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Case Report of A Infantile Tremor Syndrome in A 9 Months old Girl With Severe Nutritional Deficiencies.

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Abstract:*Infantile tremor syndrome (ITS) is a self limiting clinical disorder. This disorder commonly affects infants and young children. This disorder seems to affect children of African and SoutheastAsian countries with poor socio-economic conditions. It's an uncommon condition and in India it accounts for 0.2% to 2% of hospital admission. This syndrome presents with mental and psychomotor changes associated with tremors, pigmentary changes of skin and hair along with regression of milestones. Many etiological theories have been put forth but the most commonly accepted theory is nutritional theory. Here we discuss such similar findings in a 9 months old girl child who was suffering from severe Vit B12 deficiency and malnutrition. Apart from the above mentioned signs the child was also found to be floppy and hypotonic. Computed tomography showed generalized cerebral atrophy. The child showed significant improvement with intensive therapy for correction of malnutrition and Anaemia.*

I. Introduction

Infantile tremor syndrome (ITS) is a self limiting clinical disorder in infants and young children, seen most commonly in Indian subcontinent, Southeast Asia and African countries. It's a disorder seen in developing countries with poor socio-economic conditions. It is a syndrome of acute or gradual onset of mental and psychomotor changes, pigmentary disturbances of hair and skin, pallor, tremors and regression of developmental milestones, beginning during later part of infancy in malnourished children aged between 5 months and 3 years [1].Improvement in nutritional status, living conditions and better weaning practices could explain the reducing incidence rates over the years and is now less frequently encountered. In India, it accounts for 0.2 to 2 % of paediatric hospital admissions (1-2% in 1960s, 1.1 % in 1975-77 and 0.2% in mid-1990s) [2]. 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 [3] The etiology of ITS is still elusive. Among various theories, nutritional theory is the most accepted. The other etiologic possibilities being VitaminB12, Mg, Zn, deficiency, Infections and toxin related. Here we report a case of 9 months old baby who presented to us with ITS associated with malnutrition showed cerebral atrophy on Neuroimaging. The child had all the classical symptoms of ITS with severe deficiency of VitB12.

II. Case report

A 9 months old baby girl who was of vertex presentation born at full term to healthy parents with non-consanguineous marriage. The child presented to our emergency department with tremors of head and limbs along with being floppy. Mother also complained of regression of milestones and child being listless. Mother also reported the child having increased pigmentation of skin along with discoloration of hair.

On clinical examination the baby was listless. The baby looked plump and weighed 4.5kg (expected 9 kg for age) with grade III malnutrition. There was pallor, dark pigmentation of skin more over knuckles of hands and feet.

The hairs were sparse, brown, thin and easily pluckable. Baby had coarse tremors involving her head and limbs. The tremors were slow and of low amplitude, which disappeared during her sleep but worsened during her activities, thereby disturbing her sitting and crawling. On further examination the child was found to be floppy with hypotoniaand associated with regression of milestones. The milestones achieved were neck holding at 3 months but now head holding was unsteady, sitting with support was attained at 8 months but now sitting with support momentarily. There was no mental delay but the child showed less interest in the surroundings.

Clinical Photos:

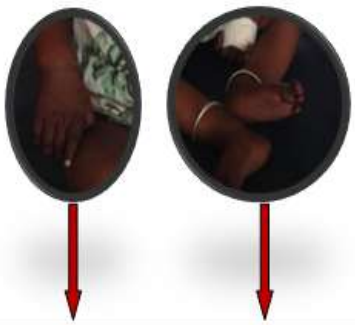
Baby at 3 months of age with neck holding

Baby appearing listless

Brown and sparse hair of the baby

Baby appearing plump on general survey

Baby requiring support to sit showing regression of motor skills



Hyperpigmentation of the skin seen classically at knuckles

Investigation

Routine blood investigations showed Hb%- 5.3 mg/dl,TLC-2600 cells/cmm, Platelet count- 4.4 lakhs, MCV – 103 fl,MCH-32 Pg ,MCHC-31 g/dl. Blood picture shows dimorphic pattern with relative lymphocytosis and Leucopenia. Bone marrow showed megaloblastic picture. Serum Vit B12 assay - 83 pg/ml (range: 198 – 883 pg/ml), Folic acid levels- 20 ng/ml (2.52 – 17.56 ng/ml). Virological studies were negative.Ultrasound abdomen was normal. CT brain showedprominent cerebral sulci,sylvian fissures and lateral ventricle more in the right cerebral hemisphere suggestive of cerebral atrophy.

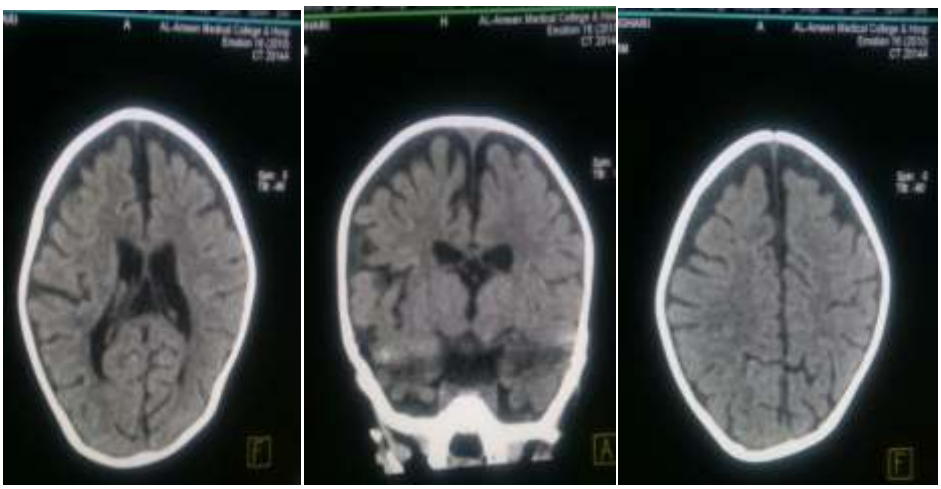


Fig: CT images showing cerebral atrophy at different

Treatment:

Once the diagnosis was confirmed the baby received treatment as per WHO protocol for malnutrition. She received VitB12, Folic acid, Mg and Zn supplements. She also received 400 ml of packed red blood cells to improve her anemia. Propranolol was started at 1 mg/kg/day and increased up to 2mg/kg/day. Tremors improved in intensity and frequency once the dose of propranolol was increased to 2mg/kg/day. The general condition of the baby improved over a period of 2 weeks of treatment. The patient was discharged home with the above medications and advised for follow up at regular intervals.

III. Discussion

A classical picture of ITS is a plump looking infant between 5 months to 3 years with the presence of malnutrition who has been exclusively breast fed. Usually, these children are listless, apathetic, and disinterested in surroundings. Scalp hair is sparse and light colored. Dark pigmentation is present over dorsal aspects of hands, nail folds, feet, knees, ankles, buttocks, and axillae. There is regression of milestones in the recent past. Tremors have an acute onset following an acute infection or stress. Initially, they are intermittent but become continuous in a few days. They are more prominent in distal parts of limbs, head, face, and tongue. These tremors disappear during sleep. There is presence of anemia, which may be macrocytic, microcytic, or normocytic. Most of these classical findings were present in our case.

The etiology of ITS is still elusive. Malnutrition, vitamin and mineral deficiency (e.g., Mg and Zn), infections, toxins, enzyme defects (e.g., tyrosine) all have been postulated as the causation of ITS[1]. Among various theories, the nutritional theory is the most accepted. Vitamin B12 deficiency has been found to be associated with ITS in many studies[1]. It is usually seen in children who are exclusively breast-fed for prolonged periods by vegetarian mothers.

ITS is essentially more of a clinical diagnosis with peripheral smear suggestive of anemia (mostly megaloblastic anemia, macrocytosis ,MCV >95). Serum Vitamin B12 levels was found to be low in most of the reported cases. Also seen was low CSF levels of Vitamin B12 along with low TC II levels.[4] Vitamin B12 levels in the mother may also be low suggesting low levels in the breast milk. Serum levels of Zn, ascorbic acid, and Mg may be low.Only a few reports of neuroimaging in ITS are available. Neuroimaging in our case had features suggestive of cerebral atrophy. Most reports have shown normal neuroimaging findings or cerebral atrophy, ventricular prominence and/or prominence of the subarachnoid space, pontinemyelinolysis, and cerebral hyperintensities.

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[5]. Garewal *et al*. (1988) described ITS as a vitamin B12 deficiency 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 [6].

Generally this condition is a self limiting disorder which gets corrected once the nutritional/ therapeutic replacement for Vit B12, Zn, Mg, Iron and protein takes place [1]. Phenobarbitone (3 – 5 mg/kg/day) or carbamazapine can be used to medically manage severe forms of tremors. Propranolol and chlorpromazine are the other drugs that can be used [7]. Over a period of time the tremors subside gradually in terms of severity and frequency to become intermittent and finally stop as the nutritional status improves.Antiparkinsonian drugs have also been tried in the past. Pigmentary changes in skin and hair take months to clear. Mental dullness and sluggishness takes years to come back to normal [8].

IV. Conclusion

ITS is no doubt less frequent but is still prevalent. Its prevalence is due to malnutrition, lack of knowledge of mothers on weaning from breast milk and poor socio economic conditions.Its a self limiting disorder but improvement in psychomotor functions is relatively slow. Prompt diagnosis and early intervention may help the child in early recovery. Mortality is never directly related to the disease but may be attributed to concurrent infections. Subnormal intelligence is the only long term sequelae.

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