University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2019 , Vol. 5(3)

A CASE REPORT OF EXTRAPONTINE MYELINOLYSIS IN A PATIENT WITH ADDISON'S DISEASE

ROBERT WILSON S

Department of Neurology,MADRAS MEDICAL COLLEGE AND GOVERNMENT GENERAL HOSPITAL

Abstract : ABSTRACT .Osmotic demyelination syndrome can occur as a complication of therapy of chronic hyponatremia. Isolated extrapontine myelinolysis occurring in patients with hypoadrenalism has been quoted in the literature only rarely. Here, we present a patient with addisons disease who developed extrapontine myelinolysis during his recovery from his primary illness. He developed features of basal ganglia involvement as the manifestation of extrapontine myelinolysis. Our patient developed this complication when he was treated with iv fluids for his vomiting and dehydration. He developed parkinsonian features following the intravenous fluid therapy. The safe rate of correction of hyponatremia has not been accurately determined. The prognosis of this condition varies considerably, either complete recovery or mortality

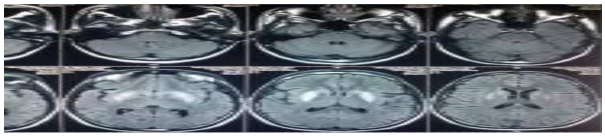
Keyword : central pontine myelinolysis(CPM), extrapontine myelinolysis(EPM), hyponatremia

INTRODUCTION

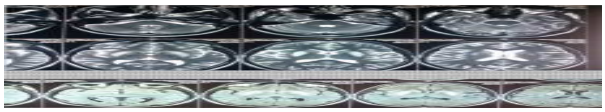
Central pontine myelinolysis was described by adams and colleagues in 1959. It was initially described in alcoholics with malnourishment. Later, it was found that lesions can occur outside the pons, so called extra pontine myelinolysis. The firm establishment of the link between these myelinolytic disorders and rapid correction of hyponatremia came in 1982 [1]. Isolated extrapontine myelinolysis is rare when compared to CPM or CPM with EPM. Adrenal insufficiency is one among the causes of chronic hyponatremia.

CASE REPORT

A 37 years old male was admitted with complaints of generalized hyperpigmentation for one year duration and difficulty in swallowing and speaking for one month. The hyperpigmentation was progressive, involving the whole body and he had lost weight significantly in the past one year. He developed postural giddiness for the past few months and during the evaluation he was found to have hyponatremia and further work up revealed addison's disease. He was started on fludrocortisone and hydrocortisone as replacement therapy. During the hospital stay he developed recurrent episodes of vomiting for which he was treated with iv fluids for several days. Two weeks after the hospitalization, he developed slurring of speech with reduced voice volume. He also had difficulty in swallowing with drooling of saliva and tremulousness of speech. There was no seizures, coma, bladder/bowel disturbances or higher function disturbances. On examination, the patient was conscious and oriented with normal MMSE and cognitive functions. He had hyperpigmentation over the entire body. Cranial nerve examination revealed no abnormality. He had bilateral cogwheel rigidity and extrapyramidal features like diminished blink rate, non accomodative glabellar tap and tongue tremor. Swallowing and gag reflexes were normal. Speech was of low volume and monotonous. Investigations revealed normal blood counts, renal and liver function tests. His serum sodium values were in the low normal range. Potassium levels were normal. No KF ring was detected on slit lamp examination. No acanthocytes were found in peripheral smear. USG abdomen and chest Xray was normal. His MRI revealed bilateral symmetrical T2, FLAIR hyper intensities in basal ganglia. Serum pyruvate and lactate were normal.



Extrapontine myelinolysis



Extrapontine myelinolysis

These images show bilateral symmetrical T2, FLAIR hyperintensities in basal ganglia.

DISCUSSION

This patient has been diagnosed with osmotic demyelination-extrapontine myelinolysis due to correction of hyponatremia of addison's disease. The common sites of osmotic demyelination in descending order of frequency are pons, cerebellum, lateral geniculate body, external capsule, extreme capsule, hippocampus, putamen, thalamus , cerebral cortex and caudate nucleus[2]. Caudate and putamen involvement is relatively rare. Pontine lesions constitute almost half of the cases of osmotic demyelination. Around three fifth of the remaining cases are due to combined CPM and EPM. The remaining small proportion is due to isolated EPM. Among EPM also, isolated basal ganglia involvement presenting with parkinsonism is even more

uncommon, with only few case reports in the literature. There has been very few case reports of EPM due to correction of hyponatremia of addison's disease presenting as parkinsonism (Gujjar et al,2010., Al-Mamari et al,2009, Okada et al, 2005) Pathogenesis involves oligodendroglial dehydration and apoptosis and metabolic stress imposed on cells that try to prevent osmotic degradation[3]. This entity has a biphasic clinical course. First phase is characterised by encephalopathy, seizures, then recovering rapidly as the hyponatremia is corrected. The second phase occurs few days after the improvement and consists of dysphagia, dysarthria and quadriparesis due to involvement of basis pontis. Extension into tegmentum may cause additional cranial nerve involvements. Locked in syndrome has also been reported in CPM. In EPM the manifestations may range from parkinsonism to mutism, catatonia and focal dystonias like torticollis, oromandibular dystonia and spasmodic dysphonia[4].

The prognosis of this condition varies considerably from mortality to complete recovery[5]. Many therapeutic modalities like thyrotropin releasing hormone, methyl prednisolone, plasmapharesis and ivig are being tried. Our patient was treated with antiparkinsonian drugs and his extrapyramidal features are decreasing.

CONCLUSION

Isolated EPM occurring during correction of hyponatremia in cases of adrenal insufficiency, presenting as parkinsonism are rare in the literature. This entity should be considered in a patient who fails to recover as expected after a severe illness requiring iv fluids. No safety limit for the rate of correction of hyponatremia has been determined. Prognosis of this condition is not uniformly bad and complete recovery can be seen.

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