CASE REPORT

Opsoclonus-myoclonus-ataxia Syndrome Associated with Dengue Fever: A Case Report

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Abstract

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare neurological disorder characterized by irregular multidirectional eye movements, myoclonus, cerebellar ataxia, sleep disturbances, and cognitive dysfunction. Although most commonly occurring as a paraneoplastic syndrome, this condition has occasionally been described following infectious illnesses. This article reports a rare case of OMAS in association with dengue virus infection. This report represents an unusual clinical syndrome associated with a viral infection and reinforces the need for clinical vigilance with regard to neurological syndromes in the context of emerging arboviruses.

Keywords: Dengue, Intravenous immunoglobulin, Opsoclonus-myoclonus-ataxia syndrome, Steroids.

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Case Description

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is a rare neurological disorder, characterized by chaotic, multidirectional, arrhythmic, high amplitude jerky eye movements (opsoclonus), limb and body jerks which can be diffuse or focal (myoclonus), and cerebellar ataxia along with cognitive and sleep disturbances in some cases (Video 1). It was described as dancing eyes-dancing feet syndrome by Kinsbourne in children[1] and is more common in them. The onset is usually abrupt and often severe. About 50% of children with OMAS have occult or manifest neuroblastoma[2] whereas only 20% of adults have underlying cancer.[3] The etiologies of OMAS are paraneoplastic, parainfectious, toxic, and metabolic causes (Video 2). No single antibody marker has been identified. Antineuronal nuclear antibody type 2 (anti-R1 antibody) antibodies are reported in some cases of paraneoplastic OMAS and anti-GAD65 antibodies are rarely found in idiopathic OMAS.[4] It occurs mostly as a parainfectious syndrome and the most common infections reported are streptococcal infections, mycoplasma, Varicella zoster virus, coxsackievirus B3, and Saint Louis encephalitis.[5] It has also been reported after autoimmune anti-N-methyl-D-aspartate receptor encephalitis[6] and rarely after malaria[7,8] and dengue fever.[9,10] Here we report a case of OMAS after dengue fever.

A 37-year-old male presented with a history of fever for 7 days which was diagnosed and treated for dengue outside (Table 1). On the 7th day of fever, he started developing severe jerky eye movements and jerky limb movements with an inability to stand or walk and was referred to our center. His fever had subsided by the 7th day. There were no other systemic symptoms. On admission; he was conscious, afebrile (98˚F), with tachycardia (heart rate 130/minute). Blood pressure was 136/80 and oxygen saturation was 95% on room air. Cardiovascular, respiratory, and gastrointestinal systems were otherwise normal. On neurological examination, he was anxious, and restless, with normal speech. On cranial nerve examination, he had multidirectional, chaotic, and jerky bilateral eye movements suggestive of opsoclonus. There were frequent and severe limb jerking which were both focal and at times involved multiple limbs and trunk. There was ataxia which could not be assessed in detail due to the jerks. Motor power and deep tendon reflexes were normal and planters were flexor.

He was investigated report mentioned in table below, MRI brain was normal. Cerebrospinal fluid (CSF) showed (total cell count—123 cells/µL, lymphocytes 65%, neutrophils 35%, protein 115 mg/dL, glucose 74 mg/dL, matching blood sugar 137 mg/dL, chloride 120 mEq/dL, and lactate dehydrogenase 39 U/L). CSF BioFire assay was negative and cultures were negative. Antineuronal antibodies (paraneoplastic panel) were negative. Antiglutamic acid decarboxylase antibody was minimally raised (value 28.74, normal <17) He was diagnosed with OMAS and started on treatment with injection methylprednisolone 1 gm IV which was given daily for 5 days. He was given gabapentin and clonazepam for myoclonic jerks. He did not show significant improvement after 5 days. After due consent of relatives, he was then given intravenous immunoglobulin (IVIG) 400 mg/kg body weight for 5 days. On the 5th day of IVIG, he showed significant improvement with a decrease in the frequency and severity of eye movements and body jerks.

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Table 1: Dengue report by enzyme-linked immunosorbent assay method

Parameter	*Result*	*Reference interval*
	Dengue IgG	
Dengue IgG serum	6.59 (negative)	Negative: <18 Equivocal: 18–22 Positive: >22
	Dengue IgM	
Dengue IgM serum	11.02 (positive)	Negative: <9 Equivocal: 9–11 Positive: >11
	NS1 antigen	
NS 1 antigen serum	4.03 (negative)	Negative: <9 Equivocal: 9.0–11 Positive: >11

He showed progressive improvement and was discharged from the hospital after 15 days. On discharge, he was able to walk with support and had minimal eye and body jerks.

On subsequent follow-up in Outpatient Department (Video 3), he had improved fully after 1 month was independent in all activities, was able to drive and there were no eye or limb jerks. There was no ataxia as well (Video 4).

Discussion

Opsoclonus-myoclonus-ataxia syndrome (OMAS) is an autoimmune central nervous system disorder, which predominantly affects young children and rarely adults, and may cause lifelong neurological disability. Early detection and better management may yield good results. Adult OMAS is mainly described in case reports and current literature is sparse about this entity. A systematic approach needs to be followed while diagnosing OMAS. Encephalitis of viral and paraneoplastic or idiopathic are the predominant causes of OMAS. Common neoplasms associated with OMAS are breast, uterine, or neuroblastoma. Parainfectious and idiopathic causes account for almost half of all cases of OMAS. There are no validated biomarkers that may help in identifying OMAS.

There are various neurological complications associated with dengue fever that has been noted and such complications lead to remarkable morbidity and mortality. Neurological complications occur in 0.5–6% of the cases with dengue fever. Both direct (neurotropism) and immunological mechanisms are responsible for neurological manifestations in dengue infection.[11]

Though the exact pathophysiology of this entity is unknown it is most likely of an autoimmune nature anti-R1 antibody and other similar antibodies have been described in this disease entity and may be mediators of motor neuron dysfunction, but these antibodies are not commonly found.[2,3] Although the pathogenesis of opsoclonus-myoclonus associated with neuroblastoma and other viruses remains speculative, these patients clearly have a good prognosis for survival.[2]

The diagnosis is nevertheless not a difficult one and can be made with confidence on clinical grounds alone and there may be a good response to steroids/immunotherapies.[1]

Like the treatment given by Rosario et al., we had given pulse steroids and IVIG for the management of OMAS, and the patient responded well to the treatment.[9] As per Pranzatelli et al., the goal of treatment of OMAS is early and aggressive immunotherapy with the goal of gaining a durable complete neurological remission, the best responders appear to be those who received early combination therapy and were only of mild to moderate severity, as in our case we treated aggressively with steroids followed by IVIG.[5]

Like Kurian et al.[6] and Bhalodiya et al.,[7] our case also responded dramatically, and immunomodulatory therapies it has a relatively good prognosis with IVIG.

Supplementary Videos

The supplementary videos 1–4 are available online at www.ijccr.org.

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