Research Paper

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Extra Pontine Myelinosis with Hypokalemia–a Rare Case Report

MEDICINE

KEYWORDS : Extra Pontine Myelinosis(EPM), Hypokalemia, Hyponatremia

DR SANKET SONI	MD 2ND YEAR INTERNAL MEDICINE, VS HOSPITAL, AHMEDABAD
DR SNEHA PATEL	MD 3RD YEAR INTERNAL MEDICINE, VS HOSPITAL, AHMEDABAD
DR PALTIAL PALAT	ASSOCIATE PROFESSOR OF INTERNAL MEDICINE,VS HOSPITAL, AHMEDABAD

ABSTRACT

Extra pontine myelinosis is commonly associated WITH rapid correction of sodium . We describe a patient with quadriparesis with predominant hypokalemia with mild hyponatremia. MRI imaging revealed the extrapontine lesions which disappeared on repeat study with symptomatic treatment. Hypokalemia as the predisposing factor in the pathogenesis For extrapontine myelinosis is highlighted.

INTRODUCTION:

Extrapontine myelinosis is characterized pathologically by non-inflammatory demyelination of various brain structures with sparing of axons. Common predisposing factors in a clinical setting of are alcoholism, malnutrition, liver disease and hyponatremia . EPM in a hyponatremic patient can develop only if it is less than 120 meq/L hrs for more than 48 hrs or it is corrected with hypertonic salines aggressively. We report a case of extrapontine myelinolysis associated with hypokalemia and mild hyponatremia wherein no rapid correction of sodium was done.

Case study:

A 38 year old hindu female with known case of hypothyroidism presented to the emergency room with sudden onset paralysis. The patient had past history of vomiting and diarrhea three days back and presented with sudden onset paralysis upper and lower extremities. The weakness was bilateral and involved both the proximal muscles of the shoulders and hips as well as the distal extremities. she had no respiratory or swallowing difficulty and was able to move her neck and facial muscles. she denied any pain or paresthesia, no urine or stool incontinence ,no bulbar weakness, no ocular complains, no periodicity and no other spinal cord symptoms. Prior to this episode, the patient had been healthy and denied any recent chest pain, shortness of breath, or weight change. she was taking tab. Thyrox. 50 µg every morning and denied use of alcohol or drugs, or significant changes in diet or activity levels. Her parents and siblings had no history of similar episodes and no other significant illnesses.

On physical exam, the patient's heart rate was 66/min and blood pressure was 140/90mm hg. She was normal in overall appearance. Her skin was cool and dry, and the oral mucosa was moist. No jugular venous distension, goiter or lymphadenopathy were appreciated. Cardiac exam revealed bradycardia with a regular rhythm and no murmurs. Examination of the lungs and abdomen were unremarkable. There were no deformities or edema of the extremities and distal pulses were present and equal bilaterally. Neurologic exam revealed normal higher functions ,flaccid paralysis of all extremities which involved the proximal and distal muscles and included the hips and shoulders. Sensation was intact but deep tendon reflexes were diminished to +2 in all four extremities. Cranial nerve function was grossly intact.

Blood sugar within normal limits. Blood urea 21mg/dl, Serum creatine 1.1mg/dl, Vit B12 level 523 pg/dl, magnesium 2.3mg/dl. liver profile normal.TSH level 10 IU/ml.CPK 469U/L. Mild hyponatremia (124-129 meq/l) but hypokalemia(2.3-2.4 meq/l) were found at repeated occasions. Plasma osmolality 288-290mmol/L throughout. CSF parameters were normal. Her EMG-NCV study was done and found to unremarkable. The patient was diagnosed with Hypokalemic Periodic Paralysis associated with hypothyroidism. After 6 hours of initiation of intravenous potassium replacement, the patient's neurologic symptoms improved.Gradual correction of potassium was done over a period of time.(fig 3)

Later on MRI was done that revealed relatively well defined hyperintense lesion on T2W and FLAIR sequences were seen at periphery of left cerebellar hemisphere and a small lesion at right cerebellar hemisphere inferomedial to cerebellar tonsil (Figure 1). Patient was treated conservatively with gradual correction of potassium and sodium as per protocols. The patient recovered clinically Repeat MRI done after two weeks after correction of electrolytes showed almost complete resolution of the lesions (Figure 2). And blood investigation like hemogram routine blood chemistry like liver profile, kidney profile and electrolytes (sodium potassium) was normal

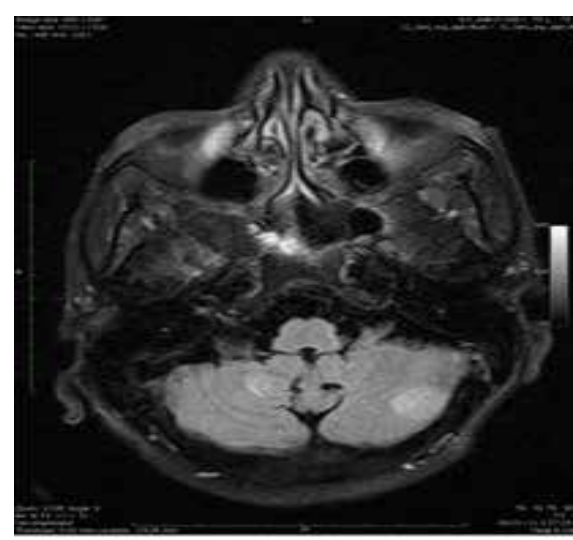

FIG 1: MRI show foci of bright signals in left inferior cerebeller hemisphere, right cerebeller hemisphere

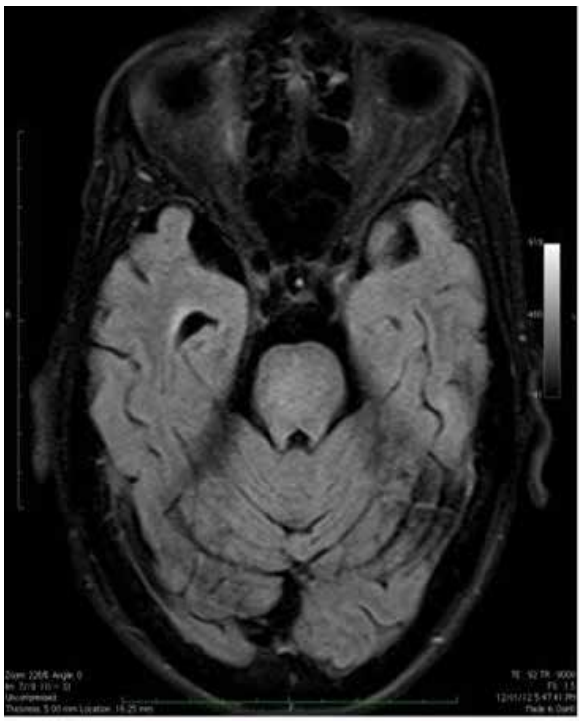

FIG 2: MRI taken a week after show almost complete resolution of bright signals of cerebellum in flair sequence

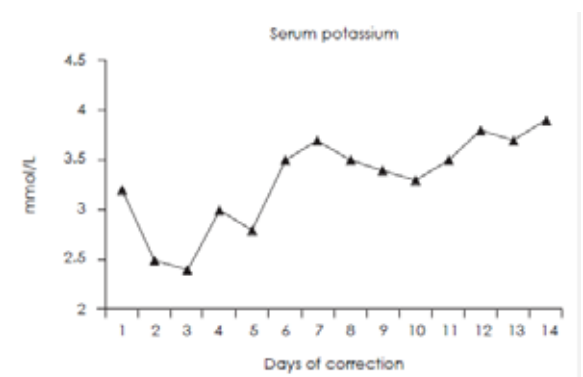

Fig 3. Daily level of potassium

DISCUSSION:

Extra-pontine myelinolysis is a uncommon variety of osmotic demyelination syndrome involving extrapontine structures with or without involvement of the pons. A variety of sites may be involved e.g., pons, cerebellum, lateral geniculate body, external capsule, hippocampus, putamen, cerebral cortex, thalamus, caudate nucleus, claustrum, internal capsule, midbrain, etc.Microscopically, the lesions show degeneration and loss of oligodendrocytes with preservation of axons and nerve cells without any evidence of inflammation.

Our case showed changes of extrapontine myelinosis on imaging studies with significant hypokalemia and mild hyponatremia, with no history of correction with hypertonic saline in the recent past . Hypokalaemia has been reported as a possible trigger in osmotic central pontine and extrapontine myelinosis. It is seen that EXTRAPONTINE MYELINOSIS tends to occur in hyponatraemia complicated by hypokelemia because a decreased concentration of Na, K-ATPase in endothelial cell membrane during hypokalemia predisposes the cell susceptible to injury by osmotic stress associated with the rapid rise in the serum sodium . We propose to highlight with this index case that even hypokalemia can also be an important predisposing factor in central and extrapontine myelinosis. Rapid clinical and radiological recovery in our patient and its association with hypokalemia makes it unusual.

CONCLUSION:

EXTRA PONTINE MYELINOSIS IS A RARE DISORDER.MANAGEMENT OF THIS INCLUDE RECOGNIZING HIGH RISK PATIENT ,GRADUAL CORRECTION OF ELECTROLYTES,MAKING PROMPT DIAGNOSIS AND MANAGING ASSOCIATE COMPLICATION.DECREASE MORTALITY DEPEND ON EARLY DIGNOSIS ,TREATMENT AND PREVENTION OF COMPLICATION.

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