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Case Report

Post infectious opsoclonus-myoclonus-ataxia syndrome-A case report

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Abstract: Opsoclonus-myoclonus syndrome is a rare neurological disorder, usually found in children and associated with neuroblastoma. In adults it is rare and associated with malignancies, toxic medications, metabolic disorders, brain anoxia or autoimmune causes. Here we present a case of post infectious opsoclonus-myoclonus-ataxia syndrome who recovered completely after immunotherapy.
Keywords: opsoclonus-myoclonus, post infectious.

INTRODUCTION

Opsoclonus-myoclonus syndrome (OMS), also known as the "dancing eye syndrome" is a very rare neurological disorder, affecting 1 in 10,000,000 people per year [1, 2]. It is more common in children and is associated with neuroblastoma. In adults it may be associated with malignancies [3], toxic medication, metabolic disorders, hydro electrolytic disorders, brain anoxia, Hashimoto's encephalopathy, and auto antibodies against cerebellar Purkinje cells, celiac disease, and paraneoplastic syndromes [4]. All of above mentioned causes of OMC are very difficult to treat and have bleak prognosis. Although OMC is not considered an infectious disease, it can be associated with some viral infections [5]. Here we report a case of post infectious opsoclonus myoclonus syndrome who recovered completely with treatment.

CASE REPORT

A 35 year female presented with chief complain of blurring and shaking of vision and severe difficulty in walking for last 7 days. She also had high grade fever 15 days back which recovered in next 4 to 5 days. At the time of admission patient was alert, well oriented to time, place and person. Examination of ocular motor nerves revealed conjugate, arrhythmic, chaotic large amplitude eye movements, predominantly in horizontal direction. Patient could not cooperate for optokionetic nystagmus testing. However, voluntary ocular movements were full but tended to exacerbate the opsoclonous. Patient had severe truncal and appendicular ataxia, along with truncal and limb myoclonus. Her myoclonic movements tend to exacerbate on movements. Remaining neurological and systemic examination was within normal limits. On investigations routine blood, urine and cerebrospinal fluid examination were normal. CSF viral markers for herpes virus, Epstein–Barr virus, Coxsackie virus, measeles, mumps, varicella-zoster virus and rubella virus were negative. Blood examination for human immunodeficiency virus, hepatitis B, and C were negative. Thyroid function test and anti TPO antibodies were normal. Toxicological profile was normal. Serum electrolytes were normal. Blood smear for malaria parasite was negative. . Chest radiograph, abdominal ultrasonography, CT scan of Chest and Abdomen and MRI brain were normal. She was diagnosed with idiopathic OMS. In this case, the onset of her OMS was thought to be related to preceding episode of febrile illness. Soon after admission patient was treated with high dose steroid for three days .Plasma paresis was initiated subsequently. She showed marked improvement after 5 plasma exchanges. Clonazepam 0.5 mg three times a day was also started. After 3 months her opsoclonous and myoclonus were reduced and she was able to sit and stand without support. The treatment with Clonazepam was continued and patient became symptom free after 6 months.

DISCUSSION

Opsoclonous-myoclonus syndrome is rare neurological disorder. Occult neuroblastoma may be detected in more than 50% of children's with OMS [6]. In adults it may be associated with breast carcinoma, small-cell lung carcinoma, uterus or ovarian cancers. In 20% of adult patients this syndrome may be presentation of paraneoplastic syndrome [4]. It also occur consequent to drugs (amitryptyline, haloperidol, and diazepam) intake, metabolic disorders, hydro electrolytic disorders, brain anoxia or autoimmune cause i.e. Hashimoto's encephalopathy and celiac disease. Although it is not considered an infectious disease, It can be associated with some viral infections such as the Epstein–Barr virus, coxsackie virus, Saint Louis encephalitis virus,

human immunodeficiency virus, hepatitis C infection, rubella, mumps, varicella-zoster and West Nile virus. In adolescents it appears following Mycoplasma pneumoniae, Rickettsia, Borrelia burgdorferi infections and sometimes after vaccination [6]. In India, few cases in relation to malaria have also been described [7]. The immuno pathogenesis of OMS is poorly understood. There appears to be humoral and cell-mediated immune mechanisms involved both in paraneoplastic and idiopathic syndromes. The pathology underlying OMS is immune mediated damage to purkinge cells in cerebellum. In idiopathic OMS cases, most patients are seronegative for antineuronal antibodies, as in this case. In our case we could not find anything that was suggestive of some occult malignancy, infections or drug intake. Patient had febrile illness two weeks prior to onset of neurological symptoms points towards possible viral infection which generated an immune response and formation of anti neural antibodies causing neuronal dysfunction.

Treatment for opsoclonous-myoclonus syndrome from whatever the cause has not been uniformly successful. The conventional therapies with corticosteroids, intravenous immunoglobulin, plasmapheresis, adrenocorticotropin hormone and antiepileptic drugs are tried with variable results. The combination of cyclophosphamide and dexamethasone pulse therapy or rituximab administered in the refractory cases is promising [8]. Clonazepam, baclofen, valproate and 5 hydroxy tryptophan provides symptomatic relief in these patients.

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