INTRODUCTION:

Opsoclonus ataxia syndrome also known as dancing eye dancing leg syndrome is a very rare neurological disorder affecting 1 in 1000000 people worldwide. In children, it is commonly associated with neuroblastoma. It can also be consequent to ingestion of toxic dose of medications like haloperidol and amitryptilin. It can be associated with infections like West Nile virus, Epstein Barr Virus (EBV), Varicella zoster virus, Mycoplasma pneumoniae and listeriosis.

CASE REPORT:

2 years old male child (figure 1) with normal birth and development history had fever of 2 days duration. 5 days after subsidence of fever, child was not able to get up from bed. When made to stand or sit, child had severe truncal ataxia. Child had tremulousness and was not able to hold objects, not able to speak but was able to recognize mother. Child also had difficulty in swallowing, for which he was tube fed. After admission child had fast chaotic eye movements suggestive of opsoclonus. He had incoordination of both upper limbs. Tone was reduced, deep tendon reflexes were present and fundus was normal. Apart from routine investigations which were normal, the child was screened for occult malignancies like neuroblastoma. CT thorax and CT abdomen were normal. Urine metabolic screening, blood Tandem Mass Spectrometry for metabolic disorders were negative. Urine Vanillyl Mandelic Acid estimation was within normal limit. Vasculitic workup was negative. Serum vitamin B12 and serum lactate were normal. CSF analysis showed elevated cell count-135cells/cu.mm (predominantly neutrophils), CSF biochemistry was normal (protein-10mg/dl, sugar-110mg/dl, lactate-13.4mg/dl) CSF EBV Ig G was positive. Screening for HIV, Japanese encephalitis, Hepatitis C virus, Enterovirus, Cytomegalovirus and Herpes simplex virus were negative. Nerve conduction study was normal. He was treated with a course of IV methyl prednisolone followed by IV Immunoglobulin. Ataxia improved much and he became ambulant and opsoclonus disappeared. 2 months later, child presented with increased unsteadiness and was treated with a course of IV methyl prednisolone followed by oral steroid. Ataxia improved partially.

DISCUSSION:

In children opsoclonus ataxia syndrome may be accompanied by myoclonus and typically presents as paraneoplastic syndrome that is usually related to neuroblastoma, which has been excluded in our case as CT thorax and abdomen, urine Vanillyl Mandelic Acid were normal. Metabolic screening for Maple Syrup Urine Disease and Mitochondrial disorders which can present with intermittent ataxia were negative. Other etiologies are infections, multiple sclerosis and SLE. In our case, MRI brain was normal (figure 2). CSF showed positive EBV IgG. EBV infection is associated with a polyclonal B lym-
phocyte proliferation and a spectrum of neutrophilic and non specific antibody response. On occasion EBV infection can give rise to autoimmune / lymphoproliferative disorders. Autoantibodies will be directed against cerebellar purkinji cells as well as peripheral nerve axons. Thus IV immunoglobulin can be used for treatment. Although symptoms are typically steroid responsive, children experience relapse and often suffer cognitive and behavioural problem as long term outcome. Rituximab can be used as an adjunctive therapy. Plasmapheresis may be useful.

CONCLUSION:

In a child presenting with opsoclonus and ataxia, apart from neuroblastoma one should also consider infectious etiologies like EBV.

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