Opsoclonus Myoclonus Syndrome-A Rare Case Report

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ABSTRACT

Opsoclonus Myoclonus Syndrome (OMS) is a severe Autoimmune CNS disorder which predominantly affects young children and causes lifelong neurological disability. Adult onset OMS is rare. About 50% of children with Opsoclonus Myoclonus have Neuroblastoma. In adults, OMS is most frequently due to viral infections. Only 20% of adults have underlying cancer. We are presenting one such case of probable Paraneoplastic OMS in an adult patient.

KEYWORDS: Opsoclonus, Myoclonus, Neuroblastoma, Paraneoplastic.

INTRODUCTION

Opsoclonus Myoclonus Syndrome is a presumed Autoimmune-mediated syndrome characterised by acute or subacute onset of abnormal eye movements, myoclonic jerks, ataxia, dysarthria and behavioural changes in the setting of B-cell expansion within the CSF[1].

Approximately 50% of children with OMS have Neuroblastoma, while only 20% of adults have an underlying malignancy. In adults, OMS is most often due to viral infections. However, only in a few cases specific viruses have been identified. Psittacosis, St.Louis encephalitis, Rickettsia and Coxsackie infections are some of the implicated infections in OMS[2].

CASE REPORT

A 66 year old male patient presented to the Department of Internal Medicine with history of giddiness and ataxia since 1 month. Patient was treated elsewhere as accelerated hypertension but his symptoms persisted and gradually progressed. Patient had no other neurological symptoms. There was no history of fever or headache prior to the onset of symptoms. Patient had no diabetes. He was detected to have hypertension recently.

On Examination: Patient was hemodynamically stable. He was conscious and oriented. Patient was found to have chaotic eyeball movements without any purposive gaze[Fig-1]. He was also found to have ataxia and inability to walk without support. There was no focal neurological deficit, other cerebellar signs or bowel/bladder disturbances.

Patient’s routine blood investigations and brain MRI was normal. The possibility of Opsoclonus without Myoclonus was considered and was referred for neurological evaluation. After 2 months patient came back to us with jerky movements of the upper and lower limbs[Fig-2] along with the abnormal eyeball movements.

This time the possibility of opsoclonus myoclonus syndrome(OMS) was considered and patient was investigated to rule out underlying malignancy. Routine tests like complete blood count and ESR along with electrolytes, chest X-ray, ultrasound abdomen and Prostate Specific Antigen were done and found to be normal. MRI of the brain was repeated and found to be normal.CSF analysis done was also normal. In spite of the normal tests the possibility of Paraneoplastic OMS could not be completely ruled out and patient was given a trial of Methylprednisolone infusion at a dose of 1gm/day for 3 days. Patient did not improve significantly and hence was referred to a higher centre for further management.

About 2 months later patient returned to us with aggravated symptoms since the last couple of days. He was unable to sit or lie still due to the myoclonic jerks. Patient had not been taken to NIMHANS(Bangalore) as advised during the last admission since patient had improved significantly following the last discharge. Hence another trial of Methylprednisolone was tried but patient showed no improvement. We considered IVIg as the next line of treatment but could not do so due to financial constraints. The patient was once again referred to NIMHANS for further management.
DISCUSSION

OMS is a severe autoimmune CNS disorder, which predominantly affects young children and causes lifelong neurological disability. Early recognition and treatment may yield better outcomes[3]. Adult onset OMS is rare. Little is known about adult onset OMS outside of individual case reports[4]. Hence diagnosis of OMS requires a high level of suspicion and a systematic approach for diagnostic testing[3].

Among various etiologies of OMS, paraneoplastic, paraviral or idiopathic encephalitis are the most common causes. OMS in adults may evolve with Lung, Breast or Uterine cancer or Neuroblastoma. Para infectious and idiopathic forms account for about 50% of cases[5]. There is no diagnostic biomarker for OMS currently[3]. As OMS is rare, there is no standard treatment recommendation. Some cases resolve spontaneously or with symptomatic treatment including Clonazepam and Valproic acid[5]. In our patient Clonazepam was tried but with no significant improvement.

In adult onset OMS, the role of immunotherapy is less well established[5]. Approximately 80% of reported patients typically treated with conventional therapies such as ACTH, Corticosteroids and/or IVIG, develop long term neurological morbidity[4]. Our patient appeared to improve with steroids initially, but he did not show a sustained response. His symptoms had worsened on follow up. Newer treatment approaches using early, aggressive therapy with Cyclophosphamide or Rituximab looks promising[3].

CONCLUSION

Opsoclonus Myoclonus Syndrome is a rare entity in adults. It requires a high index of clinical suspicion for diagnosis. The treatment outcomes are not gratifying; but with the introduction of newer treatment modalities, prognosis appears to be promising.

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