



CASE REPORT

Myoclonus-Opsoclonus-Ataxia Syndrome Secondary to Advanced HIV Infection: A Rare and Atypical Case with Management Considerations

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Abstract

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare but serious neurologic disorder that commonly presents with both spontaneous multidirectional saccades (opsoclonus) and involuntary shock-like muscular contractions (myoclonus), but may or may not present with ataxia. This disorder has an incidence of 0.18 cases per million in the total population but has a higher incidence of 0.27-0.40 per million in children due to its link to specific pathologies [1]. OMAS is mostly idiopathic or associated with paraneoplastic syndromes such as neuroblastoma, small cell lung cancer, and ovarian cancer [2]. However, there have been few instances linking OMAS to infectious causes such as HIV [3,4]. This case presents an unusual presentation of OMAS with opsoclonus and ataxia without a myoclonic component in a previously healthy patient who was diagnosed with HIV after admission.

terovirus, West Nile virus, Salmonella, Cytomegalovirus, Coxsackievirus B3, streptococcus, and HIV [3-11]. OMAS commonly presents with both myoclonic and opsoclonic components, with or without ataxia. However, this case reports a previously healthy patient who presents with weakness, dizziness, balance dysfunction, gait instability, and multidirectional opsoclonus, without any spastic muscle contractions.

Case Description

Patient is a 43-year-old female with no past medical history, presented to the emergency department with complaints of weakness and dizziness for four days. She admits to decreased appetite, falling frequently, and a sensation that her eyes are moving. She denies fevers, chills, coughs, shortness of breath, chest pain, palpitations, abdominal pain, nausea, vomiting, difficulty swallowing, dysuria, rashes, seizures, or slurring of speech. Physical examination showed no abnormalities except for mild tachycardia, balance dysfunction, and nystagmus. She was awake, alert, oriented to person, place, and time. She has no motor deficits and sensation is intact. Initial Brain CT showed a small, age indeterminate, right frontal lobe infarction and aspirin was continued.

Throughout the course of the hospital stay, the patient develops extreme photophobia, diminished reflexes bilaterally, focal weakness, blurry vision, and diplopia, but is still alert, oriented to person, time, and

Introduction

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare disorder that presents with spontaneous arrhythmic, multidirectional saccades without a saccadic interval (opsoclonus) and rapid onset, brief, involuntary muscular contractions (myoclonus) which may or may not present with ataxia. Hence, this disease has been referred to as “dancing eyes, dancing feet”. OMAS is mostly idiopathic in origin or in association with cancers such as neuroblastoma in children and breast and small cell lung cancer in adults [2]. However, other reports also link OMAS to infections such as Lyme disease, En-

place, and with gross sensation and CN II-XII still intact. Due to the downbeating nystagmus on ocular motility test, the neurologist considered a cervicomedullary junction pathology but head MRA showed no evidence of pathology and cervical spine MRI reported degenerative bulges at C4-C5 and C5-C6 junction, but otherwise negative. Due to the persistent ataxia and nystagmus, a history of heavy smoking, and weight loss, hematology and oncology was consulted and a paraneoplastic workup was done. However, paraneoplastic markers and antibodies such as rheumatoid factor, antinuclear antibody, Anti-Hu, Anti-Yo, and dsDNA were not detected.

Given her constantly low levels of WBC, an HIV workup was ordered and showed that the Antigen/Antibody HIV screen test was positive but HIV Antibody test was non-reactive, which was deemed as a probable false positive. The HIV-1 viral load was then checked and reported an HIV viral load of 427,459 copies/mL bDNA. At this point, a clinical picture of OMAS secondary to advanced HIV infection was elicited which prompted the start of the treatment for HIV with Raltegravir and Truvada (Emtricitabine and Tenofovir) and pulse high dose steroids for nystagmus. Patient reported mild alleviation of the nystagmus but persisted until discharge.

Discussion

Opsoclonus myoclonus ataxia syndrome is an extremely rare neurological disorder that usually presents idiopathic or as a paraneoplastic syndrome. Its reported incidence is 1 case for every 10,000,000 in the total population but is slightly higher in children. Idiopathic OMAS presents in younger adult patients and have a better clinical outcome compared to paraneoplastic OMAS that presents in older adult patients around the age of 50 [12]. This disease has no apparent predilection for any race and ethnicity but there are 10% more females than males who suffered from OMAS [2].

In most patients with idiopathic OMAS, there are no associated autoimmune antibodies [13]. However, there are OMAS cases where antibodies have been linked with specific neoplasms such as anti-Ri antibody with breast cancer and Anti-Hu and Anti-Glycine receptor (Gly-R) antibody with small cell lung cancer [13,14]. Other autoimmune markers reported are directed against neuronal surface antigens such as dipeptidyl-peptidase-like protein-6 (DPPX), antibodies to non-synaptic surface puncta on neuronal dendrites, glutamic acid decarboxylase (GAD), gamma-aminobutyric acid type A (GABA-A) and type B (GABA-B) receptors, N-methyl-D-aspartate receptor (NMDAR), and antibodies against the surface of cerebellar granular neurons [13,15-19]. In paraneoplastic OMAS, bladder cancer, gastric adenocarcinoma, ovarian teratoma, and malignant melanoma have been associated with this syndrome [12,20-22].

Although rare, OMAS can lead to pervasive and permanent neurological deficits such as dysregulated be-

haviors and affect, irritability, poor attention, impulsivity, and cognitive impairment [23]. Thus, early detection and treatment of the underlying pathology is essential and has improved OMAS and its neurologic sequelae [24]. In the pediatric population, the most commonly prescribed treatments are ACTH, steroids, and IVIg with ACTH having the best initial response [25]. Other cases also used azathioprine, cyclophosphamide, and plasmapheresis which also showed positive results [25]. In the adult population, immunotherapy is less effective but there have been responses to corticosteroids, cyclophosphamide, IVIg, clonazepam, and topiramate [26,27]. However, most adults who experience OMAS are more likely to have relapses [12]. There are ongoing Phase II clinical trials for the efficacy of rituximab for OMAS and also a Phase II clinical trial for carmustine, etoposide, cytarabine and melphalan together with antithymocyte globulin before a peripheral blood stem cell transplant [28,29].

OMAS usually presents with both opsoclonic and myoclonic components, however this patient only complains of balance dysregulation and a sensation that her eyes are moving but denies any spastic muscle contractions. In addition, this patient has no past medical history which is an unusual presentation of OMAS due to its association to neoplasms. Even then, the paraneoplastic workup essentially came back negative but her low WBC and history of weight loss warranted an additional workup for HIV. In testing for OMAS, all possible etiologies should be considered even though the patient presentation does not perfectly fit the typical OMAS picture. This atypical presentation should be highlighted because early detection and treatment of OMAS proves to be essential in the prognosis of the syndrome and any attributed pathologies, which can affect the patient permanently throughout life.

Conclusion

OMAS typically presents with both opsoclonus and myoclonus, with optional ataxia. However, in patients who do not complain of spastic muscle contractions, OMAS should not be retracted from the differential diagnoses, especially in a patient with balance dysregulation. Moreover, OMAS can present in seemingly healthy patients. Early detection and treatment is empirical in this disease so a quick extensive workup should be started to decrease the chances of developing neurological deficits.

Conflict of Interest

The author(s) declare(s) that there is no conflict of interest.

Author's Declaration

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