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Case Report

Infantile tremor syndrome

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ABSTRACT

Infantile tremor syndrome (ITS) is defined by the tetrad of pallor, developmental delay/regression, skin pigmentation, and brown scanty scalp hair. Involuntary movements in the form of tremors supervene in the natural course of the illness in a significant number of cases. The disorder is common in exclusively breast-fed infants of vegetarian mothers. Most of the children eventually recover but are frequently left with long-term cognitive and language neurodeficits. This review attempts to provide comprehensive and up-to-date information on the subject incorporating recently published studies.

Keywords: Acrodermatitis, Infantile tremor syndrome, Vitamin B_{12} deficiency

INTRODUCTION

Infantile tremor syndrome is characterised by pallor, developmental regression, tremors, hyperpigmentation of skin, and sparse brown hair.[1] It has been reported in children between 5 months and 3 years of age with a male predominance.[2] Vitamin B_{12} deficiency has been found to be associated with ITS in many studies. It is usually seen in children who are exclusively breast fed.[2] In India it accounts for 0.2 to 2% pediatric hospital admissions in 1960 and currently 0.2%.[3]

Improvement in nutritional status, living condition and better weaning practices could explain the reducing incidence rate over years. Mostly reported from India and south east asia.[4] Nutrient deficiencies like Vitamin B_{12}, Zinc, Magnesium have been found to be associated with ITS. Immerslund grasbeck syndrome has similar features like ITS, it is also associated with Vitamin B_{12} deficiency. In ITS child will not have persistent proteinuria whereas immmerslund grasbeck syndrome will have persistent proteinuria. The affected children will have regression of motor and mental milestones in children of around one year of age. The purpose of this case report is to determine the factors responsible for continuing prevalence.

CASE REPORT

A 9-month-old term baby, born normally at term, on exclusive breast feeding and fully immunized was brought with history of fever, cough and cold for seven days with increased work of breathing for two days. On examination child had pallor with apathetic look, knuckle pigmentation, hypopigmented sparse hair and perioral pigmentation, honeycomb or reticulate pigmentation of the skin over limbs was noticed. His weight was 5.4 kg and height was 67 cm.

On systemic examination child had bilateral wheeze. On central nervous system child was found to be irritable with mild developmental delay of milestones. cardiovascular and abdominal examination done was normal. Child required oxygen support for three days then gradually tapered oxygen and stopped. Investigations done showed severe macrocytic

hypochromic anemia [HB-6.9], [MCV-109.2] with peripheral smear showing macrocytic anemia with lymphocytosis.

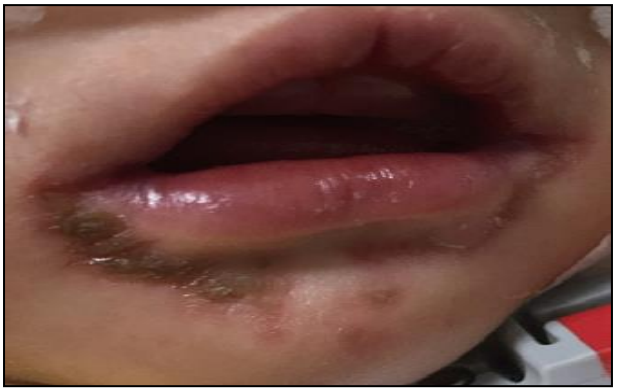

Figure 1: Acrodermatitis enteropathica before treatment.

Urine routine done showed proteinuria [1+]. Further workup done for anemia which showed normal serum iron, TIBC and ferritin. Had low B_{12} and folic acid of 10.9. Dermatology opinion obtained for skin rashes which was suggestive of acrodermatitis enteropathica and started on therapeutic dose of Zinc.

Hematology opinion for megaloblastic anemia obtained and advised to give IV Vitamin B_{12} stat and T. Folic Acid. Mother CBC was tested and showed evidence of iron deficiency anemia, she is a vegetarian, put on iron, folic acid and B_{12} tablets and was dewormed. At the time of discharge rashes disappeared and child was active, playful.

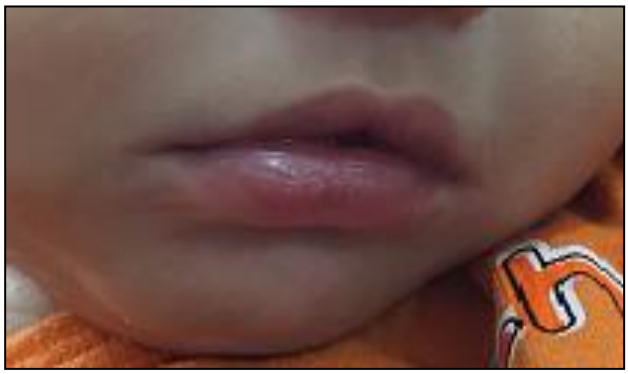

Figure 2: Acrodermatitis enteropathica after treatment.

DISCUSSION

Infants with ITS are born without any prenatal or perinatal complications and display normal neurodevelopment till the first 4-6 months of age.[5] Developmental slowing then sets in, and is followed by developmental regression, if corrective measures are not instituted.

The low levels of vitamin B_{12} and its transport protein Transcobalamin II in CSF is responsible for neurological features of this syndrome.[6] It has been observed in earlier studies the presence of LRTI, exact reason for this high incidence of LRTI is not known but may be due to presence of less antioxidants due to deficiency of Vitamin C.[7]

Infantile tremor syndrome cases always present with various vitamins deficiency and varying degree of anemia.[8] It is most commonly found in children belonging to poor socioeconomic status and who are on exclusive breast feeding of vegetarian mother. Low Vitamin B12 levels in the mother may also be low in the breast milk. Treatment of nutritional deficiency includes vitamin B_{12}, multivitamins, folic acid, iron, calcium, zinc, magnesium and high protein diet.[3]

It is easy to understand that infants with vitamin B_{12} deficiency can have predominantly hematological (megaloblastic anemia) or predominantly neurological (ITS) manifestations.[9]

To conclude, ITS is a nutritional deficiency syndrome presenting predominantly with neurological manifestations. A variable number of these children has concomitant hematological findings consistent with megaloblastic anemia. Laboratory evidence of vitamin B_{12} deficiency is found whenever adequate investigations are undertaken.

Treatment with vitamin B_{12} produces results in rapid amelioration of symptoms. Treatment of other associated nutritional deficiencies and correction of feeding practices is necessary for comprehensive management of these infants. Since long-term cognitive and language impairments can occur, early diagnosis and treatment are necessary.

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