



POST-INFECTIOUS OPSOCLONUS-MYOCLONUS ATAXIA SYNDROME IN RICKETTSIAL DISEASE

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Abstract

Background: Opsoclonus-Myoclonus Ataxia (OMA) syndrome is a rare neurological disorder that occurs in approximately 1 in 5,000,000 individuals per year in the general population. We report three cases of OMA syndrome presenting post-rickettsial infection.

Discussion: The prevalence of rickettsial infections in the South Indian population is estimated to be around 10% to 30% among acute undifferentiated febrile illnesses. While headache and encephalitis are well-known neurological manifestations of this infection, "dancing feet–dancing eyes" (OMA) is rarely seen.

Methods & Results: We present three such unique cases. Rickettsial disease was confirmed using the Weil-Felix test. All patients were thoroughly screened for underlying paraneoplastic conditions. Every patient responded exceptionally well to a combination of pulse intravenous methylprednisolone (IVMP) and doxycycline.

Key Words: opsoclonus-myoclonus-ataxia syndrome, rickettsial illness

Introduction

Dancing eyes-dancing feet syndrome (Opsoclonus-Myoclonus Ataxia) is a rare clinical entity seen across all age groups, though it is traditionally more common in children. It is frequently associated with paraneoplastic conditions but can also manifest as a parainfectious phenomenon.

Rickettsial infections typically present with fever, rashes, eschars, and multisystem involvement. While headache, altered sensorium, and encephalitis are established neurological manifestations, OMA syndrome developed post-infectiously in all of our cases within one week of the onset of febrile illness. Neuroimaging and cerebrospinal fluid (CSF) analyses were largely normal or non-specific.

Patients were systematically screened for other co-infections and paraneoplastic conditions. They were treated with pulse methylprednisolone and doxycycline. All patients showed a clinical response within a week and achieved complete recovery within a month.

Case Reports

Case Report 1 (IP No. 1911180001)

A 50-year-old gentleman presented with an acute febrile illness of 3 days' duration. He had no neurological deficits upon admission. While being treated symptomatically, he developed abnormal movements in his upper and lower limbs, alongside walking difficulties on day 3.

- **Examination:** Revealed opsoclonus, myoclonus involving all four limbs, gait ataxia, and bilaterally impaired finger-to-nose testing.
- **Investigations:** Routine labs showed thrombocytopenia. Brain MRI revealed bilateral cortical FLAIR hyperintensities. CSF analysis showed elevated proteins (81.5 \text{ mg/dL}). The Weil-Felix test was positive (OX19: negative; OX2: 1:80; OXK: 1:160). Serological screening for autoimmune and paraneoplastic encephalitis was negative.
- **Treatment & Outcome:** He was started on doxycycline and high-dose methylprednisolone (1 gram per day) for five days. His myoclonus gradually subsided by second day, and his ataxia improved by day three of IVMP. Following a 5-day course of IVMP, the patient was able to walk with support, and OMA symptoms drastically reduced. He was briefly prescribed with amantadine. At his 2-week outpatient review, symptoms had significantly decreased, and he achieved complete recovery at 4 weeks.

Case Report 2

A 35-year-old lady, initially treated for a viral fever at a local hospital, presented to the neurology outpatient department (OPD) one week later with progressive walking difficulty and involuntary hand movements.

- **Examination:** Demonstrated prominent opsoclonus, limb myoclonus (predominantly upper limbs), and severe gait ataxia.
- **Investigations:** Routine blood work was normal. The Weil-Felix test was positive (OX19: 1:80; OX2: 1:160(positive); OXK: negative). CSF analysis showed elevated proteins without pleocytosis. Brain MRI was unremarkable.
- **Treatment & Outcome:** She was treated with doxycycline and pulse methylprednisolone (1 gram per day). Significant improvement in her opsoclonus and myoclonus was noted on second day, followed by improvement in ataxia on fourth day. After five days of IVMP, she could ambulate with minimal assistance. Complete recovery was documented at her 3-week outpatient follow-up.

Case Report 3

A 50-year-old lady presented with a 4 days history of fever, giddiness, headache, and difficulty in walking and imbalance.

- **Examination:** On admission, she was febrile with gaze-evoked nystagmus, impaired finger-to-nose testing, gait ataxia, and neck rigidity.

- **Investigations:** Initial brain MRI was normal. CSF analysis revealed lymphocytic pleocytosis (272 cells/mm^3 with 92% lymphocytes) and raised proteins (164 mg/dL). Routine blood investigations were unremarkable.
- **Progression & Treatment:** She was initially started on intravenous ceftriaxone. During her hospital stay, on day 3, her ataxia worsened, upper limb myoclonus emerged, and classic "dancing eyes" were observed, prompting a diagnosis of OMA syndrome. Subsequent workup revealed a positive Weil-Felix test (OX19: 1:80; OX2: 1:160; OXK: negative). Contrast brain MRI remained normal.
- **Outcome:** She was immediately switched to doxycycline and pulse methylprednisolone (1 gram daily). By day 2, a significant decline in myoclonus and opsoclonus was noted, and she began ambulating with support by day 4. She fully recovered by her 4-week outpatient follow-up.

Discussion

Opsoclonus consists of involuntary, irregular, chaotic, continuous, large-amplitude, multidirectional saccades. It persists during fixation, smooth pursuit, convergence, and even during sleep or eyelid closure. Myoclonus typically affects the limbs and trunk, often accompanied by ataxia, irritability, and altered sensorium.

In 1962, Marcel Kinsbourne, an Austrian-born pediatric neurologist, first described OMA in children with neuroblastoma and noted the efficacy of ACTH therapy in achieving remission.

Epidemiology: The overall incidence of Opsoclonus-Myoclonus-Ataxia syndrome is estimated at 0.18 cases per million population per year. It is most frequently recognized in children associated with neuroblastoma, with a mean onset age of 1.5 years and a slight female preponderance. Only about 20% of OMA cases occur in adults. Adult-onset OMA is rare, presenting at a mean age of 49 years, and is highly associated with paraneoplastic conditions (e.g., anti-Hu or anti-Ri antibodies secondary to small cell lung cancer, ovarian teratomas, and breast cancer). anti Ri antibodies has more aggressive and rigid myoclonus.

OMA can also be triggered by infectious agents. HIV is the most common viral etiology, though it has been reported with the herpes virus family, Cocksackievirus B3 and B2 variants, Dengue, and Chikungunya. Bacterial and atypical pathogens like *Borrelia*, *Mycoplasma*, *Rickettsia*, and *Chlamydia psittaci* are exceptionally rare causes.

While paediatric and paraneoplastic OMA cases often necessitate complex immunotherapy regimens (ACTH, IVIG, rituximab, or cyclophosphamide), our series highlights three cases of post-rickettsial OMA that achieved complete remission using only targeted antibiotics and high-dose corticosteroid monotherapy.

Pathogenesis of OMA is possibly autoimmune causing loss of Purkinje cells, and dentate demyelination which is noted in autopsy. Brainstem and cerebellum together, have also been implicated. Three different premotor neurons, burst, tonic and pause cells, which are located in pontine reticular formation, ultimately control the eye movements. Tonic cells holds the eye in new position where as Burst cells initiate saccades and pause cells inhibit saccades. Disease of burst cells produces abnormal saccades. Disease of pause cells produces an abnormal oscillation like flutter or opsoclonus. A lesion in dentatorubral pathway may be responsible for occurrence of opsoclonus and myoclonus of limbs and palate'.

In developing nations like India, where endemic and scrub typhus are highly prevalent (primarily transmitted by vectors like mites and ticks), rickettsial diseases frequently present as pyrexia of unknown origin. Diagnosis relies heavily on the Weil-Felix test, IgM ELISA, or immunofluorescence assays (IFA). Complications like encephalitis are well known in rickettsial diseases however, OMA is a rare entity. They respond well to doxycycline or tetracycline along with pulse corticosteroids and supportive care.

Conclusion

Rickettsial infection is a prevalent cause of acute febrile illness in India and can manifest with varied neurological complications. Although OMA syndrome remains an exceedingly rare complication of rickettsial diseases, clinicians should maintain a high index of suspicion. Most post-rickettsial OMA cases respond remarkably well to IVMP and doxycycline. However, underlying neoplastic or autoimmune etiologies must always be thoroughly ruled out before assuming a parainfectious origin.

Consent: - obtained from all the patients in their local languages

Conflict of interest: - Nil

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