



THE MITE AND THE EYE ROCK AND ROLL – AN INTERESTING CASE OF OPSOCLONUS MYOCLONUS SYNDROME IN A PATIENT WITH SCRUB TYPHUS

Neurology

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ABSTRACT

Scrub typhus is a common yet under-recognized tropical infectious disease commonly seen in rural India that can affect various body systems, all of which have endothelial cell dysfunction as the common pathophysiology. Of which central nervous system manifestations are quite rare. Here, we report a This case was presented with Opsoclonus myoclonus syndrome due to scrub typhus infection. The patient was treated with doxycycline and pulse methylprednisolone which showed dramatic improvement in his clinical condition.

KEYWORDS

parainfectious, neuroinfectious disease, scrub typhus, opsoclonus myoclonus syndrome

INTRODUCTION

The term Opsoclonus Myoclonus syndrome (OMS) was first described by Marcel Kinsbourne in the year 1962[1].

Out of various causes of Opsoclonus Myoclonus syndrome, paraneoplastic, parainfectious, or idiopathic encephalitis accounts for most of the causes of OMS and an immune-mediated dysfunction of the brainstem has been suggested as the underlying pathophysiology [2].

Among infectious causes of OMS, Epstein-Barr virus, human immunodeficiency virus, and streptococcal infections are often associated with opsoclonus myoclonus syndrome (OMS).

CASE VIGNETTE

64-year-old male presented with complaints of fever for a week duration, following which he developed involuntary jerky movements of all four limbs which were aggravated by movement, and also had a rapid involuntary movement of eyeballs in all directions. An initial diagnosis of Opsoclonus myoclonus syndrome was made. CSF analysis was done which revealed cells – 50 cells/cu.mm, predominantly lymphocytes, protein – 52.8 mg/dl (Normal: 15 – 45 mg/dl), and glucose 50 mg/dl. The Vasculitic profile was normal. EEG showed a non-specific slowing of waves. MRI brain with contrast was normal. The patient was initially treated with ceftriaxone, Acyclovir, and dexamethasone but the patient didn't show much improvement. A paraneoplastic (anti-Hu, anti-Ri, Anti Yo, Anti ma2, and Anti CV2/CRMP5) and autoimmune panel were sent which was also negative. And finally, scrub typhus serology was done which turned out to be positive. The patient was started on Doxycycline along with pulse methylprednisolone for 5 days followed by gradual oral taper off after which he showed a significant improvement. Doxycycline therapy was continued for two weeks. At the end of two weeks, the patient had significantly reduced opsoclonus and myoclonic jerks.

DISCUSSION

Scrub typhus is caused by *Orientia tsutsugamushi* and it is transmitted by the larval form of the vector *leptotrombidium* mite commonly referred to as chiggers, and it is called "chiggerosis"[3]. It is a zoonotic disease and one of the most common re-emerging Rickettsial infections in the Indian subcontinent [3]. The non-specific nature of its presentation and low index of suspicion often lead to delayed diagnosis and treatment. Pathophysiologically, human endothelial cells are the primary target of *O.tsutsugamushi*. Scrub typhus is predominantly a multisystem disorder initially presenting with a febrile illness, thrombocytopenia, liver dysfunction, and hallmark skin lesion at the site of the bite termed "eschar".

Neurological involvement ranges from meningoencephalitis to encephalopathy, and seizures. Rare manifestations include cerebellar involvement, opsoclonus myoclonus syndrome, cranial neuropathies, ADEM, etc[4]. In the central nervous system, endothelial dysfunction produces a plethora of complications. Another mechanism of underlying neurologic manifestation is immune-mediated, which explains certain late-onset neurological manifestations including opsoclonus, myoclonus, GBS, and myelitis[5].

Scrub typhus is a rare parainfectious cause of opsoclonus myoclonus syndrome. The first case was reported by Nam et al. in 2010[6]. The important diagnostic testing for scrub typhus is serologic testing. *O.tsutsugamushi* being an intracellular pathogen, cannot be isolated via standard bacterial culture. So, nucleic acid amplification tests form the mainstay of diagnosis. ELISA is a frequently used test because of its wide availability and need for less technical input.

Doxycycline is the treatment of choice. Azithromycin is an alternative agent used for scrub typhus. Most neurological features respond well to these antibiotics. However, some of the manifestations that have immune pathogenesis like opsoclonus myoclonus syndrome may require high-dose steroids or intravenous immunoglobulins.

Learning Points For Clinicians

The causes of adult-onset opsoclonus myoclonus are myriad including paraneoplastic and parainfectious etiologies. This case report highlights the consideration of scrub typhus in the differential diagnosis of OMS, especially in endemic regions like the Indian subcontinent, where the presentation is an acute febrile illness along with multi-organ dysfunction because of its highly treatable nature yet potentially fatal.

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