Case Report

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MRI Findings in Osmotic Demyelination Syndrome- A Report of Two Cases

Radiology Section

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ABSTRACT

Osmotic Demyelination Syndrome (ODS) is a condition seen due to the loss of normal myelin in the central nervous system. Multiple etiologies have been identified as causing this condition, however, the most reported cause is the rapid correction of hyponatraemia. Different levels of the central nervous system are involved in the condition. Despite variable etiologies and clinical outcomes of the disease, the Magnetic Resonance Imaging (MRI) findings are well-established and characteristic. Hereby, two cases (45 years and 25 years old males) of ODS are described, to demonstrate the characteristic magnetic resonance imaging findings of the brain. Both cases were chronic alcoholics with a history of few episodes of vomiting, who later developed neurological symptoms. Their biochemical investigations showed severe hyponatraemia. The hyponatraemia was corrected rapidly, following which they developed neurological symptoms. Non contrast MRI brain was done for both patients, which showed abnormal hyperintense T2/Fluid-attenuated Inversion Recovery (FLAIR) signals in central Pons. Both cases showed variable, however characteristics findings of ODS on MRI. Despite, multiple causative factors and clinical outcomes of ODS, MRI with its characteristic findings plays a key role in diagnosis.

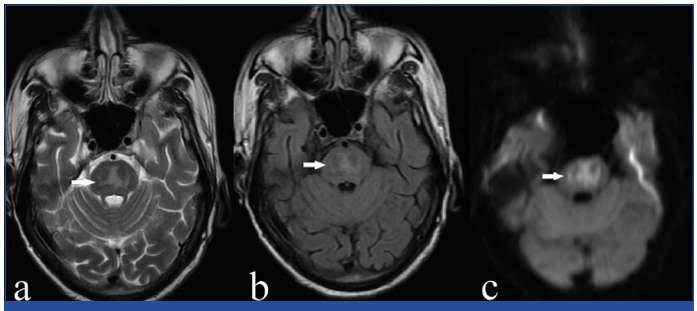
Keywords: Hyponatraemia, Myelin, Magnetic resonance imaging

CASE REPORT

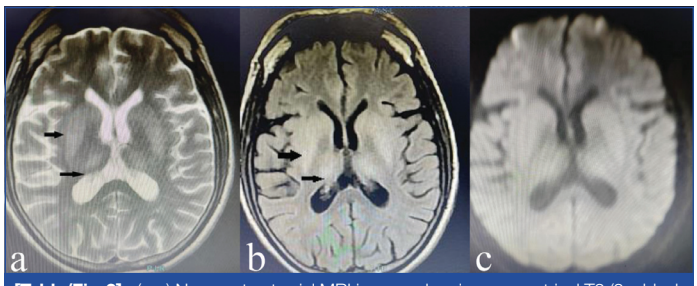
Case 1

A 45-year-old male patient, presented with generalised weakness and vomiting for four days, followed by tremors and difficulty in speaking. The patient was a chronic alcoholic for 7 years, patient gave 3-4 episodes of vomiting and pain in the abdomen but gave no past history suggesting any neurological symptoms. Clinical examination, showed normal pulse (85/minute) and blood pressure (130/80 mmHg). Biochemical investigations revealed severe hyponatraemia with a serum sodium level of 95 mmol/L. The hyponatraemia was corrected with 3% hypertonic saline infusion and his serum sodium levels were normalised (135 mmol/L) within 48 hours.

However, on 4th day of admission, the patient's mental status started deteriorating with the development of seizures and he became stuporous. Non contrast MRI brain was advised and done on the same day. Axial T2 wt/FLAIR MRI images showed hyperintense signals, showing restricted diffusion in central pons with relative sparing of peripheral cortico-spinal tract resembling "piglet sign" [Table/Fig-1]. Symmetrical T2 and FLAIR hyperintensities were also seen in bilateral basal ganglia and thalami. The basal ganglia and thalami lesions did not show any restricted diffusion on diffusion-weighted images (DW images) [Table/Fig-2]. Diagnosis of ODS (both central pontine myelinolysis and extra-pontine myelinolysis) was made based on clinical, biochemical and radiological findings. Supportive and conservative therapy was started, in form of commencement of 5% dextrose water infusion and desmopressin 2 µg intravenously 6 hourly, to which the patient responded. In the next three days, patient's neurological symptoms showed improvement and after a week's time patient became asymptomatic.



[Table/Fig-1]: (a-c): Non contrast axial Magnetic Resonance Imaging (MRI) images- T2 wt image (1a, white arrow),FLAIR image (1b , white arrow) showing hyperintense signals in central pons with relative sparing of peripheral cortico-spinal tract, showing restricted diffusion in Diffusion Weighted (DW) images(1c, white arrow)- giving "piglet sign".

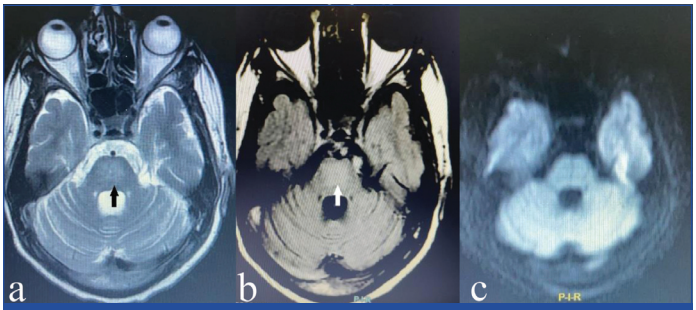


[Table/Fig-2]: (a-c) Non contrast axial MRI images showing symmetrical T2 (2a, black arrows) and FLAIR (2b black arrows) hyperintensities seen in bilateral basal ganglia and thalami showing no restricted diffusion on DW images (2c).

Case 2

A 25-year-old young male presented in the Emergency Department with chief complaints of vomiting for two days and three episodes of seizures with the development of altered sensorium for last one day, his vitals were stable though. He had a history of chronic alcohol abuse for the past four years, Biochemical investigations showed hyponatraemia with serum sodium level of 102 mmol/L. He was managed conservatively and the sodium levels were rapidly corrected over a period of 24 hours, and his sodium levels reached 140 mmol/L after 24 hours with intravenous infusion. On 5th day of hospitalisation, the patient's mental status deteriorated with the development of delirium and tetraplegia. Non contrast MRI brain showed hyperintense signals in central pons on T2 wt and FLAIR images. No evidence of restricted diffusion was seen [Table/Fig-3]. The clinical, biochemical, and radiological findings were consistent with ODS (central pontine myelinolysis). Over the next two weeks of hospitalisation, the patient's clinical condition showed

no improvement and later the patient developed hospital-acquired infection with acute kidney injury and could not be saved.



[Table/Fig-3]: (a-c)-Non contrast axial MRI images showing hyperintense signals in central pons on T2 wt (3a, black arrow) and FLAIR images (3b, white arrow). No evidence of restricted diffusion was seen on DW images (3c).

DISCUSSION

Osmotic Demyelination Syndrome (ODS), is a condition occurring secondary to the loss of normal myelin in different levels of central nervous system [1]. Depending upon the level of central nervous system involved, it is subdivided into Central Pontine Myelinolysis (CPM) and Extra-pontine Myelinolysis (EPM) [1,2]. These two conditions can co-exist or present in isolation [2,3]. In the former condition, pons is mainly involved while in the latter, basal ganglia and thalami are predominantly involved [2]. The history of myelinolysis dates back to 1949 when a young male with delirium tremens and pneumonia was treated by two neurologists Raymond Adams and Maurice Victor as the patient developed quadriplegia, facial weakness, dysphagia, and a positive Babinski sign, indicative of affection of corticospinal tract pointing to pontine lesion [4]. The lesion could not be attributed to pontine infarct or basilar arterial thrombosis based on clinical signs. Autopsy findings revealed bilaterally symmetrical abnormality involving central pons and microscopic examination showed destruction of myelin-sparing neurons and axons [4].

After evaluating three similar cases, ten years later in 1959, Adams and colleagues named this condition “central pontine myelinolysis”. Due to the predominant involvement of the central pons, they termed it as “central pontine” and the word “myelinolysis” was used to avoid any confusion with demyelination of multiple sclerosis and similar diseases causing myelin loss [4]. Later as a few similar cases showing symmetrical involvement of extra-pontine structures were reported, the concept of “extra-pontine myelinolysis” was introduced from 1962 onwards [4,5].

The exact attributing factor for ODS is still not very clear, however rapid correction of hyponatraemia has been found to be the most common etiology. The other attributing factors are alcoholism, post-liver transplant, severe burns [3], and hyperglycaemia with associated acidosis [6]. Due to rapid change in serum osmolality, there occurs sudden shrinkage of the brain cells [6,7]. The cell shrinkage makes brain cells, especially oligodendrocytes susceptible to death, which causes myelinolysis [7] Gray matter and white matter are anatomically intermixed in the pons, making pons highly vulnerable to osmotic myelinolysis [7].

Amongst available imaging modalities- Computed Tomography (CT) and MRI, MRI has been found most sensitive in evaluating radiological changes in ODS. The radiological changes which are reported in literature are mostly seen after 7-14 days of acute osmotic shift [2,7]. The most common MRI findings seen in CPM are bilaterally symmetrical hyperintensities in T2 and FLAIR in central pons with sparing of cortico-pyramidal tracts [4,8,9]. Many of these lesions are seen to show restricted diffusion [8,10,11]. Conditions like acute disseminated encephalomyelitis, multiple sclerosis, and pontine infarcts are radiological mimics; however clinical history in association with biochemical parameters and the course of disease progression can help differentiate it from CPM. A pontine neoplasm can also mimic CPM, however, the absence of postcontrast enhancement and mass effect in CPM helps to rule out pontine neoplasms [6]. In agreement with the above literature on MRI findings, the second case, on MRI showed a hyperintense pontine lesion with no mass effect. In correlation with clinical history, biochemical parameters (showing hyponatraemia), and history of rapid correction of hyponatraemia, a diagnosis of CPM was made.

In both the present cases, the lesions (the extrapontine lesions in the first case and the pontine lesion in the second case), did not show restricted diffusion. In contrast, Ruzek KA et al., presented a case report showing restricted diffusion in lesions (seen on day 12) preceding the changes in form of hyperintensities on T2/FLAIR images (seen on day 17). The radiological changes in pons on DW were seen in their case within 24 hours of the onset of tetraplegia [10]. Cramer SC et al., and Ruzek KA et al., advocated use of DW imaging for early diagnosis of ODS. [8,10]. In most of the cases, the pons hyperintensity showed a characteristic “trident shape” on axial images [9-11]. In lack of any literature of variability of this finding, the authors believed that the presence of restricted diffusion on MRI could be a variable finding based on the chronology of events, which could be evaluated on a larger cohort of patients in a temporal analysis.

Case reports by Wagner J et al., [12] and Sonjjay pande et al., [13] found hyperintense signal abnormalities in pons partially sparing tegmentum, ventrolateral pons and corticospinal tracts on FLAIR, which on axial images resemble face of a pig-hence named as “piglet sign” (both the carotid arteries being eyes, temporal lobes representing ears, and the abnormal pons signal resembling-pig snout, the fourth ventricle is the mouth of the pig) [12,13]. In the first case of this report, piglet sign was well demonstrated, which correlates well with this literature.

The radiological findings seen in EPM are bilaterally symmetrical T2 and FLAIR hyperintensities in basal ganglia, thalami [4]. The radiological mimics showing similar MRI features are hypoglycaemia, hypoxic ischaemic syndrome and carbon monoxide poisoning which can well be differentiated by hypoglycaemic state, evaluating blood sugar levels and history of hypoxia or poisoning, respectively. Chronic history of vitamin B12 deficiency with MRI features showing patchy and asymmetrical basal ganglia lesions helps differentiate wernick’s encephalopathy from EPM [7]. Infective condition such as Japanese encephalitis shows similar MR features however this condition is associated with fever and clinical features of encephalitis. Creutzfeldt-Jakob disease, another mimicker, seen in immunocompromised cases, can be differentiated from EPM with the help of T1 images which shows hyperintensities in involved regions [7]. In the first case of the present report, clinico-biochemical and MR features leads to diagnosis of ODS with both CPM and EPM involvement.

Y Tatewaki reported a case of ODS involving cortical and subcortical regions of temporal lobes in addition to signal abnormalities in pons [14]. Babanrao SA et al., [7] and Lee C and Ko C [2] individually reported cases where the extrapontine myelinolysis preceded central pontine myelinolysis. Individual case reports by Bhatia S et al., [11] and Saroja AO et al., [15] cited case of ODS following hypernatremia, involving the posterior limb of internal capsule, anterolateral midbrain, anterior pons and middle cerebellar peduncles and splenium of corpus callosum. On T2 WT coronal images, hyperintense signals were seen symmetrically involving cortico-spinal white matter from internal capsule to pons giving “wine glass appearance” and extending to middle cerebellar peduncle with splenium.

CONCLUSION(S)

Osmotic demyelination syndrome can occur due to multiple different causes with variable clinical outcome, however, MRI with its characteristic findings play a key role in the diagnosis.

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