

Rare association of central pontine myelinolysis with infantile tremor syndrome

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Abstract

Central pontine myelinolysis (CPM) is an acute demyelination within the central basis pontis. Though exact mechanism is not known it is seen commonly with rapid correction of hyponatremia and also with pontine ischemia or infarction, demyelinating diseases, pontine neoplasm and different metabolic diseases. We report a rare association of CPM in a patient of Infantile Tremor Syndrome (ITS). ITS is a syndrome of tremor, mental and physical retardation, pigmentary changes of hair and skin and anemia in malnourished children. Though first reported in Indian subcontinent many identical cases were reported from around the world. Our case is a 15 month old child with generalized tremor, mild hepatosplenomegaly with features of grade II malnutrition including skin and hair changes. All the signs and symptoms of tremor improved after treatment with the World Health Organization (WHO) protocol for protein energy malnutrition (PEM) and administration of propranolol without any side effects.

Key Words

Central pontine myelinolysis, infantile tremor syndrome, malnutrition

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Introduction

Infantile tremor syndrome (ITS) is a clinical syndrome of acute or gradual onset of mental and psychomotor changes, pigmentary disturbances of hair and skin, pallor, and tremor in malnourished children aged between 5 months and 3 years.^[1] Though first reported in Indian subcontinent many identical cases were reported from around the world.^[2,3] Clinically the presence of tremor has been attributed to structural and functional alterations of extrapyramidal system but routine neuroimaging studies with CT scan and MRI in past revealed non specific structural changes in ITS.^[4]

Here we report a classical case of ITS with grade II malnutrition with a rare association of central pontine myelinolysis (CPM) in MRI. So far as our knowledge this is the first reported case of such an association.

Case Report

A 15 month old male child born normally at full term to non-consanguineous parents of low socio-economic status was admitted in pediatric ward with complaints of continuous and generalized tremor involving all limbs and head for 2 days. The baby had grade II malnutrition. He had hyperpigmentation specially over hands, feet, and moderate pallor with mild hepatosplenomegaly. He had recent hair-cut for graying and fall of hair. His tremor was generalized, coarse, fast, and of low amplitude. Tremor disappeared during sleep but aggravated during activities. Tremor was disturbing his normal activities of sitting, walking, and feeding. The baby had history of motor delay but no mental delay and he took interest in surroundings. His other neurological examination findings were normal.

Investigation

Routine blood revealed pallor (Hb - 6.8 gm/dl) with microcytic hypochromic anemia. Chest X-ray revealed mild hilar infective changes. USG abdomen showed hepatosplenomegaly without ascites. Slides for malaria parasite and dual antigen were negative. The Mantoux test was negative. Serum urea, creatinine, and other biochemical parameters (serum Na, K, Ca) were within normal limits. MRI showed a typical hyperintense lesion in T2-weighted image of central pons with sparing of periphery of pons consistent with CPM [Figure 1]. EEG was normal. The cerebrospinal fluid (CSF) study was normal. Serum vitaminB12 and mineral (Mg and Zn) levels were within normal limits.

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Treatment and follow-Up

The baby was treated according to the WHO protocol for malnutrition, vitamin (A, D, B-complex, and C) and mineral (Zn, Mg, Ca) supplementation, antibiotics, and propranolol for 2 weeks. Propranolol was started at a dose of 0.5 mg/kg/day 2 days after starting nutritional therapy as there was no considerable improvement of tremor. Propranolol was gradually increased up to 2 mg/kg/day. During this period tremor decreased in intensity and general condition of the baby improved substantially. The baby was discharged with above medications and iron and followed-up for 2 weeks with disappearance of tremor without any side effects of propranolol.

Discussion

The causation of ITS is of much debate for ages. Malnutrition, vitamin and mineral deficiency (e.g., Mg and Zn), infections, toxins, degenerative brain disease, enzyme defects (e.g., tyrosine) all have been postulated as the causation of ITS.^[1]

But neuroimaging in the case of ITS is a very little studied subject. The presence of gyral atrophy and mild ventricular dilatations on CT scans were described previously in the literature. Thora *et al.* presented a series of eight cases of ITS with cranial neuroimaging (CT scan and MRI) and found similar results. Interestingly one MRI scan showed hyperintense signals in frontal and periventricular white matter on T2-weighted images which were attributed to demyelination due to viral infection or acute disseminated encephalomyelitis (ADEM).^[4] In the MRI study of our case of ITS we found typical hyperintense lesion in central pons with sparing of the periphery of pons suggestive of CPM [Figure 1: Axial T2-weighted MR image].

CPM has recently been described as osmotic demyelination syndrome (ODMS) as it commonly occurs in rapid correction of hyponatremia. It has also been reported in pontine ischemia or infarction, demyelinating diseases, pontine neoplasm (like astrocytoma or metastasis), and different metabolic diseases (like Wilson, Leigh, diabetes and hypertensive encephalopathy).^[5] CPM is also seen in hyponatremic patients with vomiting, diarrhea, diuretic therapy, postoperative overhydration, psychogenic water intoxication, and severe malnutrition (sometimes with extreme burn injuries).^[6] Along with hyponatremia other electrolyte disturbances like hypophosphatemia and hypokalemia have been reported.

Similarly various vitamin and mineral deficiencies have also been reported in association with ITS.

Kahn (1954) described tremor syndrome with reference to extrapyramidal nervous system disorder in pellagrins (Lewy *et al.*, 1940). According to him this syndrome was only observed during convalescence from malnutrition (particularly with kwashiorkor) due to rehabilitation with food which has an imbalance of high protein and low vitamin content.^[7]

Garewal *et al.* (1988) described ITS as a vitamin B12 deficiency

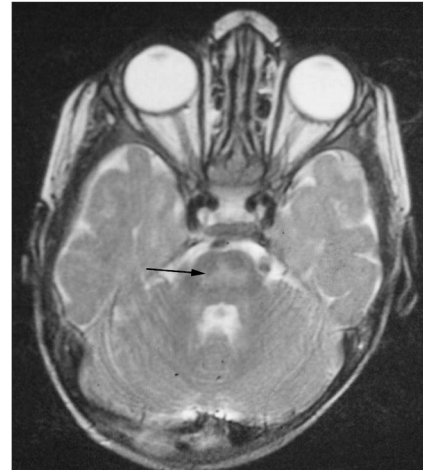


Figure 1: Axial T2-weighted MR of brain demonstrates hyperintense signal of central pons with peripheral sparing

syndrome in infants. He postulated the low levels of vitamin B12 and its transport protein TCII in the CSF may be responsible for the neurologic features of the syndrome.^[8]

Edema and various electrolyte abnormalities like hyponatremia, hypokalemia, and hypophosphatemia (specially during refeeding), etc. are known to occur in ITS/PEM (protein energy malnutrition).^[9]

We have observed the unique association of CPM with ITS for the first time and further studies would be required to establish the relationship.

Conclusion

The treatment of ITS being largely empirical along with nutritional supplements various drugs like anticonvulsants, antiparkinsonian drugs, beta blockers have been tried in the past. In our case we found a favorable response with propranolol. However further studies are required to find the exact cause of this disorder and establish the association of CPM with ITS.

Already known: Cranial neuroimaging using CT and MRI in the case of the infantile tremor syndrome reveals only non-specific structural changes.
 What is new: Neuroimaging using MRI in the cases of ITS may show central pontine myelinolysis.

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