

A Rare Case Presentation of Infantile Tremor Syndrome

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Abstract

Introduction: 9 month old girl of an Asian origin admitted with abnormal movements of the left side of the body of 10 days of duration prior to admission disappearing in sleep with disinterest in surroundings with moderate fever of 15 days of duration. Birth history was unremarkable. She was exclusively breast fed baby. Mother was purely vegetarian. There was gross motor delay in the form of not able to sit without support. Neuroregression was noted in the form of loss of head control and social smile after starting of the abnormal movements. She was malnourished with weight of 6.0 kilograms. She was noted to have pallor, thin sparse hair with knuckle hyper pigmentation Neurological examination revealed hypotonia with unilateral tremor of the left side of the body with twitching of the left side of the face.

Material and Methods: Her complete blood work up including metabolic investigations, vitamin B12 and folic acid assay was negative. Urine homocysteine was positive. MRI Brain showed generalized atrophy. Child was started on injection Vitamin B12 in high doses.

Result: Tremors gradually reduced and completely stopped after 3 weeks of starting of the treatment. Child regained the social smile and head control. She started to sit without support.

Discussion: Infantile tremor syndrome is a rare entity which can be presented from 6 months to 18 months of age with neuroregression, malnutrition and acute tremor which can be attributed to the structural and functional alteration of extra pyramidal system. Though Vitamin B12 blood levels were normal, there was deficiency at tissue level so she was treated successfully with Vitamin B12 in high doses. If recognized early then neurological consequences can be completely reversible with appropriate treatment.

Conclusion: Infantile Tremor Syndrome needs to be considered in a young child with tremors, malnutrition, developmental delay and neuroregression.

Keywords: Infantile Tremor Syndrome; Vegan Diet; Malnourished; Abnormal Movements

Introduction

Infantile tremor syndrome (ITS) is a rare clinical disorder characterized by coarse tremors, anaemia and regression of motor and mental milestones in children of around one year of age. If recognized early, then neurological sequences can be reversible.

Case Report

A 9 month old girl of Asian origin admitted with abnormal movements of the left lower limb of the body followed by upper limb and face for 10 days of duration prior to the admission. The movements were not associated with up rolling of eyeballs and used to disappear during sleep. She had moderate fever prior to admission 15 days. She was disinterested in surroundings.

Birth history was normal. She was exclusively breast fed child prior to admission Mother was strictly vegetarian.

There was gross motor delay in the form of head holding at 5 months of age and not able to sit without support. She didn't achieve pincer grasp. Neuroregression was noted in the form of loss of head control and social smile with onset of illness. She was poorly nourished baby with weight of 6.0 kilograms. She had pallor; thin sparse hair with knuckle hyper pigmentation. On neurological examination she was hypotonic in all four limbs. Intermittent unilateral tremor of the left side of the upper and lower limb with twitching of the left side of the face was seen.

Material and Methods

Her complete blood work up including full blood count, renal and liver function test was normal. Her metabolic work up including Serum methylmalonic acid, Serum biotinidase, Serum methionine, Serum cysteine, Serum homocysteine, vitamin B12 and folic acid assay was negative. Urine homocysteine was positive. Ophthalmic examination revealed normal fundus with optic disc with normal anterior segment.



Figure: MRI Brain shows in axial T2 images generalized atrophy with mild to moderate prominence of ventricular system.

Taking in to account the history and examination, the diagnosis of Infantile Tremor Syndrome was considered. Child was given a course of injection of vitamin B12- 1000 micrograms daily intramuscular for one week followed by weekly for 4 weeks followed by monthly for 6 months with multivitamins.

Result

With appropriate treatment her tremors gradually reduced and completely stopped after 3 - 4 weeks of starting of the treatment. Child slowly regained the social smile and head control. She started to sit without support and she was completely recovered.

Discussion

Infantile tremor syndrome is a rare clinical disorder. Exact incidence of this syndrome is unknown. 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) [1].

Improvement in nutritional status, living conditions and better weaning practices could explain the reducing incidence rates over the years.

It can be presented from 6 months to 18 months of age. It is manifested as neuroregression, malnutrition and acute tremor. Tremors which can be initially intermittent or can become continuous in few days following an acute infection or stress which are more prominent in distal parts of limbs, head, face and tongue they disappear during sleep. They can be attributed to the structural and functional alteration of extra pyramidal system [2]. Hypotonia with flabbiness of muscle is common with tremulous cry.

Neuroregression in infants has varied aetiology and vitamin B12 deficiency is one of the uncommon causes. Infantile vitamin B12 deficiency is encountered in poorly nourished infants or in offspring of strict vegan mothers. In British Medical Journal two cases (*BMJ Case Reports* 2009; doi:10.1136/bcr.06.2008.0235) both infants of 10 and 8 months of age, whose mothers had vitamin B12 deficiency were reported. On admission, the patients were apathic, hypotonic and lethargic. Serum vitamin B12 levels were below normal limits. Both the infants responded to vitamin B12 treatment [3].

As Vit-B12 deficiency is predominantly found in poorly nourished children also prolonged breast feeding, pure vegetarian diet, iron, magnesium and zinc deficiency has been postulated [4]. The low levels of vitamin B12 and its transport protein Transcobalamin II in the cerebrospinal fluid may be responsible for the neurological features of this syndrome [5].

Other speculations for its etiology include viral encephalitis and degenerative processes.

Vitamin B12 levels are usually low in baby and mother. Prominence of subarachnoid space and ventricular system and cortical atrophy are the commonest findings in CT/MRI of brain [6].

Vitamin B12 in high doses may be required if B12 levels are low. Addition of multivitamins, vitamin C, iron, protein, zinc and magnesium supplements may also be important [7]. It might take months for pigmentary changes of skin and hair to

Come back to normal. Mental dullness and sluggishness takes years to come back to normal [8].

Anaemia mostly is seen which can be micro or macro or normocytic anaemia [6]. Though Vitamin B12 blood levels were normal in our case there is usually deficiency at tissue level so she was treated successfully with Vitamin B12 in high doses [9].

If recognized early the neurological consequences can be completely reversible with appropriate treatment as it happened in our case.

Conclusion

Infantile Tremor Syndrome should be considered in a young child with neuroregression, tremor, developmental delay and malnutrition.

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