Brief Communication

Central pontinemyelinosis, hyperparathyroidism, hypokalemia

Shyam Kishore, D. Kandasamy[1], Viveka P. Jyotsna

Department of Endocrinology and Metabolism, [1]Department of Radiodiagnosis, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Central pontinemyelinolysis (CPM) is one of the rare non-inflammatory demyelinating diseases of the pons and very rarely it can involve extrapontine structure as well. The exact etiopathogenesis of this condition is still unclear. Rapid correction of hyponatremia has been attributed as a most common factor, but alcoholism, malnutrition, prolonged use of diuretics, psychogenic polydipsia, post liver transplant and hypokalemia have also been attributed as a causative factor. We describe a case of hyperparathyroidism with concomitant hypercalcemia accompanied by central pontine myelinosis without hyponatremia/hyperosmolality or associated rapid corrections of sodium, which developed as a consequence of severe hypokalaemia.

Key words: Central pontinemyelinosis, hyperparathyroidism, hypokalemia

Introduction

Central pontinemyelinolysis (CPM) is a demyelinative lesion in the central pons, initially described by Adams *et al.,* in autopsy of his four patients of chronic malnutrition and alcoholism.[1] CPM is predominantly the lesion of basis pontis, but when the pathological process also involves extrapontine site it is called as osmotic demyelination syndrome (ODS). The exact incidence of this condition is not known. Rapid correction of hyponatremia has been attributed as a major cause for CPM. Here, we report a case of hyperparathyroidism with hypokalemia and normonatremia causing asymptomatic CPM.

Description of Case

A 50-year-old lady had symptoms of constipation, abdominal fullness and infrequent episodes of vomiting for last three months. Records revealed that after evaluation she was found to be a case of primary hyperparathyroidism based on biochemical tests, functional and structural imaging and underwent surgical exploration for parathyroidectomy but had a failed surgery.

She had history of one episode of expressive galactorrhoea. She did not have any history of headache, visual field defect or any features of pancreatic neuroendocrine tumor, thyroid disorders, any other neurological sign and symptom. She had persistently and progressively increasing level of serum calcium and serum intact parathyroid level with normonatremia and hypokalemia. There was no history of diabetes/hypertension/alcohol intake/malnutrition. Her hemoglobin was 7.2 mg/dl, blood urea 30 mg/dl, serum creatinine 1.4 mg/dl, and vitamin B 12 level >1200 pg/ml and serum folate > 24 ng/ml. Her thyroid function tests were normal. Her serum prolactin on three occasions were 146, 51 and 46 ng/ml, serum cortisol-24.46 mcg/dl, LH-15.38 miu/ml, FSH-34. 46miu/ml, 25(OH) vitamin D-15 ng/ml, iPTH-1634 and 2268 pg/ml on two occasions. Her liver function tests were normal. Her electrolytes were measured at frequent intervals. She had persistent hypercalcemia with serum calcium of 11.4 to 20.6 mg/dl along hypokalemia (2.1-3.3 meq/L), in spite of rich potassium diet and potassium supplementation. Her serum sodium ranged from 133 to 146 meq/L and plasma osmolality was 288 mmol/L.

Access this article online	
Quick Response Code:	Website: www.ijem.in
	DOI: 10.4103/2230-8210.119523

Corresponding Author: Dr. Viveka P. Jyotsna, Department of Endocrinology and Metabolism All India Institute of Medical Sciences, New Delhi, India. E-mail: vivekapjyotsna@gmail.com

On evaluation of association of possible MEN-1 syndrome, her MRI revealed [Figures 1a and b] T1 weighted post contrast sequence in axial plane (a) shows hypointense foci (arrow) in the pons with sparing of the periphery without any appreciable enhancement and fluid attenuated inversion recovery (FLAIR) sequence in axial plane (b) shows a hyperintense area (arrow) in the central part of pons with characteristic sparing of periphery. The imaging findings are consistent with central pontine myelinosis.

Her hypercalcemia was initially treated conservatively with hydration with normal saline but her serum calcium remained high. She received two doses of bisphosphonate along with calcitonnin, and low dose of diuretics, in spite of which her serum calcium steadily went up and she finally required three consecutive hemodialysis prior to her parathyroidectomy. Post parathyroidectomy, her serum calcium dropped down to 8.9 mg/dl and serum PTH to 47 pg/ml.

Discussion

The exact etiopathogenesis of CPM is unknown. Most CPM cases have been reported in association with rapid correction or overcorrection of hyponatremia.[2] Laureno *et al.*, on their animal based experiment suggested that the condition could be prevented by correcting hyponatraemia by less than 10 mmol/L in 24 h. Hypokalemia as a causative factor for CPM has also been reported in recent past irrespective of serum sodium level.[3,4]

Association of CPM with hypokalemia had been reported in patients of anorexia nervosa after starting feeding in them.[5] Clinical suspicion of CPM arises once there are neurological symptoms, but in our patient there were no neurological symptom and CPM was found once patient was evaluated for possible MEN syndrome due raised prolactin levels which later came normal. The initial raised prolactin level could be due to initial raised serum creatinine and associated medication. With recent advancement of radiological diagnostic modalities like MRI, cases of asymptomatic CPM are now being reported more.[6,7]

In our patient there was persistent hypercalcemia for which she required hydration, small dose of diuretic, calcitonin and bisphosphonate to lower down her calcium level. Due to her hypercalcemia she had off and on vomiting and polyuria. She had constantly received normal saline as an infusion to maintain hydration that might have maintained her sodium level. Patients who vomit lose hydrogen, chloride and water and develop hypokalemia not only because of shift of potassium ions into cells as a result of alkalosis, but also because of increased potassium ion excretion in the urine as a result of re-absorption of hydrogen ions.[8]

Aldinger KA, *et al.*,[9] studied a large group of patients of normal renal function with hypercalcemia to determine the prevalence of hypokalemia and reported that 16.9% had hyperparathyroidism, and the degree and frequency of hypokalemia were greatest at the higher serum calcium levels. It was postulated that the calcium delivery increases the sodium delivery to the distal tubule which in turn results in Na-K exchange with loss of potassium.

Secondary aldosteronism attributable to the decrease of total body water also causes an increase of potassium ion excretion in the urine.[10] Our review of literature showed many patients of CPM with hypokalemia but only few patients with hypokalemia and normonatremia.

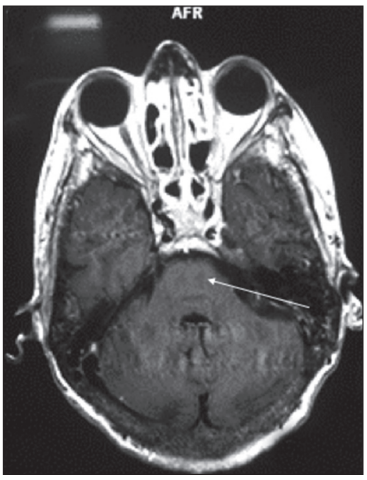

Figure 1a: T1 weighted post contrast sequence in axial plane shows hypointense foci (arrow) in the pons with sparing of the periphery without any appreciable enhancement and FLAIR sequence in axial plane

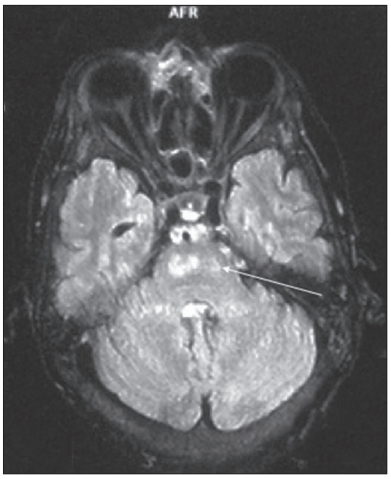

Figure 1b: T1 weighted post contrast sequence in axial plane shows a hyperintense area (arrow) in the central part of pons with characteristic sparing of periphery. The imaging findings are consistent with central pontine myelinosis

Conclusion

By reporting this case, we try to highlight the correlation of hyperparathyroidism with hypokalemia and subsequent asymptomatic radiologically detectable CPM which may go unnoticed and may have grave neurological consequences later.

References

1. Adams RD, Victor M, Mancall EL. Central pontine myelinolysis: A hitherto undescribed disease occurring in alcoholic and malnourished patients. AMA Arch Neurol Psychiatry 1959:81:154-72.
2. Laureno R, Karp BI. Pontine and extrapontine myelinolysis following rapid correction of hyponatremia. Lancet 1988;1:1439-41.
3. Bähr M, Sommer N, Petersen D, Wiethölter H, Dichgans J. Central pontine myelinolysis associated with low potassium levels in alcoholism. J Neurol 1990;237:275-6.
4. Lohr JW. Osmotic demyelination syndrome following correction of hyponatremia: Association with hypokalemia. Am J Med 1994;96:408-13.
5. Sugimoto T, Murata T, Omori M, Wada Y. Central pontine myelinolysis associated with hypokalemia in anorexia nervosa. J Neurol Neurosurg Psychiatry 2003;74:353-5.
6. Pfister HW, Einhäupl KM, Brandt T. Mild central pontinemyelinolysis: A frequently undetected syndrome. Eur Arch Psychiatry Neurol Sci 1985;235:134-9.
7. Girmenia F, Colosimo C, Di Biasi C, Faroni J, Pozzilli C. Central pontine myelinolysis: Report of an asymptomatic case. Ann Ital Med Int 1995;10:53-4.
8. Koh E, Onishi T, Morimoto S, Imanaka S, Nakagawa H, Ogihara T. Clinical evaluation of hypokalemia in anorexia nervosa. Jpn J Med 1989;28:692-6.
9. Aldinger KA, Samaan NA. Hypokalemia with hypercalcemia: Prevalence and significance in treatment. Ann Intern Med 1977;87:571-3.
10. Warren SE, Steinberg SM. Acid-base and electrolyte disturbances in anorexia nervosa. Am J Psychiatry 1979;136:415-8.

Cite this article as: Kishore S, Kandasamy D, Jyotsna VP. Central pontinemyelinosis, hyperparathyroidism, hypokalemia. Indian J Endocr Metab 2013;17:S114-6.

Source of Support: Nil, **Conflict of Interest:** None declared.