

Central Pontinemyelinolysis: Report of a Rare Case with a Good Outcome.

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Abstract- Central pontinemyelinolysis is a noninflammatory demyelinating disorder characterized by the loss of myelin in the center of the basis pontis usually seen in those with chronic alcoholism and in malnourished persons and by rapid correction of chronic hyponatremia. The clinical features vary depending on the extent of involvement. Demyelination can occur outside the pons as well and diagnosis can be challenging if both pontine and extrapontine areas are involved. We herein report a case of myelinolysis involving pons.

Index Terms- Central pontinemyelinolysis (CPM), Extrapontinemyelinolysis (EPM), Osmotic demyelination syndrome(ODS).

I. INTRODUCTION

Central pontinemyelinolysis (CPM) was described by Adams and colleagues in 1959 as a disease affecting alcoholics and the malnourished¹. The concept was extended from 1962 with the recognition that lesions can occur outside the pons, so-called extrapontinemyelinolysis (EPM). In 1976 a link between these disorders and the rapid correction of sodium in hyponatraemic patients was suggested, and by 1982 substantially established².

The precise incidence of CPM is not known, but the ability to diagnose it during life has been helped by modern neuroimaging, particularly with an MRI of the brainstem. Clinical manifestations of CPM include truncal and gait ataxia, pseudobulbar syndrome, and bilateral upgoing plantar responses. There is no proven effective therapy for CPM^{3,4}.

In the past, the prognosis of CPM was thought to be poor with a mortality rate of over 50%. With greater general awareness of the disorder and a better ability to diagnose it, the prognosis is improving. In some instances, complete recovery of CPM has been reported.⁴

We report an unusual case of CPM affecting the brain in a man with chronic alcohol as known risk factor to develop this condition with normal serum sodium.

II. CASE REPORT

A 40 yr old male, K/C/O Pulmonary Koch's since 2 months(on ATT) presented with Slurring of speech since 1 week ,Choking on attempting to swallow liquids since 1 week ,Weakness of both upper limbs and lower limbs that gradually progressed over 4days.H/o weakness of limbs started as

difficulty in mixing his food,buttoning/unbuttoning his shirt & slippage of slippers with his knowledge;Later there was difficulty in combing his hair & stand and inability to get up from sitting/squatting position.There was difficulty in lifting his head/sitting from lying position without support. No H/o hoarseness of voice .No H/o S/o sensory involvement.No H/o S/o bowel & bladder involvement.No H/o S/o cerebellar symptoms.No history of similar complaints in the past.Past H/o fever & productive cough since 2 months; for which he was started on Cat I ATT.No past H/o HTN,DM.No H/o any other drug intake or previous hospitalization.Family history – Not significant.

PERSONAL HISTORY.-Chronic alcoholic-Consumed approx. 180 ml of whisky per day for the last 15 years.Non-smoker on examination patient was conscious & coherent thin built, moderately nourished(BMI-20) No pallor / icterus / cyanosis / clubbing, Nokoilonychia / lymphadenopathy / edema. No neurocutaneous markers. No peripheral nerve thickening.

VITAL DATATemp.- afebrile Pulse – 82/min;regular;normalvolume;all peripheral pulses felt;B.P – 100/70 mmHg in Lt. upper limb in supine position, Respiratory Rate – 20/min CNS examination Skull & Spine – normal, Oriented to time, place, person.

Memory-intact.Speech & Language – Dysarthria present - With abnormal lingual &velars (spastic dysarthria), Naming, comprehension, repetition, reading& writing present.

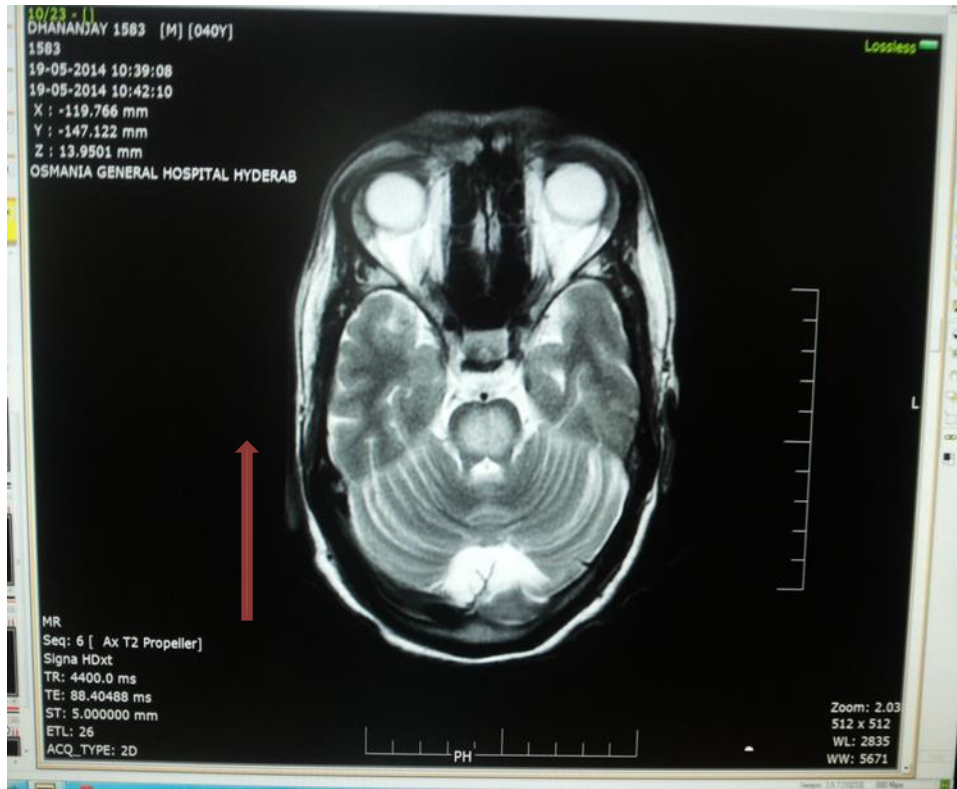
CRANIAL NERVES I - XII – Normal , Jaw jerk brisk, Gag & Palatal reflexes present, Decreased movements of tongue with spasticity; no fasciculations, all are suggestive of pseudobulbar involvement.

MOTOR SYSTEM: Bulk and tone – Normal ,Power – both upper limbs-proximal 4-/5distal – 3/5 hand grip - 50%(right);40%(left) both lower limbs – proximal-3/5distal-2/5 neck – 3/5 trunk <60%.**REFLEXES :**Superficial: Corneal & Conjunctival – Present, Abdominals and Cremasteric – Absent, Plantars – B/L Babinski's sign +Ve .All DTR – Biceps ++ ,Triceps ++, Supinator +, Knee ++ , Ankle +,Coordination & Gait- could not be tested.Sensory system – Normal.Cerebellar system – NormalAutonomic System – Normal.No meningeal signs.Other systems are normal.

On investigation CBP ESR,CUE,LFT, RBS, Blood urea, Sr. Creatinine, Lipid profile are normal, Sr. Na–137meq/lSr. K– 3.8meq/ lt. HIV/HbsAg – Non reactive, ECG, CXR, USG Abdomen– Normal. CT BRAIN - Ill defined hypodensity noted in central pons.MRI Brain with contrast – Ill defined altered signal intensity which is hyperintense on T2W1,FLAIR and

hypointense on T1W1 and restricted on DW1 seen in pons sparing the periphery S/o CENTRAL PONTINE MYELINOLYSIS .The patient was given CAT I ATT ,supportive treatment along with Inj.Multivitamin.Advised CAT

1 ATT for 6 months and multivitamin tablets at discharge along with physiotherapy & speech therapy. On follow up-Patient improved after 4weeks and became normal after 12 weeks.



III. DISCUSSION

Demyelinating disease of the brain has 3 subtypes

CPM - lesion is confined to pons. Extra Pontine Myelinolysis - lesions confined to the basal ganglia, cerebrum and cerebellum. ODS - CPM and EPM lesions are both present.

Central Pontine Myelinolysis(CPM) and Osmotic Demyelinating Syndromes(ODS) are rare medical

conditions. Exact incidence – unknown; derived primarily from autopsy series.

It is seen in all age groups affecting both males and females equally¹.

In 1976 Tomlinson and colleagues suggested that the rapid correction of serum sodium in hyponatremic patients was the causative factor⁸. Since then several cases of CPM with normonatremia particularly in alcoholics have been reported. CPM has been traditionally associated with rapid correction of hyponatremia although this is not the cause in all cases.

Chronic Alcoholism, Chronic malnutrition, Chronic debilitating conditions, Liver transplantation, Burns, AIDS, Hyperemesis gravidarum, CRF on dialysis, Cancer are risk factors^{3,4}.

The predilection of the myelinolysis to pons is thought to be the result of the grid arrangement of the oligodendrocytes in the base of the pons which limits their mechanical flexibility and therefore their capacity to swell. During hyponatremia these cells can only adapt by losing more ions instead of swelling making them prone to damage when sodium is replaced^{5,6}. Proximity to the extensively vascularized grey matter makes the pons particularly susceptible to damage caused by vasogenic edema and myelinotoxic substances from the vessels¹.

Microscopically - Destruction of myelinated sheaths with relative sparing of axons & neurons in pontine nuclei in contrast to pontine infarct and No inflammation in contrast to Multiple sclerosis noted^{7,8,9}.

Clinical features are highly variable

Typically a rapidly evolving paraparesis or quadriparesis and pseudobulbar symptoms such as dysarthria and dysphagia (most consistent), exaggerated jaw jerk and spastic tongue⁹. Horizontal gaze paralysis, Vertical ophthalmoparesis - demyelination extending through the mid brain. If the lesion extends into the tegmentum of the pons, pupillary and oculomotor abnormalities may occur. Ataxia, movement disorders or behavioral symptoms, Locked in syndrome may be seen. Coma or delirium occur if lesions in the pontine tegmentum and thalamus^{5,6}.

EPM - Postural limb tremors, myoclonic jerks, parkinsonian picture, catatonia, dystonia, or pyramidal dysfunction. These may resolve completely or partially over months or they can become permanent.

Diagnosis of CPM is based on clinical suspicion and is confirmed by MRI (positive 1-2 weeks post-onset of symptoms). Hypointense lesions on T1 weighted images, Hyperintense single central symmetric midline pons lesions on T2 with sparing of ventrolateral pons. Sparing of the ventrolateral pons, tegmentum, and corticospinal tracts results in the characteristic “trident-shaped” or “bat-winged” appearance^{5,6}. The lesions are non-contrast enhancing.

Recognising the patient at risk and preventing rapid correction of hyponatremia especially chronic severe hyponatremia. The rate of increase of serum sodium be no more than 1-2 mEq/L/hr during the first few hours and no more than 12 mEq/L during the first 24 hours¹.

The following treatment modalities have been reported:

Thyrotropin releasing hormone, Methylprednisolone, Plasmapheresis, Immunoglobulins^{6,7,8}. The exact mechanism of action of TRH, corticosteroids and plasmapheresis is unknown.

A conservative approach with treatment of the precipitating or underlying conditions and appropriate supportive care should be given.

Most of the patients survive and of the survivors, approximately 1/3rd recover; 1/3rd are disabled but are able to live independently; 1/3rd are severely disabled^{4,5}. Permanent disabilities range from minor tremors and ataxia to signs of severe brain damage, such as spastic quadriparesis and locked-in syndrome.

In summary, CPM or EPM are devastating and often preventable conditions, with considerable morbidity and/or mortality. Prevention is certainly the key, as current treatment only rarely leads to full recovery. Although imaging advances and a broader neuropathological understanding have improved diagnosis in the last few decades, CPM remains a condition for neuropsychiatrists to be aware of, particularly in view of the multiple neuropsychiatric medication classes in use and the psychiatric illnesses.

The patient in this case was a chronic alcoholic with no electrolyte abnormalities and improved totally with symptomatic treatment.

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