Infantile Tremor Syndrome

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Abstract: Infantile tremor syndrome is a self-limiting condition usually present in infants and young children of low socioeconomic status with multiple nutritional deficiencies. It presents as developmental delay along with other symptoms. Timely diagnosis and management is necessary for better neurological growth and outcome of the children. Treatment is mainly supportive with micronutrient supplementation and nutritional rehabilitation.

Keywords: Tremors, Developmental delay, Anemia, psychomotor changes

1. Introduction

Infantile tremor syndrome (ITS) is a self-limiting condition affecting infants and young children of age between 5 months to 3 years (1). It is characterized by tremors, anaemia, pigmenitary skin changes, regression of milestone stones and hypotonia (2). It affects children of lower socioeconomic status and has been predominately reported from Indian subcontinent and South East Asian regions (1). In India, it accounts for 0.2 to 2 % of paediatric hospital admissions (3). The etiology of ITS still remains elusive. Among various theories, nutritional theory is the most accepted. Due to presence of developmental regressions and psychomotor changes, sometimes it is being misdiagnosed as seizure disorder. So, we here by present case series of 3 children of ITS who were misdiagnosed as seizure disorder.

The purpose of the case series is to create awareness among the clinicians regarding infantile tremor syndrome, whose incidence is now declining owing to better education and better feeding practices.

Case 1:

Born at term, as a product of non-consanguineous marriage by normal vaginal delivery with birth weight around 2.7kg, with no perinatal adverse events, exclusively breast fed, fully immunised, previously healthy ten months old male child presented to our Out Patient Department with history of tremors, excessive crying, delayed milestones. Prior to presenting to us she was taken to a peripheral hospital where she was started on anti-epileptics, as there was no improvement in the tremors she was brought to our OPD. There was no previous hospitalization. On examination, pallor was there with pigmentation of hair and knuckle pigmentation was present. Child was vitally stable and anthropometry was as given in table (1). There was no organomegaly, milestone stones were delayed for her age. Child was exclusively breast fed till 8 months of age and following that she was predominantly on milk based diet. Lab investigations were as in table 2. In v/o of history, physical examination and lab work up possibility of ITS was kept and she was started on vitamin D, multivitamin, iron supplementation and vitamin B12. The child started improving, as the irritability improved and oral intake improved and on subsequent visits to OPD the tremors had stopped and she was catching up on her milestones.

Case 2:

Previously healthy, fully immunised with normal anthropometrically normal parameters, eighteen months old female child was admitted with complaints of delayed milestone stones compared to her elder sibling. Antenatal period was uneventful, was delivered by vaginal delivery with no perinatal adverse events. child was exclusively breast fed till 9 months of age and complementary feeding was started thereafter which was predominantly milk based diet. On examination, child was vitally stable, pallor was present, frontal bossing was there. Knuckle pigmentation was present. There was no organomegaly and rest of the systemic examination was normal. The lab investigations and anthropometric parameters were as in the table (1, 2) respectively. While the investigations were awaited, child started having tremors during the hospital stay and possibility of ITS was kept and was managed as per guidelines and she improved. On subsequent visits child was thriving well with improvement in general well being as well as improvement in the lab parameters.

Case 3:

One-year old male child was referred to us from private practitioner as case of seizure disorder, as the child was taken to a private hospital with complaints of acute onset of tremors involving the peripheries and it was absent while in sleep and used to aggravate on crying. Developmental milestone stones were delayed for his age, other than that the period of life till date was uneventful. On examination, child was hemodynamically stable, euthermic, and euglycemic. Pallor was there, hair pigmentation was present along with frontal bossing. Rest of the systemic examination was normal. Lab work up was as given in table 2. All the antiepileptics were stopped at once and child was managed as per protocol along with

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dietary rehabilitation started. The child was discharged once there was satisfactory improvement.

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2. Discussion

Infantile tremor syndrome is a self-limiting entity most commonly reported in 5 months to 3 years of age group. It is usually seen in infants and children of lower socioeconomic strata with a slight male preponderance. Clinical signs and symptoms are variable which include pigmentary changes of skin and hair, psychomotor changes and tremors. The hair changes include pigmentation along with sparse hair. Dark pigmentation is present over dorsal aspects of hands, nail folds, feet, knees, ankles, buttocks and axillae. There is regression of mile stones or there may be delay in achievement of the milestones. Usually the presenting complaint is acute onset of tremors which may be aggravated by stress or infection. Tremors are intermittent to begin with and then become continuous during the course of the illness. They are more prominent in distal part of limbs, head face and tongue. They get worse while the child is crying and disappear during sleep. Most of the classical findings were present in our cases.

The aetiology of ITS is still not clear. Of the various theories nutritional theory is the most accepted. Vitamin B12 deficiency has been found to be associated with ITS in many studies (4). It is mostly seen in prolonged breast fed babies (5) of vitamin B12 deficient mothers. The low levels of vitamin B12 and its transport protein Transcobalamin II (TC II) in the cerebrospinal fluid (CSF) may be responsible for the neurological features of this syndrome (6). Other micronutrient deficiency like Iron, Zinc, Magnesium, vitamin A, D, C, B complex has also been directly or indirectly implicated in ITS. Other speculations for its etiology include viral encephalitis and degenerative processes.

ITS is essentially a clinical diagnosis although there may be anemia (usually megaloblastic) with peripheral smear suggestive of macrocytosis, hypersegmented neutrophils along with megaloblastic bone marrow. Vitamin B12 levels may be low in serum and CSF, along with low Transcobalamin II in CSF (7). Vitamin B12 levels in the mother may also be low suggesting low levels in the breast milk. Serum levels of zinc, ascorbic acid and magnesium may be low.

Treatment regimen consists of therapy for anemia and for other nutritional deficits. Vitamin B12 injections if serum vitamin B12 levels are low. Multivitamins, vitamin C, iron, protein, zinc and magnesium supplements may also be necessary. Along with nutrient supplementation nutritional rehabilitation of the patient and counseling of the parents is also of utmost importance. If the tremors are severe, phenobarbitone (3-5 mg/kg/day) may be required to decrease the intensity (8). The tremors subside slowly over months, initially in decrease in tremor frequency and then they stop. Propranolol and chlorpromazine may also be used to control seizures. Pigmentary changes in skin and hair take months to clear. Mental dullness and sluggishness may take years to come back to normal.

3. Conclusion

To summarize, infantile tremor syndrome still remains under diagnosed and under reported. Although the prevalence has come down owing to better education of parents and good feeding practices in the developing countries. But infantile tremor syndrome needs to be considered high up in the differentials in a young child presenting with malnutrition, developmental delay, tremors and typical cry.

References
