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OSMOTIC DEMYELINATION SYNDROME: CASE REPORT

M. NGUNGA, R. HARANIA, K. HAMEED and J.O. JOWI

SUMMARY

We present a 40 year old man who was admitted to the hospital with convulsions. His final diagnosis was osmotic demyelination syndrome (ODS). We discuss the diagnostic and management challenges and the possible complications of this rare diagnosis.

INTRODUCTION

Osmotic demyelination syndrome (ODS) or central pontine myelinosis (CPM) is a rare condition that is characterised by varying levels of neurological deficits and is caused by neuronal loss due to diffuse non-inflammatory demyelination mainly in the midbrain, cerebrum and cerebellum. ODS is a well recognised complication of rapidly corrected severe hyponatraemia. This was first recognised by Tomlinson in 1976. Other conditions associated with increased risk of the syndrome include chronic alcoholism, malnutrition, prolonged diuretic use, liver failure, receiving organ transplant and extensive burns; all of which have inherent hyponatraemia (1).

In a study of 3000 brain autopsies, there were 15 cases of asymptomatic CPM (2). Musana and Yale (3) between 1986 and 2003; identified six cases ranging in age from 31-73 years (mean age 51.5 years); five of these patients were alcoholic and had favorable outcomes on conservative management. The exact incidence of ODS in Kenya is unknown. We reviewed medical records at the Aga Khan University Hospital Nairobi for this condition and two cases were noted; in 1994 and 2002.

CASE REPORT

A 40 year-old known alcoholic male consuming greater than 100 g alcohol per day for over 20 years presented to the hospital following an episode of generalised tonic-clonic seizure during an alcohol binge. There was no history of febrile illness or trauma. The patient was conscious but disoriented in time, place and person. He had been admitted two weeks earlier at another health facility.

On examination the patient was wasted and dehydrated. He was drowsy but arousable; with a tinge of jaundice but there was no pallor, cyanosis, finger clubbing or lymphadenopathy. His temperature was 37.5°C; blood pressure was 100/76 mmHg; radial pulse was 89 bpm regular and respiratory rate was 22 per minute. Oxygen saturation at room air was 96%. On central nervous system examination, he was found to be disoriented in time, place and person. He had dysarthria and impaired short and long term memory. The Glasgow Coma Scale was 12/15. The neck was supple and Kerning's sign was negative. The pupils were 2mm in diameter and reactive to light. Consensual light reflex was normal. There was no nystagmus or ophthalmoplegia. Cranial nerves were normal.

He had left hemi-paresis-muscle power grade 4 on the Medical Research Council (MRC) scale. Sensation was normal and co-ordination could not be appropriately ascertained. His gait was ataxic. Other systems were normal.

The differential diagnosis included hepatic encephalopathy, meningoencephalitis, and possible metabolic encephalopathies. Computed tomography scan of the brain revealed a hypo-dense lesion in the midbrain and diffuse brain atrophy but no features of meningeal enhancement or raised intracranial pressure. The initial chest X-ray was normal. Other blood work is as shown in Table 1.

The patient was admitted to the High Dependency Unit but later transferred to the ICU because of deteriorating neurological state. He was rehydrated and B-complex vitamins supplemented. He was commenced on naso-gastric tube feeding and potassium chloride was supplemented until normal levels were achieved. Cerebro-spinal fluid analysis was normal.

Magnetic resonance imaging of brain showed hyperintense signals on T2W and FLAIR sequences in the pons and both internal capsules and anterior peri-ventricular areas. There was no enhancement on contrast. Gross brain atrophy was noted. There were no signs of elevated intracranial pressure. Features were in keeping with osmotic demyelination (ODS).

Table 1

Note normal sodium level and hypokalaemia. Sodium levels may be normal at this stage of disease

Haematology	Chemistry	Liver function tests	Others
Hb 12.3 g/dL	RBS 7.9 mmol/L	T. bilirubin 54 mmol/L	Ca^{++} 2.7 mmol/L
MCV 102 fl	Na^{+} 147 mmol/L	D. bilirubin 14 mmol/L	$P04^{3-}$ 0.21 mmol/L
WBC 5 x 10^{9}/L	K^{+} 3.0 mmol/L	ALP 107 u/L	Mg^{++} 0.73 mmol/L
Neutrophils 80%	Cl^{-} 114 mmol/L	GGT 392 IU	B_{12} 124 pg/ml
INR 1.26	Urea 4.9 mmo/L	SGOT 105 IU	Folate 3.4 ng/ml
HIV negative	Creatinine 42 mmo/L	SGPT 43 IU	Albumin 32 g/dL

While in the ICU his urine output increased dramatically to the range of 150-400 ml/h, amounting to 9L in 24 hours. The serum sodium rose to 155 mmol/L and the central venous pressure dropped to <4 cmH_2O. The urine specific gravity was low and the electrolyte values are shown in Table 2.

Figure 1

T1 weighted images showing hypodense region in the pons. Note brain atrophy

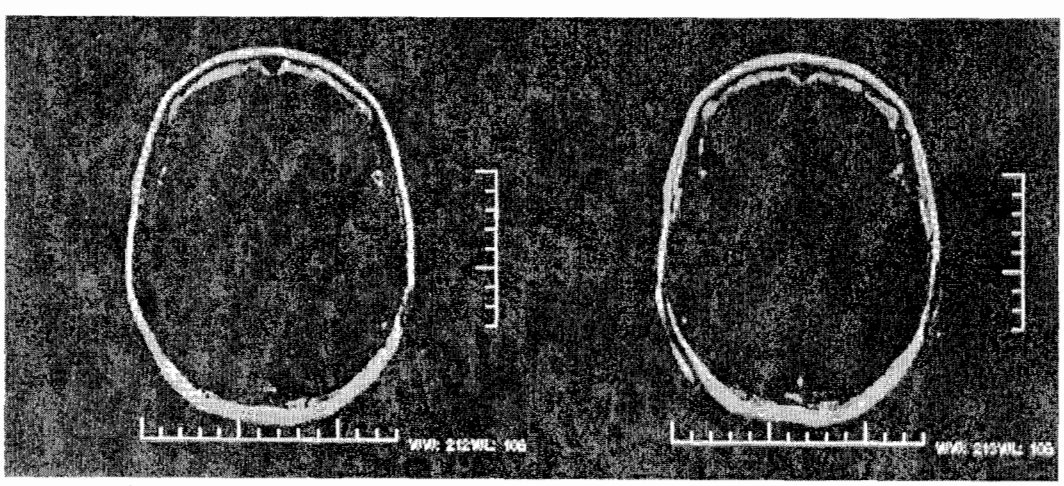


Figure 2

T2 weighted images. A high signal attenuation area in the pons (arrow) is the hallmark in ODS

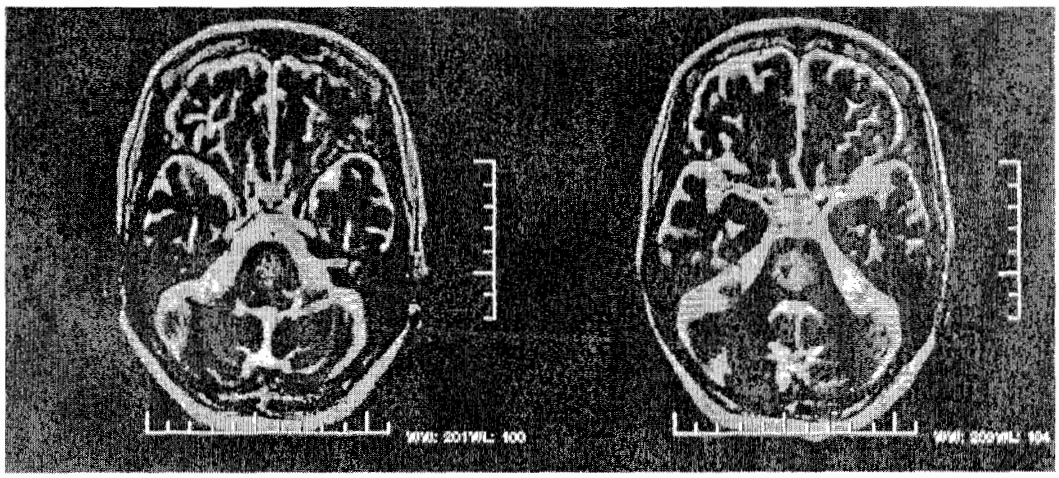


Table 2

Note marked reduction of urine osmolarity, an indirect marker of diabetes insipidus

	Urine	Serum
Na	59.5 mmol/L	137 mmol/L
K	27.2 mmol/L	3.8 mmol/L
Cl	–	105 mmol/L
Urea	51.6 mmol/L	1.3 mmol/L
Sugar	Nil	5.9 mmol/L
SG	1010↓	–
Osmolarity	224.6↓ mosm/L	288.8 mosm/L (290 mosm/L)

He was managed with intranasal desmopressin and urine output normalised. The desmopressin was discontinued after eight days of therapy; and his urine output remained between 1000-2000 ml/24 hours. Concomitantly, the patient showed marked neurological improvement; he was awake and could obey simple commands.

By the fourth week he could walk with assistance. He, however, still had dysarthria and swallowing reflex remained poor and needed naso-gastric tube feeding until one week prior to discharge. At subsequent review in the neurology out-patient clinic three weeks after hospital discharge he could walk with a tripod, and his speech had completely normalised.

DISCUSSION

Adams and colleagues first described CPM as symmetrical, non-inflammatory demyelination in the pons in 1958 (4). Extrapontine myelinolysis with or without pontine involvement was recognised in 1962 and occurs in at least 10% of patients with CPM, most often in the basal ganglia and thalamus (5,6). Although both conditions share the same pathology, the location of the lesions result in different clinical presentations. Classically CPM is associated with dysarthria and dysphagia due to corticobulbar fiber involvement as well as an initial flaccid quadriparesis. Extrapontine myelinolysis is characterised by tremor and ataxia and may be associated with other movement disorders. Mutism may be seen.

The diagnosis of ODS in this index case is mainly based on the history and the radiological findings. The bimodal form of disease characterised by hyponatraemia, seizures and confusion followed by neurological improvement and sudden deterioration in an alcoholic is almost pathognomonic. We were not able to demonstrate initial hyponatraemia in our patient but the seizure episode could be suggestive. The demonstration of the diffuse hypo-intensity seen in the *'basis ponds'* in the initial CT scan suggests development of pathology before patient was transferred to our care and was confirmed by MRI scan. Other causes of CNS disease were ruled out from investigations. Interesting to note is the

development of cranial diabetes insipidus. This can occur in theory, but has not been reported in the literature and we hereby wish to describe it as one of the complications of extra pontine demyelination. Diminished or absent ADH can be the result of a defect in one or more sites involving the hypothalamic osmoreceptors, supraoptic or paraventricular nuclei, or the supraoptico-hypophyseal tract. It is notable that as the DI improved the patient simultaneously got better neurologically. Central fever may occur because of involvement of the hypothalamus. This may have been the case in our patient since a full septic screen was negative.

Patients with chronic alcoholism are commonly admitted to hospital and given intravenous fluids as part of the treatment of alcohol withdrawal. These patients are predisposed to chronic severe hyponatraemia because of a variety of mechanisms including pseudohyponatraemia, hypovolaemia, "beer" potomania syndrome, cerebral salt wasting syndrome and the reset osmostat syndrome. If hyponatraemia is present, it is important to correct this slowly at a rate of less than 8 mmol/l/day to minimise the risk of developing ODS (7,8). MRI remains the mainstay in diagnosis and diffusion weighted images can pick early lesions and can be used for prognostic purpose to assess extent of disease (9,10). Thus, in patients with risk factors for chronic hyponatraemia and unexplained neurological deterioration a diagnosis of osmotic demyelination must be entertained. Hyponatraemia must not necessarily be demonstrated because this may be altered by previous therapy including intravenous fluids. MRI will be diagnostic in such cases; with a characteristic hyperintensity in the pons in FLAIR sequences. Other areas may too be involved and these lesions are characteristically non-inflammatory.

Microscopically the lesions show symmetrical myelin destruction affecting all the fiber tracts, with loss of oligodendrocytes. Recent studies have shown substantial axonal damage in central pontine myelinosis associated with an inflammatory infiltrate (11).

Treatment is supportive and the outcome is variable. In a study of 34 patients with osmotic demyelination syndrome two died and of the remaining 32, a third recovered; a third were debilitated but independent and a third were dependent (12).

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