Original Research Paper

Volume - 15 | Issue - 08 | August - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Neurology

ADULT ONSET OPSOCLONUS MYOCLONUS SYNDROME: A CASE SERIES

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ABSTRACT Opsoclonus-myoclonus syndrome (OMS) is a rare neurological disorder characterized by opsoclonus, myoclonus, and cerebellar ataxia. We report three cases of adult-onset OMS, all of whom presented with acute onset classical features of OMS. All patients had a favorable outcome, with complete recovery. Early recognition and treatment of OMS are crucial for optimal outcomes. This report highlights the importance of considering OMS in adults with acute onset of ataxia and ocular symptoms and the potential benefits of immunotherapy in managing this condition.

KEYWORDS : Opsoclonus, autoimmune, immunotherapy.

INTRODUCTION

Opsoclonus–myoclonus syndrome or dancing eye syndrome (DES/OMS) (also known as opsoclonus–myoclonus–ataxia syndrome, myoclonic encephalopathy, and Kinsbourne syndrome) is rare condition charecterised by opsoclonus,myoclonus and cerebellar ataxia[1]. It has an incidence of 1 per 5 million total population per year[1]. Common causes in adult onset OMS include paraneoplastic, parainfectious and autoimmune encephalitis.

CASE 1:

A 34 year old male presented with imbalance, dysarthria and abnormal jerky movements of arms and trunk for 21 days. There was a history of fever, headache and vomiting two weeks prior to onset of these symptoms. On examination he was conscious, alert, afebrile with pulse rate of 70/minute and blood pressure of 110/70mmHg. On neurological examination he had high frequency, conjugate ocular oscillations in all directions which indicated opsoclonus, arrhythmic jerky movements of arms and trunk suggesting myoclonus, head tremors, dysarthria, pancerebellar signs (dysdiadokokinesia, impaired finger nose and heel knee test bilaterally). His motor examination revealed normal power in both upper and lower limbs, spasticity and brisk deep tendon reflexes (DTRs) in both lower limbs, positive Babinski and spastic ataxic gait. His mRs was 4. His routine blood investigations (CBS,RFT,LFT,ESR,CRP,Serum Electrolytes) were normal. His CSF studies revealed 80 cells (85%Lymphocytes), protein – 25mg/dl and sugars – 53mg/dl. On the background of fever before the onset of symptoms and CSF findings, post viral OMS was considered and he was started on intravenous (IV) Methylprednisolone (MPS) 1gm for 5 days and injection acyclovir 500 mg tid for 10 days. His MRI Brain and EEG were normal. CSF PCR was negative. As there was no improvement in his symptoms, a possible autoimmune etiology was considered and was started on IVIg 2gm/kg. Screen for autoimmune encephalitis, paraneoplastic antibodies and malignancy (whole body PET) were normal. Patient started showing improvement and had mRs of 2 on discharge. On follow up after two months he had complete remission of symptoms and mRs was 0.

CASE 2:

A 21 year old female presented with a history of imbalance and oscillopsia for 15 days. She had a history of mild dull aching intermittent headache and fever for around five days 2 months prior to the symptoms. On examination she was conscious, alert, afebrile, pulse rate of 88/minute and blood pressure of 120/80 mmHg. Her neurological examination was remarkable for opsoclonus and ataxia (pancerebellar signs). The diagnosis of OMS likely due to postinfectious vs autoimmune cause was considered. Her routine blood investigations (CBS,RFT,LFT,ESR,CRP,Serum Electrolytes) were normal. CSF studies revealed 3 cells (100% lymphocytes), proteins -36 mg/dl and sugars -64mg/dl. Her MRI brain and EEG were normal. CSF PCR was negative. Screen for autoimmune encephalitis, paraneoplastic antibodies and malignancy was normal. She was started on I.V MPS 1gm for 5 days and injection acyclovir 500 mg IV three times a day for 14 days. She started showing improvement and she had complete remission of symptoms at the time of discharge and mRs was zero.

CASE 3:

A 30 year old male presented with a history of fever and headache for 2 days associated with abnormal jerky movement and imbalance. On examination he was conscious, alert, febrile (Temperature of 101 F) with pulse rate of 110/minute and blood pressure of 100/70mmHg. His neurological examination revealed opsoclonus, myoclonus and pancerebellar signs. His mRs was 4. His routine laboratory investigations revealed raised WBC counts (11,000). CSF studies showed 102 cells (96%Lymphocytes), proteins – 39 mg/dl and sugars – 58 mg/dl. His MRI brain and EEG were normal. Considering infective etiology, a viral PCR panel was sent that was negative. He was started on I.V MPS 1 gm and I.V acyclovir 500 mg IV tid . Autoimmune and paraneoplastic workup were negative. As there was no improvement in the symptoms, he was started on injection IVIG 2gm/kg. Patient started showing improvement and had mRs of 2 at the time of discharge. On follow up after 2 months he had complete remission of symptoms and mRs was zero.

Table No.1: Summary Of Clinical Features, Treatment And Outcome Of Patients

SL NO	AGE/ SEX	COMPLAINTS	FINDINGS	TREATMENT	OUTCOME
1	34/M	Ataxia,jerky movements of limbs	Opsoclonus, myoclonus, ataxia	IV steroids, IV Acyclovir, Inj.IVIg	mRS 4 to 0
2	21/F	Oscillopsia, ataxia	Opsoclonus Ataxia	I.V steroids , IV Acyclovir	mRS 2 to 0
3	30/M	Fever, Ataxia, jerky movement of limbs	Opsoclonus, myoclonus, Ataxia	I.Vsteroids, I.V Acyclovir , Inj.IVIg	mRS 4 to 0

DISCUSSION

Opsoclonus-myoclonus-ataxia syndrome (OMS) is a rare disorder of the nervous system . This condition classically presents with a combination of characteristic eye movement disorder opsoclonus and myoclonus, in addition to ataxia, irritability, and sleep disturbance[1]. Opsoclonus is characterised by involuntary, irregular, but conjugate saccadic eye movements either multidirectional or horizontal ("ocular flutter") precipitated by change of fixation. Pathophysiologically, a disordered interaction of "burst" and "omnipause" cells located in the brain stem has been suggested.[2] The associated myoclonus is typically exacerbated by muscle activation and predominantly involves the face, limbs, and trunk.

To fulfil the diagnostic criteria for OMS, at least three of four supportive findings are required, namely 1) Opsoclonus 2) Myoclonus

and/or ataxia, 3) Behavioral change and/or sleep disturbance 4) Tumorous conditions and/or presence of antineuronal antibodies.[3]

Opsoclonus-myoclonus syndrome in adults is associated with multiple etiologies including the paraneoplastic syndromes, parainfectious encephalitis, and toxic-metabolic states. However, in many cases no obvious cause can be found.[4] Idiopathic causes are more common in adults and are usually caused by infection. Mycoplasma pneumonia, salmonella enterica, HIV, hepatitis C virus, rotavirus, chickenpox, and mumps virus are among the most common infectious causative agents reported in the literature.[5]Autoimmune mediated brain stem dysfunction is the suggested underlying pathomechanism. Paraneoplastic OMS in adults may evolve with lung, breast or uterus cancer, or neuroblastoma. The autoimmune mechanism involves anti-Hu and anti-Ri antibodies targeting neuronal structures in the brainstem and cerebellum, leading to opsoclonus.[6] Anti-NMDA receptor encephalitis may present with opsoclonus in addition to psychiatric symptoms, seizures, and dysautonomia. Anti-GAD autoimmune cerebellitis has been reported in both paraneoplastic and non paraneoplastic cases. Opsoclonus has also been observed in demyelinating disorders such as multiple sclerosis and neuromyelitis optica when lesions affect the brainstem. In Sjögren syndrome and systemic lupus erythematosus (SLE), autoimmune vasculitis or neuroinflammation may involve the brainstem, leading to opsoclonus.[7] The first step for all patients with OMS is a thorough diagnostic evaluation for the underlying tumor. Brain MRI is usually normal in the early stages, but recent reports suggest that cerebellar atrophy is common in follow-up. 18F-FDG positron emission tomography (18F-FDG PET) has a high diagnostic value for ruling out neuroblastic tumors. CSF analysis may reveal mild pleocytosis and oligoclonal bands in autoimmune encephalitis, while elevated protein levels point to an inflammatory or paraneoplastic process.[8] Paraneoplastic and autoimmune screening involves testing serum and CSF for specific autoantibodies. Paraneoplastic opsoclonus is commonly associated with anti-Hu, anti-Ri, anti-Yo, and anti-Ma2 antibodies, while autoimmune forms may involve anti-NMDA, anti-GAD, and anti-amphiphysin antibodies. Infectious causes should be explored through polymerase chain reaction (PCR) and serology testing for neurotropic viruses, including EBV, cytomegalovirus, COVID-19, and herpes simplex, West Nile, and influenza viruses. If bacterial infection is suspected, CSF Gram stain and culture are warranted.

As OMS is rare there is no standard treatment recommendation. Treatment is based on case series and previous publications. Some cases resolve spontaneously or with symptomatic treatment including clonazepam, valproic acid, piracetam, thiamine, reserpine, chlormethiazole. Given the immune-mediated pathogenesis, corticosteroids such as oral prednisone or intravenous dexamethasone pulses, along with adrenocorticotropic hormone (ACTH), have demonstrated efficacy. Recent evidence supports the addition of IVIG to steroid therapy for improved response rates.[9] Second-line agents such as rituximab, cyclophosphamide, MMF (600–1200 mg/m²/day), or PLEX are considered for refractory cases.[10]

The prognosis of OMS depends on the underlying cause, severity at diagnosis, treatment response, and the presence of long-term neurological impairments. Early diagnosis and prompt intervention are critical for functional recovery. Several prognostic factors influence the outcome, including age at onset, the underlying cause of the disorder, the severity at diagnosis, and the patient's response to immunotherapy. Early and aggressive immunotherapy with steroids, IVIG, and rituximab can prevent long-term neurological impairment in OMS. Although many patients improve with timely intervention, a subset will experience chronic neurological sequelae. Long-term interprofessional care involving neurologists, oncologists, physiotherapists, and neuropsychologists is essential for optimizing patient outcomes.[11]

CONCLUSION

Our case series highlights the importance of recognizing and treating opsoclonus-myoclonus syndrome (OMS) in adults. Despite its rarity, OMS can have a significant impact on patients' quality of life. Early recognition and treatment with immunotherapy, such as corticosteroids and intravenous immunoglobulin, can lead to favorable outcomes. Our cases demonstrate that adult-onset OMS can be idiopathic, but prompt treatment can result in complete recovery. Further studies are needed to understand the underlying mechanisms and optimize treatment strategies for this rare condition.

Acknowledgement: We would like to acknowledge the patients and their family for their cooperation and consent in publishing this case report. We also appreciate the efforts of the entire medical team involved in patient care.

Financial Support And Sponsorship-Nil

Conflicts Of Interest-Nil

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