Indian Journal of Trauma and Emergency Pediatrics
Volume 6 Number 1-2, January - June 2014

Infantile Tremor Syndrome

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Abstract

Infantile Tremor syndrome is a peculiar condition most commonly seen in Indian subcontinent. Although the number of cases with this syndrome reduced in recent years, finding of additional features made us to present this series. Mean age of presentation is generally 11.2 months (range 6 to 18 months). The male to female ratio was 5: 6. There is no seasonal variation. Tremors, pigmentation, delayed milestones, pallor and hepatomegaly was present in all cases. The mean duration of tremor phase was 23.2days with range 5 to 44 days. There was no mortality

Our case is 1year infant admitted to us for bronchopneumonia with grade II malnutrition with tremors and peripheral smear showing anemia with CT finding of cortical atrophy

Keywords: Infantile Tremor syndrome; Anemia, Pigmentary skin lesions; Regression of mental development

Introduction

Various names given to this condition are "*Infantile meningoencephalitis syndrome*", "*Syndrome of tremors in infants*", "*Vitamin B12 deficiency syndrome*", "*Syndrome of mental regression, tremors and anemia*"[1]

Infantile tremor syndrome is a clinical state characterized by tremors, anemia, pigmentary skin lesions, and regression of mental development and hypotonia of muscle.

Exact incidence is not known. 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]. Improvement in nutritional status, living conditions and better weaning practices could explain the reducing incidence rates over the years. It has been primarily reported from India and South East Asia and has also been reported from other developing countries in Asia and Africa [3]. Various nutrient deficiencies (e.g. Vit B12, Magnesium, Zinc, Vit C etc.) have been found to be associated with ITS. Other causative theories include viral encephalitis and degenerative processes.

Case Report

A 1 year infant admitted to us with complaints of Fever, Cough and vomiting since 8 days with respiratory distress with coarse tremors involving limb, head and tongue since with pigmentary changes on skin. Child is 2nd product non consanguineous marriage, 1st being female of 3 years of age without any involuntary movements or pigmentary changes. Past history and birth history are not significant with delayed development.

On general examination, anthropometry suggests Grade II malnutrition by IAP grading and under nutrition by Waterlow classification. On head to toe examination

Child has dull face (Fig.1), dry skin with pigmentary changes (Fig.2) and brittle hairs. Systemic examination reveals moderate hepatomegaly and mild spenomegaly.

On investigation

- Hb: 4.4 gm%
- TLC: 1800/cmm; N=37 L=59 M=0 E=4
- Platelet count: 1.5 lakhs/cmm

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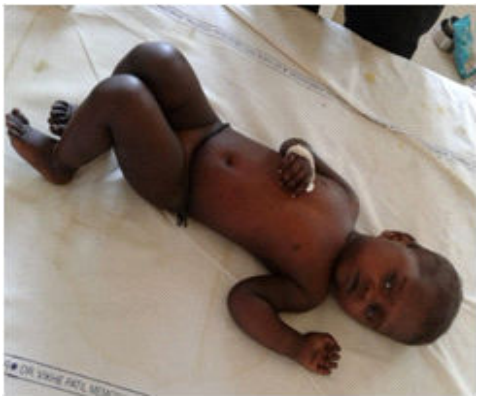

Fig. 1: Dull facies

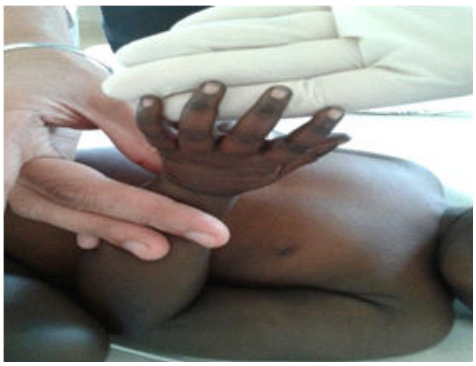

Fig. 2: Pigmentary changes

- X-Ray Chest: prominent bronchovascular markings
- CT scan s/o cerebral cortical atrophy

Discussion

Infantile tremor syndrome (ITS) is a clinical disorder characterized by coarse tremors, anemia and regression of motor and mental milestones in children around 1 Year of age. It has been primarily reported from India and south East Asia.[4]

Etiology

Vitamin B12 deficiency has been found to be associated with ITS. However iron deficiency, magnesium deficiency and zinc deficiency have also been postulated. It is seen in children who are on prolonged exclusive breast-feeding with mothers who are predominantly vegetarians [5]

Clinical Features

A classical picture of ITS is a plump looking infant between 6 months to 18 months with presence of malnutrition. These children are listless, apathetic and disinterested in surroundings[6]. 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. Hypotonia with flabbiness of muscles is common. There is presence of anemia, which may be macrocytic, microcytic or even normocytic.[7]

Diagnosis

It is essentially a clinical diagnosis with peripheral smear suggestive of anemia. Vitamin B12 levels may be low. Vitamin B12 levels in the mother may also be low suggesting low levels in the breast milk.[8]

Treatment

Treatment consists of therapy for anemia and nutritional deficiency. Vitamin B12 in high doses may be required if B12levels are low. Zinc and magnesium supplements may also be necessary. If the tremors are severe, phenobarbitone (3-5mg/kg/day) may be required to decrease the intensity. The tremors subside slowly. Initially there is gradual reduction in the amplitude and severity then the tremors become intermittent and finally stop. Pigmentary changes in skin and hair take months to clear. Mental dullness and sluggishness takes years to come back to normal.[9]

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