Osmotic Demyelination Syndrome in a Eunatremic Patient with Chronic Kidney Disease

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Abstract

Osmotic demyelination syndrome is classically associated with rapid correction of hyponatremia. However, it can occur in normonatremic patients with other electrolyte abnormalities. One must suspect osmotic demyelination syndrome in susceptible patients with other electrolyte abnormalities like hypokalemia and hypophosphatemia.

Introduction

Osmotic demyelination syndrome (ODS) also known as Central pontine myelinolysis (CPM) was described by Adams and colleagues in 1959 as a disease affecting alcoholics and the malnourished. The concept was extended from 1962 with the recognition that lesions can occur outside the pons, so-called extrapontine myelinolysis (EPM). Classically this is associated with rapid correction of hyponatremia but it can also occur in presence of normonatremia. We present a case of ODS in a normonatremic patient with chronic kidney disease.

Case Report

A 35 years old male from a remote village, who had hypertension and chronic kidney disease stage IV (serum creatinine 4.5 mg/dl) and was taking indigenous therapy from a quack, presented to us with complaints of anorexia since 4-5 months, nausea, vomiting and loose stools of 2 days duration and drowsiness since 6 hours. Diarrhoea was watery, copious, about 10 times per day and was not associated with blood or tenesmus. There was no fever, headache, head injury, hematuria, dysuria or pyuria.

On examination, his blood pressure was 120/80 mm of Hg. On central nervous system examination, he was drowsy, spontaneously opening eyes and responding to command slowly. Pupils were normal. There was no focal neurological deficit, no meningeal signs, and plantar were flexor. Other systemic examination was normal. His investigation showed haemoglobin – 8.2 gm/dl, (normocytic normochromic anaemia), total leucocyte count 9,600 cumm, polymorph 60%, lymphocyte 36%, platelet count 1,55,000 cumm. His serum urea –131 mg/dl, creatinine –9.1 mg/dL, serum electrolytes: sodium –136 meq/L, potassium 3.0 meq/L, calcium 9.6 mg/dl, phosphorous 2.0 mg/dL. PTH 940 pg/ml. He had bilaterally shrunken kidneys on ultrasonography (right –6.8 cm and left –5.5 cm). His stool examination was normal.

He was diagnosed as a case of chronic kidney disease of unknown etiology (bilateral contracted kidneys) with hypertension. He was managed with intravenous fluids (2 L of normal saline), parenteral antibiotics (ceftriaxone and metronidazole), recombinant erythropoietin, alfacalcidiol, phosphate binders, calcitriol, amlodipine and hematinics along with hemodialysis and other supportive therapy.

Over the next 24 hours his sensorium worsened with GCS dipping to E1V1M3. The pupils were equal and reacting to light, but horizontal conjugate eye movements were restricted. There was spasticity with grade 2/5 power in all limbs, brisk reflexes and bilateral extensor plantar response. An urgent computerized tomography of the brain showed diffuse hypodensities in the pons extending into the middle and superior cerebellar peduncle. Magnetic resonance imaging (MRI) of the brain showed hyperintense lesions on T2 weighted images suggestive of pontine myelinolysis (Figures 1, 2).

He remained in hospital for 15 days and was treated with repeated sessions of hemodialysis and other supportive management but his sensorium did not improve and he finally expired after 1 month due to septicaemia and bilateral pneumonia.

References

4. Wong PC, Sanders SP, Jonas RA, et al. Pulmonary valve-stenosis and other congenital cardiac anomalies but it can also occur in presence of normonatremia. So, we present a case of ODS in a normonatremic patient with chronic kidney disease.

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Discussion

Rapid correction of hyponatremia is an important risk factor for the development of ODS, but ODS has been reported in normonatremic and hypernatremic patients also, especially in association with conditions like chronic alcoholism, liver transplantation, diabetes mellitus, hypokalemia, pituitary surgery, hepatocellular dysfunction, lithium toxicity, hypophosphatemia, chemotherapy and chronic renal failure. However osmotic demyelination, does not occur with the frequency one would expect in renal dialysis. It is thought that urea acts in renal failure patients as an “ineffective solute” — i.e. it contributes to measured osmolality but as it easily crosses cell membranes does not contribute to tonicity, thus protecting from the rapid shifts in sodium which can occur in haemodialysis. The signs of CPM include dysarthria and dysphagia (secondary to corticobulbar fibre involvement), 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 and pupillary, oculomotor abnormalities may occur. There may be an apparent change in consciousness level reflecting the “locked-in syndrome” that a large lesion in this site is particularly liable to produce. In Extrapontine Myelinolysis (EPM) the pathological changes are identical to those of CPM. A variety of sites may be involved. The lesions are often strikingly symmetrical.

Proposed hypotheses of CPM include osmotic injury to the endothelium resulting in release of myelinotoxic factors or vasogenic oedema and brain dehydration resulting in separation of the axon from its myelin sheath with resultant injury of oligodendrocytes. ODS have predilection to involve areas of rich gray-white matter apposition. This is probably due to endothelial changes (due to osmosis) leading to the release of myelinotoxic factors from the gray matter. Magnetic resonance imaging (MRI) is the imaging technique of choice. Hyperintense lesions are seen on T2, and hypointense lesions on T1 weighted images. The lesions are noncontrast enhancing. The timing of the appearance of lesions on MRI may be significantly delayed, and if the diagnosis remains likely a repeat imaging study at 10–14 days may reveal lesions not apparent on early scans. Diffusion weighted imaging (DWI) may aid in early detection of disease. DWI might have the capability of detecting lesions undetectable on T2.

CPM has been reported in the presence of hypokalemia and hypophosphataemia. In a recent study, hypokalemia was found to be a predisposing factor in 7 cases of CPM seen amongst 22 cases of hyponatremia, even when rapid correction of hyponatremia and non-acuteness of hyponatremia were not found to be the risk factors. For this apoptotic hypothesis has been proposed. It is suggested that a depletion of the energy supply to glial cells might limit the function of their Na+/K+-ATPase pumps. This could reduce their ability to adapt to relatively minor osmotic stress caused by small changes in serum sodium concentration, and ultimately lead to apoptosis.

Our patient possibly developed ODS due to multiple factors that may have predisposed him to have osmotic shifts like diarrhoea, hemodialysis and hypokalemia, hypophosphatemia and malnutrition.

Conclusion

Our case shows that ODS is not exclusively linked to hyponatremia and one must suspect ODS in susceptible patients with other electrolyte abnormalities like hypokalemia and hypophosphatemia.

References


Figs. 1 and 2: Magnetic resonance imaging of the brain showing Hyperintensity in pons with sparing of peripheral rim in T2WI