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Case report

BILATERAL INTERNUCLEAR OPHTHALMOPLEGIA AS FIRST MANIFESTATION OF EXTRA PONTINE MYELINOLYSIS

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ABSTRACT

Extrapontine myelinolysis (EPM) is a rare clinical entity affecting anterior basal ganglia. This is one of the osmotic demyelination syndromes. It occurs due to rapid correction of hyponatremia and also rarely occurs in alcoholics. It generally presents with extrapyramidal symptoms. We here report a case of EPM in a 13 year old boy presenting with bilateral internuclear ophthalmoplegia and ptosis. The patient also had generalised weakness, but no psychiatric symptoms. The patient slowly recovered over six months. EPM can affect any age group, although the elderly are more likely to be affected due to frequent electrolyte abnormalities. Ocular movement disorders or brainstem signs are rarely reported in EPM. When present, it can create diagnostic confusion with multiple sclerosis. We believe this is the first report of this entity from India. The relevant literature regarding brainstem manifestations in myelinolysis syndromes is also discussed, along with the radiological findings.

Keywords: Internuclear ophthalmoplegia, Extrapontine myelinolysis, Ptosis, CIDP, Basal ganglia

INTRODUCTION

Extrapontine myelinolysis (EPM) is a rare clinical entity occurring mainly after rapid correction of hyponatremia. It is usually associated with its counterpart: central pontine myelinolysis (CPM).¹ However, very rarely, EPM can occur in absence of CPM and this makes the diagnosis challenging. The clinical manifestations of EPM vary and may range from extrapyramidal features to neuropsychiatric manifestations.^{1, 2} Such atypical features, along with the rarity of the entity often delay the diagnosis. We here report a case of EPM presenting with bilateral internuclear ophthalmoplegia (INO). To our knowledge, this is probably the first report of EPM presenting with INO from India. Other reported cases from India have shown parkinsonian features and bulbar symptoms.³ Another case was reported with flaccid quadraparesis.⁴

CASE REPORT

A 13 year old boy presented with acute onset generalised weakness without loss of consciousness for two days. He had been admitted elsewhere with increasing abdominal pain and vomiting for twenty days. He was there documented to be dehydrated and resuscitated with intravenous fluids. He apparently improved with the conservative management but deteriorated again with severe generalized weakness and blurring of vision. With this complaint, he was referred to our tertiary care center.

At our centre, on admission, the boy was found to be severely weak with power 2-/5 in all four limbs. He could not turn in bed or lift his head from pillows. His abdomen was found to be distended and his parents complained of severe constipation for the last ten

days. Immediate straight X-ray of abdomen (Fig. 1) Showed air fluid levels consistent with intestinal obstruction.

Further examination revealed bilateral ptosis (fig. 2) with bilateral internuclear ophthalmoplegia. Pupillary reactions were normal and there was no weakness of any other cranial nerve. Ophthalmoscopy was normal. The blurring of vision was probably due to nystagmus on lateral gaze. The deep tendon jerks were all depressed and plantar response was absent. There was generalised hypotonia. There was no muscle tenderness or nerve thickening. Higher functions remained normal throughout. The pulse rate was 120/min with loss of respiratory variation and there was marked postural hypotension (fall of SBP by 35 mm of Hg on sitting).

Past history revealed recurrent episodes of similar abdominal distension and constipation over three years. However, each time, he had responded to conservative management. He had no history of abdominal surgery or tuberculosis. Past CT scans of abdomen were normal. Also, he had three episodes of generalized weakness lasting for one to two months over past three years. In one such episode, he was investigated in detail and diagnosed as Acute Inflammatory Demyelinating Polyneuropathy. However, he was lost to follow up after that. His parents said that he had some residual weakness of the limbs from that episode and needed support while walking.

Laboratory examinations revealed hemoglobin of 9.9G/dl with total leukocyte count of 7100/cmm (Neutrophil 67% and lymphocyte 28%). The Platelet count was 1.9 lakhs/mm³ with normal red cell indices and normal ESR. Blood sugar 108mg/dl, /urea 31mg/dlcreatinine was 0.7 mg/dl respectively. Liver function test was normal and blood electrolytes revealed Na 135 mEq/L and K 4.2 mEq/L. Serum calcium was also normal. After admission, the generalised power improved to 3/5 but the INO persisted. A CT scan of brain was normal. CSF study revealed 8 cells/cmm with protein of >2g/dl and high globulins. CSF ACE level was normal and TB-PCR done from CSF was negative. Also, CSF VDRL was negative. Blood for HIV, Hepatitis B, C and Herpes Simplex serologies were negative. A nerve conduction study was done which revealed decreased amplitudes of mainly motor nerves in all four limbs with relatively normal conduction velocity,

suggestive of axonal degeneration. Also, needle EMG revealed spontaneous fibrillation, suggestive of denervation. This picture, along with the CSF report was suggestive of Chronic Inflammatory Demyelinating Polyneuropathy. This could also explain the autonomic dysfunction as manifested in cardiovascular examination. Probably the intestinal obstruction was a manifestation of autonomic involvement in CIDP. Repeat ultrasonography of abdomen and barium meal study did not reveal any mechanical obstruction.

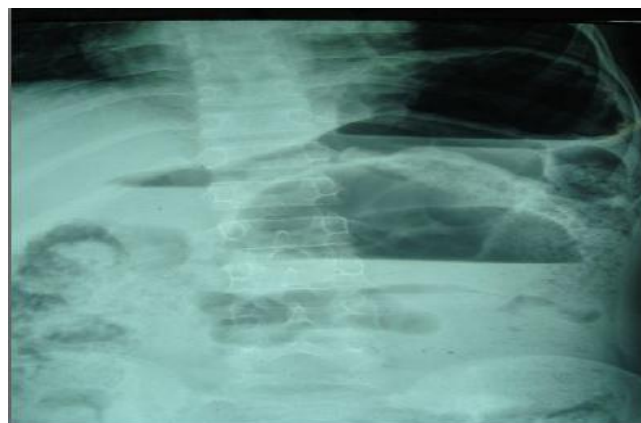


Fig 1: straight X ray abdomen showing multiple air fluid levels



Fig 2: Face of the patient with bilateral ptosis (Photo was taken with consent of patient)

Chest X ray was normal. MRI scan of brain was done which revealed symmetric marked hyperintensity in T2 images in anterior part of basal ganglia involving putamen and anterior part of caudate nucleus (Fig. 3). Also there was some hyperintensity in tegmental part of midbrain involving periaqueductal grey matter (Fig. 4). However, the T1 images were completely normal and coronal section of pons in T2 imaging did not reveal any signal changes also. MRI spectroscopy was done, but was reported to be essentially normal. Blood lead levels and porphyrin levels

(Porphobilinogen and delta ALA) were normal. Serum magnesium, thyroid function tests and vitamin B12 levels were also normal.

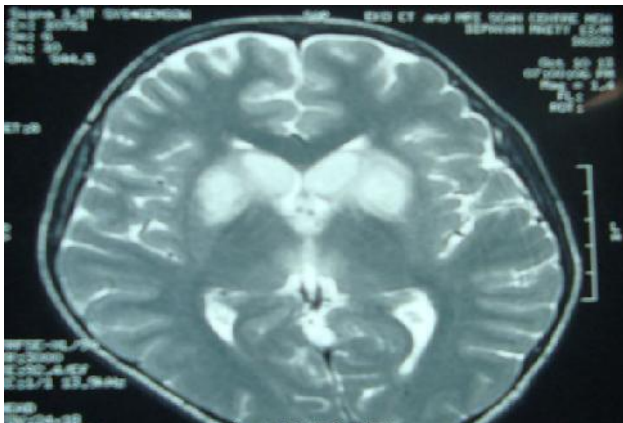


Fig 3: T2 weighted MRI images showing bilateral anterior basal ganglia hyperintensity



Fig 4: FLAIR image of midbrain showing the lesion (red arrow)

Thus, based on the imaging findings, the case was diagnosed as extra pontine myelinolysis probably due to overzealous correction of hyponatremia (normal 135–145 mEq/L.) in dehydration in the background of CIDP with autonomic features.

The patient was treated with physiotherapy and braces. At six months follow up, his INO had improved. Repeat MRI showed resolution of the lesions. Also, he had not developed any Parkinsonian features. However, the power in his limbs remains 4-/5 and he can now walk only with support.

DISCUSSION

EPM is a rare entity occurring mainly after rapid correction of hyponatremia.⁵ Thus, this can occur in disease states like renal failure, diarrhea, diuretic abuse, heart failure, vomiting and salt losing states. It is due to osmotic damage to brain tissues which occurs due to rapid shift of osmotically active particles across neuronal cell membranes. CPM and

EPM can also occur rarely in chronic alcoholics or malnourished persons.⁶ In our patient, the hyponatremia was never documented, but since the patient had a prolonged history of vomiting with intestinal obstruction, this was probably the most likely underlying abnormality. Our patient had the typical feature of early improvement followed by sudden deterioration, which is found in osmotic demyelination syndromes. In EPM, the manifestations can vary from extrapyramidal features like tremor or dystonia to psychiatric illness.^{2, 5} Even quadriplegia has been reported in this disease.

INO is a manifestation of brain stem dysfunction at the level of medial longitudinal fasciculus. The chief causes of INO are multiple sclerosis and brain stem infarction, although it can also be seen rarely in any local tumour or congenital malformation.⁷

In CPM, brain stem features like nystagmus and gaze palsy are reported.⁸ This is due to edema in pons and its connections with the cerebellum. Also, presence of nystagmus in CPM, especially in an alcoholic patient, should prompt a search for coexisting Wernicke's encephalopathy (WE).⁹ In these cases, MRI imaging of brain can help in differentiation. In CPM, we get trident shaped lesion in T2 image in pons on sagittal section. In WE, there will be additional hyperintense lesions in FLAIR and T2 in bilateral thalami.⁹ Nystagmus has very rarely been reported in EPM. One case report from Denver showed a patient of EPM with medullary lesions, presenting with downbeat nystagmus.¹⁰ Like our case, this case also had resolution of brain stem symptoms with time. Like CPM, the prognosis of EPM is variable. Some patients recover completely while others may have residual motor, psychiatric or memory related dysfunctions. The mortality rate has decreased now with early diagnosis.

INO is almost never reported with CPM or EPM. Sometimes, a patient with INO is first thought to have CPM, but later new features emerge and a diagnosis of multiple sclerosis is made. Thus, in a patient presumed to have CPM, the presence of INO should alert the clinician to the possibility of multiple sclerosis. However, in our case, the presence of clear basal ganglia lesions in MRI was conclusive. Literature search revealed only one other reported case of EPM with presence of INO.¹¹ In that case, there was also gaze palsy with gaze evoked rotatory nystagmus.¹¹ However, due to rarity of EPM, the

ocular movement disorders in this disease have not been well studied.

In INO, the lesions are usually found in paramedian pontine tegmentum or periaqueductal region.¹² In our patient, the lesions in periaqueductal midbrain in MRI accounted for the INO. In EPM, the typical MRI features include symmetrical bilateral hyperintensity in T2/FLAIR in putamen and caudate nucleus with relative sparing of globus pallidus.¹ Also T1 images in these areas will be normal and this helps to differentiate this condition from similar presentations with CO poisoning. The diagnosis of EPM is mainly clinical with added MRI findings.

Our patient recovered slowly over time. The MRI lesions also resolved. This temporal profile of EPM was also documented in other reported cases.^{8, 10} However, since our patient had underlying CIDP, he did not regain full power of the limbs.

This is probably the second reported case of EPM with INO. This case depicts the possible varied presentation of osmotic demyelination syndromes with brain stem signs.

CONCLUSION

Central nervous system osmotic demyelination is a rare complication of electrolyte correction. It may present with atypical features like ocular movement disorders. Thus, clinicians should have a low threshold for brain imaging if atypical neurological signs appear in a patient of hyponatremia.

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Conflict of interest: Nil

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