients with alcohol dependence, although hyponatraemia remains the most common cause. It should be suspected in chronic alcoholics and especially in those with protracted delirium tremens. Magnetic Resonance Imaging (MRI) is the modality of choice to diagnose this condition with a trident or pig snout appearance of pons being the characteristic finding. This publication discusses the case of a 27-year-old male who reported to the casualty ward in a delirious state following an episode of tonic clonic seizures and was diagnosed with alcohol withdrawal with pontine myelinolysis.

Keywords: Hyponatraemia, Pig's snout appearance, Pons, Tonic clonic seizures

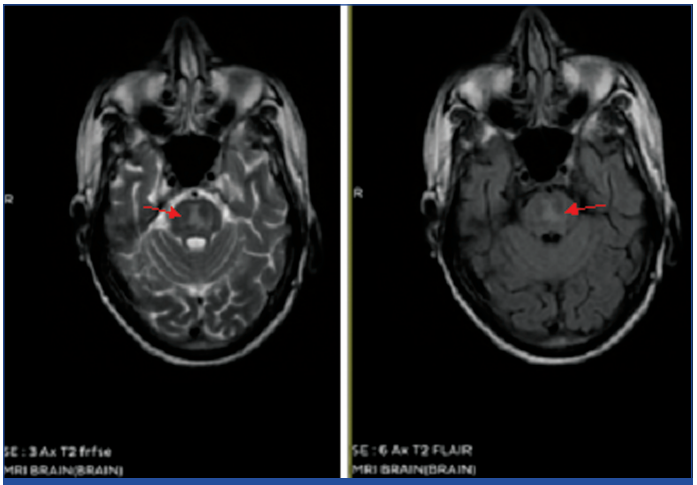
CASE DETAILS

A 27-year-old male was brought to the casualty in a delirious state following an episode of generalised tonic clonic seizure. He had no previous history of fever, trauma, headache, vomiting, loss of consciousness or seizures. On inquiring about his drinking pattern, it was established that he was an addict with the minimum consumption of at least 250 mL of country liquor a day since eight years. Following an attempted to cease consumption of liquor for three days, he developed a tonic clonic seizure and was brought to the emergency department of our hospital. There were no co-morbidities like diabetes, hypertension, bronchial asthma or tuberculosis.

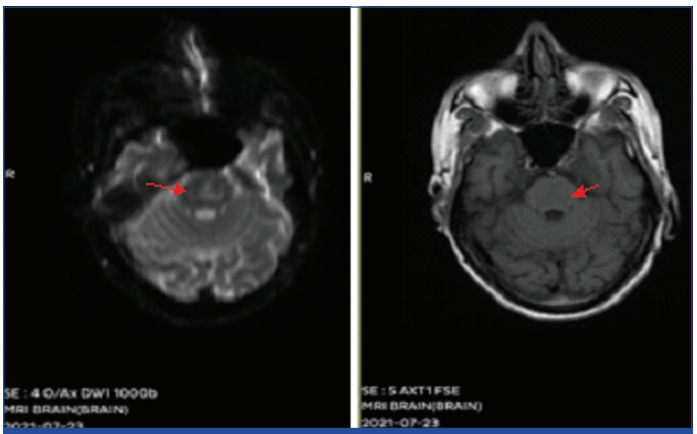
On examination he was conscious but disoriented with a Glasgow Coma Scale (GCS) score of $E_4V_4M_5$. His pulse was 88/min, Blood Pressure (BP) was 100/70 mmHg and respiratory rate was 33 breath/min. The Central Nervous System (CNS) examination revealed gaze evoked horizontal nystagmus, hypertonia in all four limbs, power of 5/5 with exaggeration of reflexes and an extensor planter response. On auscultation of the chest, breath sounds were equal on both sides with no adventitious sounds, normal heart sounds and no murmur. The abdomen was soft and non tender with no evidence of free fluid or organomegaly.

Laboratory investigations revealed a haemoglobin of 10 mg/dL, mean corpuscular volume of 100 fL, White Blood Cell (WBC) count of 13800/mm^3 a platelet count of 2,58,000/mm^3, Glutamic Oxalacetic Transaminase (SGOT) of 67 IU/L, Glutamic-pyruvic Transaminase (SGPT) of 49 IU/L, serum bilirubin of 1.4 mg/dL, serum protein of 4 mg/dL, serum ammonia of 12 micromol/L, serum lipase of 24 U/L, serum amylase of 35 U/L with the urea as 24 mg/dL, creatinine as 1.4 mg/dL, sodium as 137 mmol/L and potassium as 3.8 mmol/L. He was managed as a case of alcohol withdrawal with cefotaxim, thiamine, antacids, antiemetics, lorazepam and i.v. fluids. Following the above treatment, patient improved; he was conscious, oriented, showed spasticity in all four limbs, catatonia, hyperreflexia, an extensor planter response, nystagmus and dysarthria.

An MRI brain was done on the 2nd day which was suggestive of altered signal intensities in the mid brain and pons on T2WI/Fluid Attenuated Inversion Recovery (FLAIR) [Table/Fig-1] with corresponding hypo intensity in T1WI with restriction on Diffusion Weighted Imaging (DWI) [Table/Fig-2] and a corresponding dark signal on Apparent Diffusion Coefficient (ADC). The signals were bilaterally symmetrical in the upper pons sparing the peripheral pons and corticospinal tract that gave it the classical pig snout appearance a.k.a "trident sign" seen in ODS. A neurologist was consulted who advised continuation of thiamine, antacids, antiemetics and lorazepam. Following this the patient was discharged on request. He was treated in the hospital for a total duration of 10 days during which there was only a slight improvement in spasticity and dysarthria.



[Table/Fig-1]: The T2WI/FLAIR with altered signal intensities in the pons. The signals were bilaterally symmetrical in the upper pons sparing the peripheral pons and corticospinal tract- classical pig snout appearance a.k.a "trident sign".



[Table/Fig-2]: Hypo intensity in T1WI with restriction on DWI.

DISCUSSION

The ODS is characterised by demyelination of pons most commonly due to hyponatraemia. A few case reports published have shown this to be an occurrence though rare among hypernatraemic individuals postpartum or those on haemodialysis for chronic renal failure. It has also been observed in those with alcohol withdrawal, burns, anorexia and severe liver disease [1]. Chronic alcoholics with severe liver disease and Wernicke's encephalopathy are more likely to develop this condition [2]. The proposed hypothesis to explain this syndrome is osmotic injury to the endothelium which may cause release of myelinotoxic substances that damage the grey matter. We report a case of Central Pontine Myelinolysis (CPM) in a young male with alcohol withdrawal and no electrolyte abnormalities.

Hyponatraemia with rapid correction of sodium levels is the best-known cause of ODS. With hyponatraemia of longer duration, the brain can remain isotonic by reducing the intracellular levels of osmolytes. With correction there is an increase in the extracellular tonicity which ultimately increases the intercellular tonicity. In rapid correction there isn't sufficient time for the brain to adjust causing osmolytes to continue moving out into the extra cellular space which causes demyelination [3]. In alcohol withdrawal, ODS is unrelated to the sodium levels. Commonly these patients present with tremors, anxiety, tonic-clonic seizures, psychomotor agitation and gait disturbances [4]. Withdrawal is distinguished from CPM by intact orientation and absence of visual hallucinations. Development of spastic quadriparesis and pseudo bulbar palsy indicates the involvement of pons and midbrain as evidenced in our case. Inability to maintain cerebral protective mechanisms against osmotic stress has been postulated as one of the reasons for CPM due to alcohol withdrawal [4,5]. Refeeding syndrome is a metabolic condition that occurs in patient who is starved or malnourished following reinstitution of nutrients. During prolonged periods of starvation most intracellular minerals are depleted. Consumption of food following this period of starvation leads to an osmotic stress which may cause shrinkage of cells with shearing of oligodendrocytes provoking demyelination especially in the region of pons [6]. A case of refeeding syndrome has been reported in an alcoholic with one week of fasting prior to presentation [6]. This theory can be ruled out in the index patient as the history obtained suggests no nutritional deficit or fasting. The MRI brain is used most commonly as a diagnostic modality in CPM with the classical trident sign or pig snout appearance of the pons confirming the diagnosis. Intensive rehabilitation and conservative management remains a key in managing a case of osmotic demyelination with the prognosis being slightly better when it is caused due to alcohol withdrawal than due to rapid correction of sodium.

CONCLUSION(S)

The ODS or central pontine myelinolysis is a condition seen in patient with alcohol withdrawal with the characteristic trident sign or pig snout appearance of the pons which is a characteristic finding on MRI brain. Thus, MRI brain helps in confirming the diagnosis and further proceeding with the management.

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