

Case Report

Extrapontine Myelinolysis: A Case Report

Dr. Mohan. D. Kashinkunti^{1*}, Dhananjaya M²

¹Professor, Department of Medicine, SDM College of Medical Sciences and Hospital, Sattur, Dharwad-580009, Karnataka, India.

²Postgraduate student, Department of Medicine, SDM College of Medical Sciences and Hospital, Sattur, Dharwad-580009, Karnataka, India.

Corresponding author

Dr. Mohan D Kashinkunti

Email: drmohandk@gmail.com

Abstract: Central pontine and extra-pontine myelinolysis are a well-known complication of hyponatremia. The term "osmotic demyelination syndrome" is similar to "central pontine myelinolysis", but also includes areas outside the pons. Osmotic demyelination usually occurs in patients with rapid correction of hyponatremia. It is usually irreversible and can only be managed by prevention. We report a 57-year old man in whom osmotic demyelination occurred in the extrapontine area (extrapontine myelinolysis) after rapid correction of hyponatremia.

Keywords: Extrapontine, Central pontine, Hyponatremia, Osmotic demyelination

INTRODUCTION

Central pontine myelinolysis (CPM) was described by Adams and colleagues in 1959 as a disease that affects alcoholics and the malnourished [1]. The concept was extended from 1962 with the recognition that lesions can occur outside the pons, so-called extrapontine myelinolysis (EPM). In 1976 a link between these disorders and the rapid correction of sodium in hyponatraemic patients was suggested, by 1982 substantially established [2]. Whenever a patient is gravely ill with alcoholism and malnutrition or a systemic medical disease develops quadriplegia, confusion, pseudobulbar palsy, and pseudo coma over a period of several days, it is justified in making a diagnosis of central pontine myelinolysis (CPM) [3].

In extrapontine myelinolysis (EPM) the pathological changes are identical to those of CPM. Studies had shown that he lesions can occur with or without CPM [4]. The lesions are often strikingly symmetrical. The age of lesions in the various sites in EPM is contemporaneous. CPM and EPM are the same disease, sharing the same pathology, associations, and time course but differing in clinical manifestations [3].

When osmotic demyelination occurs, it can be lethal which is usually irreversible and has no proper management. Thus, prevention is most important. It is generally suggested that the rate of increase of serum sodium be no more than 1-2 mmol/liter/hour during the first few hours and should not be more than 8 mmol/liter during the first 24 hours [5].

We report on a patient who developed extrapontine myelinolysis after treatment for hyponatremia.

CASE REPORT

A 67-year-old man with a history of chronic schizophrenia disorder came to emergency room with complaints of slurring of speech, altered sensorium, excessive drinking of water, vomiting.

He had hypertension. There was past history of hyponatremia following intake of antipsychotic therapy and correction done by 3% saline and he improved. Physical examination revealed mild drowsiness but he could be aroused (GCS E2V3M4). The upper and lower limbs had generalized mildly decreased muscle power and normal deep tendon reflexes. The brain stem reflexes were all normal. His skin was dry and his ankles were not edematous. His blood pressure was 154/90 mmHg, pulse rate 80 beats per minute, respiratory rate 20 per minute, and body temperature 36.8°C. No other abnormalities were found on physical examination.

The laboratory data were blood hemoglobin 13.6 gm/dl, white cell count 12,300 cells/ μ L, urea 23 mg/dl, creatinine 0.7 mg/dl, sodium 101 mmol/L, potassium 3.0 mmol/L.

Because of drowsiness and past history of similar episode patient was started on 3% saline was injected in at a rate of 30 ml per hour to correct hyponatremia. There was slight improvement in GCS. The ensuing plasma sodium levels were 112 mmol/L 6 hours later, 120 mmol/L 12 hours later, 128 mmol/L 18 hours later, 135 mmol/L 24 hours later. The 3% saline was discontinued in 6 hours and was changed to normal saline and 5% dextrose water according to the plasma sodium level, to lower the rate of increase of the sodium level. After 24hrs he became stuporous (GCS E2V3M4)

and had seizures. He had 3-4 seizures per hour, each lasting about 3 minutes.

On examination patient had spastic quadriplegia. Patient was mechanically ventilated to support airway. Brain stem reflexes were intact and his vital signs were within normal limits. MRI scan diffusion and T2 weighted images showed hyperintensity in caudate and lentiform nucleus but no abnormalities in the brain stem including the pons [Figure]. The differential diagnosis

from the magnetic resonance imaging included extrapontine myelinolysis with cortical laminar necrosis, hypoxic-ischemic encephalopathy, or encephalitis. The clinical course and radiological findings were compatible with extrapontine myelinolysis.

Patient had recurrent seizures followed by aspiration pneumonia and patient expired after 72 hours.

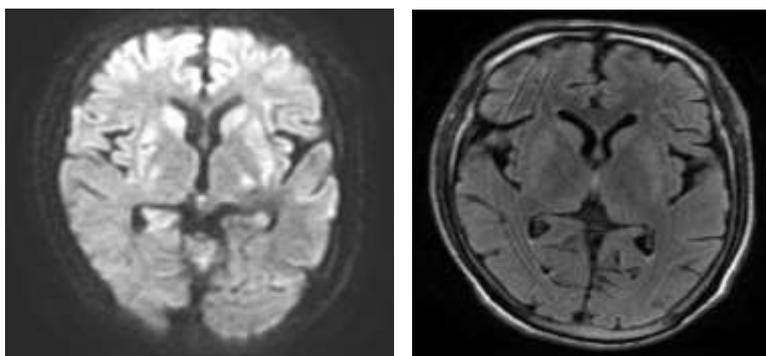


Fig.: Showing hyperintensity in Diffusion and T2W images in caudate and lentiform nucleus

DISCUSSION

Adams and colleagues first described CPM in 1959. In 1976, pontine and extrapontine myelinolysis were first found to be associated with rapid correction of low serum sodium levels [1, 2]. The clinical course following rapid correction of hyponatremia is biphasic with initial encephalopathy or seizures from hyponatraemia, followed by a rapid recovery as normonatremia is restored, with deterioration several days later. In CPM, the initial signs include dysarthria and dysphagia (secondary to corticobulbar fiber involvement), and flaccid quadriplegia (from corticospinal tract involvement) which later becomes spastic, all from involvement of the basis pontis. If the lesion extends into the tegmentum of the pons, pupillary and oculomotor abnormalities may occur. Also there may be changes in the level of consciousness [3].

EPM has identical pathological findings but the involved areas are extrapontine, so clinical manifestations are different [4]. AS EPM is rare, manifestations of it continue to attract further studies. It may manifest as postural limb tremors, a parkinsonian picture, myoclonic jerks, dystonia, catatonia or pyramidal dysfunction (secondary to the different areas involved). These may resolve completely or partially over months or they can become permanent [2].

The mechanisms of CPM and EPM are poorly understood, but the proposed mechanisms include osmotic injury to the vascular endothelial cells and this causes the release of myelinotoxic factors, the production of vasogenic edema and/or brain dehydration. This then causes separation of the axon from its myelin sheath with resultant injury to the

oligodendrocytes, particularly at the interface of the gray and white matter [6].

The most common causes of hyponatremia are therapy with thiazides, syndrome of inappropriate secretion of antidiuretic hormone in postoperative state, polydipsia in psychiatric patients, gastrointestinal fluid loss, ingestion of dilute fluid, and accidental ingestion of excessive water [7, 8]. These cases are mostly hypovolemic hyponatremia, as seen in our patient.

The most important aspect of hyponatremia management is a proper rate of correction. When hyponatremia develops rapidly and is accompanied by neurologic symptoms, more rapid correction is required. When hyponatremia develops slowly, slow correction is needed [9]. Generally, acute symptomatic hyponatremia should be treated by hypertonic saline and the plasma sodium level should be raised by only 1-2 mmol/L/hour. It should not be more than 8mmol/L during the first 24 hours. In Chronic asymptomatic hyponatremia, the plasma sodium level should be raised more slowly and not more than 8 mmol/L should be given during the first 24 hours [4]. The rate of increase in the sodium level is usually rapid if 3% saline is used intravenously, but it can also occur rapidly with only intravenous normal saline if the initial sodium level is profoundly low [11].

CONCLUSION

Hyponatremia is a manifestation of various disorders. Osmotic demyelination can be prevented. Proper and correct guidelines are needed to first identify the cause of hyponatremia and the correction should be done accordingly. Hypertonic saline should not exceed a rate of 1-2 mmol/L/hour and not more

than 12mmol/L/day. Especially when using intravenous 3% saline, the plasma sodium level should be checked frequently. If the sodium level increases very rapidly, water can be administered intravenously or orally.

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