Case Report

Opsoclonus Myoclonus Syndrome: A Rare Manifestation of Dengue Infection in a Child

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INTRODUCTION

Opsoclonus myoclonus syndrome (OMS) is an inflammatory neurological disorder, which is characterized by chaotic uncontrolled movements of the eyes and involuntary jerk-like movements of the body caused by contractions of a group of muscles. It was described first by Marcel Kinsbourne in 1962. It is also known as “dancing eyes–dancing feet,” “Kinsbourne syndrome,” “myoclonic encephalopathy,” or “opsoclonic encephalopathy.” It is associated with ocular, motor, behavioral, sleep, and language disturbances. OMS may be associated with an occult body tumor or infections or it may be idiopathic in nature.

We report a case of OMS along with dengue infection. Although dengue can present with some neurological manifestations such as meningoencephalitis, transverse myelitis, or along with Guillain–Barre syndrome (GBS), it is very rare to find OMS associated with dengue.

Case History

A 14-year-old boy presented with high-grade fever accompanied with headache, arthralgia, and myalgia for 4 days. No associated shortness of breath, cough, urinary disturbance, or ear discharge was reported. On the 4th day, he started having abnormal movement in his eyes and arms, with imbalance while walking or sitting without support. On neurological exam, he was oriented to self, relatives, and place but disoriented to time. On extraocular eye movement examination, bilateral, involuntary, chaotic, multidirectional eye movements were noted. Rest of the cranial nerve and motor examination was normal. His motor exam revealed appendicular as well as truncal myoclonus along with severe symmetric ataxia of trunk, gait, and limbs.

KEYWORDS: Dengue, immunotherapy, modalities of treatment, neurological complications, opsoclonus myoclonus syndrome

Abstract

Opsoclonus myoclonus syndrome (OMS) is an inflammatory neurological disorder, which is characterized by chaotic uncontrolled movements of the eyes and involuntary jerk-like movements of the body. Different modalities of treatment have been described in medical literature to treat OMS. Immunomodulatory treatment with either steroids or intravenous immunoglobulin has been considered. Our case was a 14-year-old boy who presented with fever, mild confusion, without any seizures or focal deficits. On examination, he had opsoclonus in his eyes and had cortical myoclonus in his hands and body. On evaluation, he had low platelets, normal metabolic workup, normal brain imaging, and cerebrospinal fluid showed lymphocytic pleocytosis. He was managed conservatively and had spontaneous improvement in opsoclonus myoclonus by 5th day of his illness and complete recovery in 2 weeks. Although dengue is primarily considered hematotropic virus, it can involve nervous system as well and manifest with OMS.

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Sensory exam and deep tendon reflexes were normal. He had neck rigidity and positive Kernig’s sign. His hemogram revealed thrombocytopenia with a platelet count of 50000/mm³ with normal values for all other blood cells. His renal functions, liver functions, electrolytes, urine analysis, peripheral smear for malaria, blood and urine culture, and X-ray chest were normal. IgG and IgM antibody for dengue virus and NS1 antigen specific for dengue infection were positive. In the presence of fever, thrombocytopenia, and positive dengue seromarkers, a diagnosis of dengue was made. However, as dengue is not commonly known to cause these neurologic abnormalities, further workup was carried out. Magnetic resonance imaging (MRI) of the brain with contrast was normal. Cerebrospinal fluid (CSF) examination revealed 25 cells with lymphocytic pleocytosis with 68 mg/dL protein and normal glucose. Over the next 4–5 days, he was given supportive management with intravenous fluids and paracetamol. Subsequent hemogram showed gradually recovering thrombocytopenia. He became afebrile after 5 days of hospitalization and his platelets improved to 270,000/mm³. His opsoclonus, myoclonus and ataxia improved spontaneously and he did not require any immunomodulation or symptomatic treatment.

**Discussion**

Dengue is a mosquito-borne viral disease caused by dengue virus, which belongs to Flaviviridae family, having four closely related but antigenically different serotypes: DENV1, DENV2, DENV3, and DENV4.[1,2] Serotypes commonly associated with neurologic manifestations are DENV1 and DENV2.[3-5] DENV4 was also detected in the immunochemistry of CSF of a patient with encephalitis.[6] Dengue is endemic in almost all tropical and subtropical regions and is the second most common mosquito-borne disease globally.[6] Dengue can manifest itself in varied presentations including asymptomatic state, dengue fever, dengue hemorrhagic fever, and dengue shock syndrome; it may rarely cause neurological manifestations.

Neurological manifestations associated with dengue include encephalitis, myelitis, meningitis, GBS, Miller Fisher syndrome, neuromyelitis optica, and optic neuritis.[7] In a study conducted in Lucknow, Uttar Pradesh, India, several neurological manifestations have been described in association with dengue infection. OMS was found in only two cases in that series of patients. Our case was a child with confirmed dengue infection who developed opsoclonus–myoclonus–ataxia. We had ruled out other causes for opsoclonus–myoclonus–ataxia by performing an MRI, which was unrevealing, and no history of malignancy was reported with his opsoclonus–myoclonic movements, which resolved spontaneously over a week. Although his opsoclonus and myoclonic movements occurred a few days after the onset of his fever suggesting a postinfectious immune injury, CSF pleocytosis may suggest dengue encephalitis.

Neurological complications in dengue infection have been hypothesized through three pathogenic mechanisms: neurotropism, systemic complications, or postinfectious immune-mediated mechanism or antibody-dependent enhancement. It is likely that the OMS could be secondary to both encephalitis and postinfectious immune injury. Once a host is infected with dengue infection, the infection provides lifelong immunity to the same subtype but no protection against the other three subtypes. This phenomenon is called antibody-dependent enhancement. The antibodies form complexes with the virus and infect mononuclear phagocytes, which consequently result in the infection of higher number of host cells, thus improving viral replication and worsening clinical signs, which may lead to neurological manifestations of dengue infection.

OMS is usually recognized as a manifestation of neuroblastoma in children[8] and paraneoplastic syndrome in adults.[9] Increasing evidence that OMS can occur as a manifestation of bacterial or viral infections such as coxsackievirus, Epstein–Barr virus, adenovirus, herpes virus, influenza virus, human immunodeficiency virus (HIV), as well as post-streptococcal or mycoplasma infection is available.[10] OMS is rarely seen as a manifestation of dengue. Table 1 enlists cases in medical literature describing OMS in parainfectious settings. To the best of our knowledge, OMS has only been reported in three cases with dengue infection (two adults and a child).[5,11] Of the two adults, presence of NS1 or IgG seroconversion was not determined in one of the cases, whereas in the child, MRI and CSF were not performed to exclude other causes.

Different modalities of treatment have been described in medical literature to treat OMS. In patients with OMS in the setting of infection, along with treatment of infection, immunomodulatory treatment with either steroids or intravenous immunoglobulin has been considered. Symptomatic treatment with clonazepam.
Our case is unique in that it has features of both direct neurotropic and immune-mediated damage, and it improved spontaneously without any immunomodulatory treatment.

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Conflicts of interest
There are no conflicts of interest.

References
1. WHO/TDR. Dengue guidelines for diagnosis, treatment, prevention and control. New ed. Geneva, Switzerland: World Health Organization; 2009.
2. Carod-Artal FJ, Wichmann O, Farrar J, Gascón J. Neurological complications of dengue virus infection. Lancet Neurol 2013;12:906-19.
3. Verma R, Sahu R, Holla V. Neurological manifestations of dengue viral infection: a review. J Neurol Sci 2013;327:1-6.
4. Verma R, Sahu R, Jain A, Garg RK, Singh MK, Mehrotra HS, et al. Neurological complications in dengue virus infection: a prospective cohort study. Neurology 2014;83:1601-9.
5. Tan AH, Linn K, Ramli NM, Hlaing CS, Aye AM, Sam IC, et al. Opsoclonus-myoclonus-ataxia syndrome associated with dengue virus infection. Parkinsonism Relat Disord 2014;20:1309-10.
6. Ramos C, Sánchez G, Pando RH, Baquera J, Hernández D, Mota J, et al. Dengue virus in the brain of a fatal case of hemorrhagic dengue fever. J Neurovirol 1998;4:465-8.
7. Miagostovich MP, Ramos RG, Nicol AF, Nogueira RM, Cuzzi-Mayo T, Oliveira AV, et al. Retrospective study on dengue fatal cases. Clin Neuropathol 1997;16:204-8.
8. Tate ED, Allison TJ, Pranzatelli MR, Verhulst SJ. Neuroepidemiologic trends in 105 US cases of pediatric opsonoclonus-myoclonus syndrome. J Pediatr Oncol Nurs 2005;22:8-19.
9. Bataller L, Graus F, Saiz A, Vilchez JJ; Spanish Opsoclonus-Myoclonus Study Group. Clinical outcome in adult onset idiopathic or paraneoplastic opsonoclonus–myoclonus. Brain 2001;124:437-43.
10. Pruitt AA. Infections of the cerebellum. Neurol Clin 2014;32:1117-31.
11. Verma R, Sharma P, Garg RK, Atam V, Singh MK, Mehrotra HS. Neurological complications of dengue fever: experience from a tertiary center of north India. Ann Indian Acad Neurol 2011;14:272-8.
12. Dassan P, Clarke C, Sharp DJ. A case of poststreptococcal opsonoclonus-myoclonus syndrome. Mov Disord 2007;22:1490-1.
13. Peter L, Jung J, Tilikete C, Ryvlin P, Mauguiere F. Opsoclonus-myoclonus as a manifestation of Lyme disease. J Neurol Neurosurg Psychiatry 2006;77:1090-1.
14. McCarthy VP, Zimmerman AW, Miller CA. Central nervous system manifestations of parainfluenza virus type 3 infections in childhood. Pediatr Neurol 1990;6:197-201.
15. Kuban KC, Ephros MA, Freeman RL, Laffell LB, Bresnan MJ. Syndrome of opsonoclonus-myoclonus caused by Coxsackie B3 infection. Ann Neurol 1983;13:69-71.
16. Syrbe S, Merkenschlager A, Bernhard MK, Grosche J, Liebert UG, Hirsch W, et al. Opsonoclonus-myoclonus syndrome after adenovirus infection. Springerplus 2015;4:636.
17. Naselli A, Pala G, Cresta F, Finetti M, Biancheri R, Renna S. Acute post-infectious cerebellar ataxia due to co-infection of human herpesvirus-6 and adenovirus mimicking myositis. Ital J Pediatr 2014;40:98.
18. Kang BH, Kim JI. Opsoclonus-myoclonus syndrome associated with mumps virus infection. J Clin Neurol 2014;10:272-5.

19. Bîrluțiu V, Bîrluțiu RM. Opsoclonus-myoclonus syndrome attributable to West Nile encephalitis: a case report. J Med Case Rep 2014;8:232.

20. Morita A, Ishihara M, Kamei S, Ishikawa H. Opsoclonus-myoclonus syndrome following influenza A infection. Intern Med 2012;51:2429-31.

21. Ertekin V, Tan H. Opsoclonus-myoclonus syndrome attributable to hepatitis C infection. Pediatr Neurol 2010;42:441-2.