ORIGINAL RESEARCH PAPER

Radiodiagnosis

OSMOTIC DEMYELINATION SYNDROME WITH KETOSIS AND WITHOUT ELECTROLYTE IMBALANCE, A RARE COMPLICATION OF JUVENILE DIABETES MELLITUS.

KEY WORDS: Central pontine myelinolysis (CPM) without electrolytes imbalance is an infrequent presentation of juvenile-onset diabetes mellitus. In the majority of central pontine myelinolysis (CPM) in young patients is associated with electrolytes imbalance. where in the older patient is associated with multisystem atrophy (MSA). I am presenting a case of young 10-year-old female child.

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ABSTRACT

Central pontine myelinolysis (CPM) is rarely reported in paediatric patients with diabetes. We report this case of a 10-year-old female with new onset diabetes. Patient presented with flaccid quadriparesis and bilateral lower motor neuron facial nerve palsy with associated history of difficulty on swallowing. MRI Brain showing central pontine myelinolysis (CPM) and bilateral parietal lobe edema. On biochemical examination blood glucose level 418mg/dl, urine ketone bodies insulin, patients condition improved. After ten days coarse of subcutaneous insulin twice a day HbA1c level was 7.1and mean plasma glucose level was 175mg/dl. MRI BRAIN shows significant improvement in CPM after 22 days compared to previous MRI scan and more improvement after 50 days with subcutaneous insulin twice a day. Though CPM is very rare with poor outcome.in our case prognosis of patient is good. And it will help us to plan a better protocol in future for CPM patient with diabetes.

INTRODUCTION

Central pontine myelinolysis (CPM) is rarely reported in paediatric patients with new-onset diabetes with ketoacidosis. Morbidity and mortality are very high. But in our case report, 10-year-old girl child patient presents with new-onset diabetes mellitus with ketosis which showing good prognosis.

Case History

10-year-old female child present with flaccidity and weakness of all four limbs more on the right side with a deviation of angle of mouth to left side since five days with history of difficulty of swallowing since three days, behavioural disturbance since two days, past history of polyurea and polydipsia since 15 days. MRI brain showing: Central pontine myelinolysis (CPM). Pons demonstrates an eventual high T2 signal. This region has a classic trident shaped appearance on T2/FLAIR axial MR images has been likened to the face of a pig, referred to as piglet sign. DWI showing diffusion restriction in the pons. There is T2/FLAIR hyperintensities in the bilateral parietal lobes. On biochemical examination blood glucose level 418mg/dl, urine ketone bodies positive, Na+ 134mmol/L, K+3.6mmol/L ,Ph-7.422, pCO2-30.5mmHg(low). After receiving subcutaneo us insulin ,patients condition improved .After 10 days course of subcutaneous insulin twice day Hba1c level is 7.1% and mean plasma glucose level was 175.46mg/dl noted . MRI brain shows significant improvement in MRI finding after twenty-two days compared to previous MRI scan and more improvement after fifty days with subcutaneous insulin twice a day.

DISCUSSION

Adams et al. first described CPM in 1959 in alcoholics and malnourished patients. CPM usually occurs with rapid correction of severe chronic hyponatremia, typically with serum sodium below 120 mmol/L. Alcoholism, malnourishment, and severely debilitating illnesses are risk factors for the development of CPM. The composition of the pons with maximum grey and white matter elements makes it more susceptible, however extrapontine involvement is also well described also is known as 'osmotic demyelination syndrome. Despite the pronounced fluctuations in serum osmolality, CPM is rarely seen in diabetes. To date, there are only few CPM cases associated with hyperosmolar hyperglycaemia in the absence of sodium abnormalities. Most recently in 2015, Saini et al. reported a case of CPM secondary to hyperglycaemia, which presented with ataxia and pseudobulbar affect and evolved sub acutely over a duration of two weeks, similar to the clinical presentation of our patient. initially in our hospital department of paediatrics thinks as a Guillain–Barré syndrome and treatment as I/V Ig with no significant improvement. Urine ketone bodies present with normal blood pH makes a conclusion of diabetic ketosis. Theories proposed behind the pathogenesis of osmotic demyelination in the setting of hyperosmolar hyperglycemia are: Subacute changes in brain cells secondary to hyperglycemia leading to demyelination; rapid changes in osmolality associated with the fluctuations in serum glucose causing osmotic demyelination and hypertonic insult associated with hyperglycaemia itself causing demyelination. A biphasic clinical course is seen in CPM usually, initially encephalopathic phase followed by deterioration of condition of patient. The clinical manifestations vary based on area of brain involvement and include dysarthria and dysphagia with corticobulbar fibre involvement, flaccid quadriparesis with corticospinal tract involvement, and, in severe cases, 'locked-in syndrome' Tremor, ataxia, movement disorders, and psychiatric and behavioural changes may be seen with extrapontine involvement. Rarely CPM can present as isolated gait ataxia MRI is the imaging technique of choice for diagnosing CPM. Typically, non-contrast enhancing, symmetric hyperintense lesions on T2-weighted images and FLAIR sequence, and hypointense lesions on T1 weighted images are seen. The treatment of CPM is mainly supportive. in our case, the patient's condition improved with subcutaneous insulin twice a day. The outcome of CPM varies from complete recovery despite initial dramatic presentation to death or permanent disability, but fortunately, in our case, now the patient is normal without any disability.

CONCLUSIONS

Central pontine myelinolysis (CPM) also known as osmotic demyelination syndrome is associated with seen in the setting of osmotic changes, typically with the rapid correction of Hyponatremia and It is rarely associated with diabetic ketoacidosis. in both condition prognosis is very poor survival rate. In our case report study patient with type-1 diabetes mellitus with ketosis showing very good prognosis in with subcutaneous insulin twice a day.

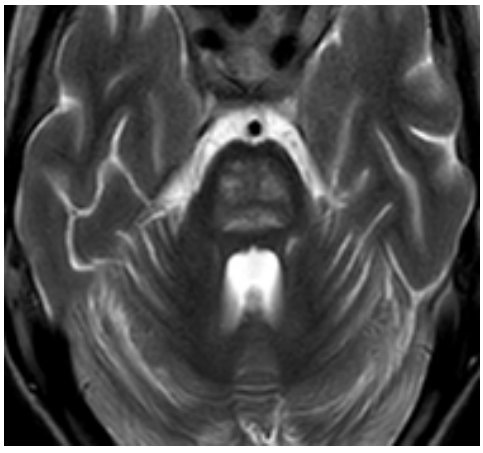

Figure-1 (a)t2w-mri Axial Image Of Pons Showing Classic Trident Appearance

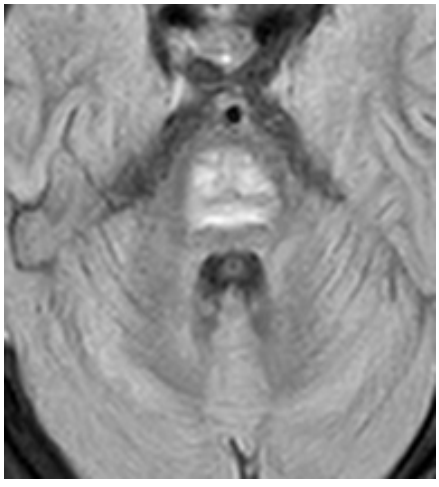

Figure-1 (b) Flair-mri Axial Image Of Pons Showing Classic Trident Appearance

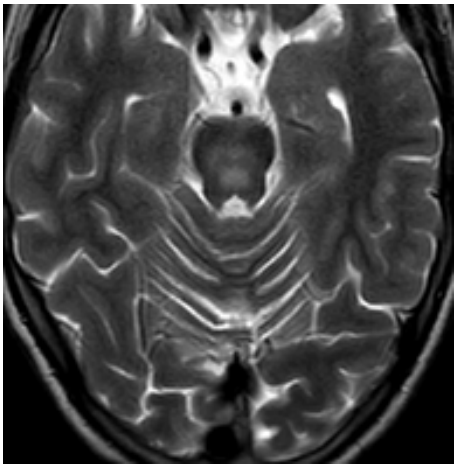

Figure-2 (a) T2w-mri Axial Image Of Pons After 22 Days With Subcutaneous Insulin Twice A Day Showing Improvement Compare Previous Scan

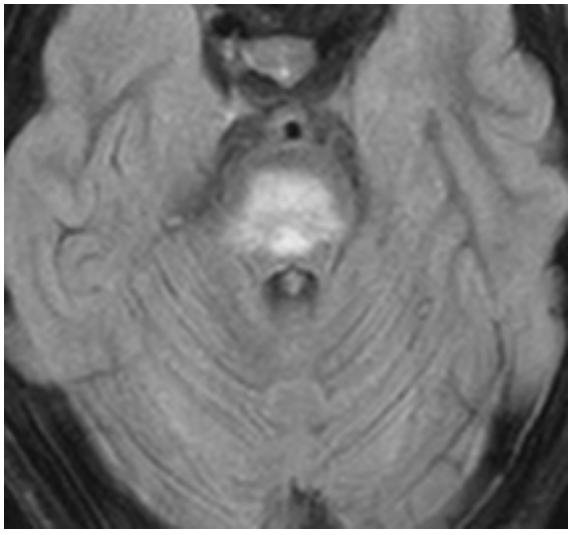

Figure-2 (b) Flair-mri Axial Image Of Pons After 22 Days With Subcutaneous Insulin Twice A Day Showing Improvement Compare Previous Scan

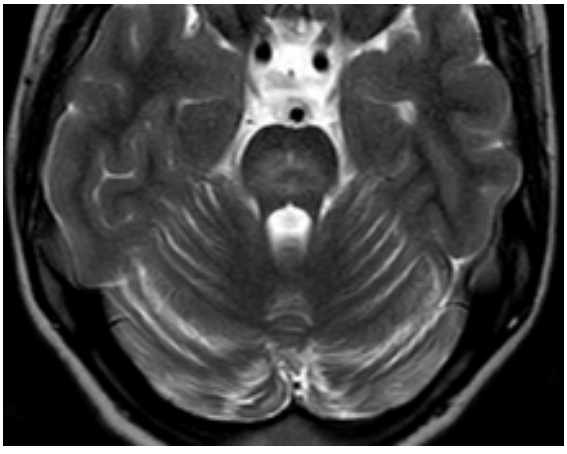

Figure-3 (a) T2w-mri Axial Image Of Pons After 50 Days With Subcutaneous Insulin Twice A Day Showing Improvement Compare Previous Scan

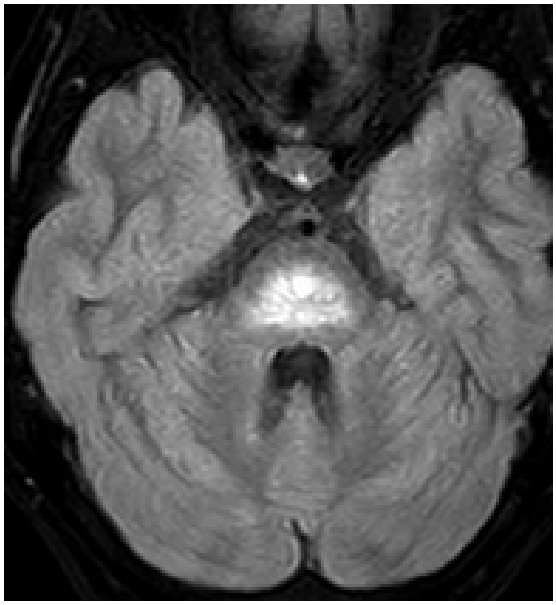

Figure-3(b) Flair-mri Axial Image Of Pons After 50 Days With Subcutaneous Insulin Twice A Day Showing Improvement Compare Previous Scan

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