

## Case Report

# Acute cerebellar ataxia in a 3-year-old Bengali girl: a novel presentation of scrub typhus in pediatric age group

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### ABSTRACT

Scrub typhus is known to cause local and systemic vasculitic response in almost all the systems of the body. In central nervous system it most commonly causes meningitis and encephalitis although several other atypical presentations have been documented. We present a case of a 3 years old child presenting with fever and isolated acute cerebellitis. Serum showed IgM antibodies to scrub typhus by ELISA. MRI scan of brain also corroborated with clinical findings. Patient showed rapid response to doxycycline therapy.

**Keywords:** Cerebellitis, MRI, *Orientia tsutsugamushi*, Rickettsia, Scrub typhus

### INTRODUCTION

Scrub typhus, a rickettsial infection caused by *Orientia tsutsugamushi* commonly found in Indian subcontinent has got a wide spectrum of presentations and complications. Though it involves almost all the organs and systems of the body, interstitial pneumonia, acute renal failure and multiple organ failure, gastrointestinal bleeding etc. are some of the serious complications.<sup>1</sup> Central nervous system involvement usually comprises of meningitis and encephalitis.<sup>1</sup> Febrile illness due to scrub typhus presenting with acute cerebellar ataxia is rare.

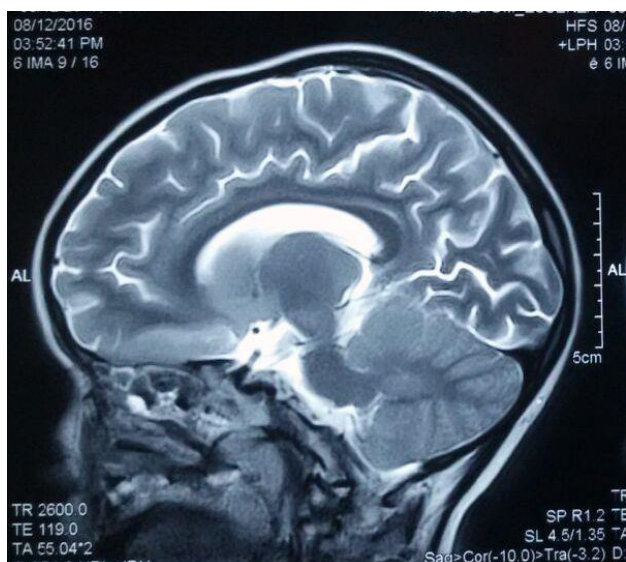
### CASE REPORT

A 3-years-old previously normal female child from rural West Bengal presented with history of fever for last 5 days and inability to sit or stand for last 2 days. There was no history of trauma, low back pain, loss of consciousness or seizure. Mother also noticed change in quality of speech. On general examination, child was febrile, irritable and was having considerable photophobia. Moderate pallor along with generalised

lymphadenopathy was present. Vitals were within normal limits.

Systemic examination revealed: Signs of meningeal irritation present. Child was ataxic having signs of truncal as well as peripheral ataxia involving all the limbs. Horizontal nystagmus was evident bilaterally. Tone, power, superficial and deep tendon reflexes, cranial nerves were normal. No root pain could be elicited by straight leg rising test. Liver was 3cm palpable below the right subcostal margin along mid clavicular line and spleen was palpable 2cm along the splenic axis. Cardiovascular and respiratory systems were within normal limits.

Initial investigations revealed: Hb- 9.7 gm%, Total leukocyte count- 9,500/cmm (Neutrophil-59%, Lymphocyte-30%), Platelet - 1.56 lakh/cmm; C-reactive protein- 45 mg/dL (cut off 5 mg/dL), Albumin- 2.5gm/dl, Na- 130 mEq/L, K- 4.3mEq/L, alanine aminotransferase(ALT)- 164 U/L, aspartate aminotransferase(AST)-204 U/L, renal function tests-normal.



**Figure 1: Parasagittal T2 weighted MRI image showing bilateral signal changes and hyperintensities involving both the cerebellar cortices suggestive of acute cerebellitis.**

Lumbar puncture report was suggestive of aseptic meningitis: clear CSF with 10 cells (100% lymphocytic), glucose and protein were normal. Widal test and tuberculin test were negative. Blood and urine culture didn't show any growth. Weil-Felix test revealed OXK positive in titres of 1:320 and scrub typhus IgM level came out to be 2.377 (cut off : 0.500).

Chest X-ray was within normal limits. Ultrasonogram showed hepatosplenomegaly. MRI brain was done (to find out the cause of ataxia) which showed (Figure 1) signal abnormality in the cerebellum with effacement of cerebellar folia in T2 weighted and FLAIR images which gave an impression of acute cerebellitis.

Apart from supportive treatments, patient was initially started on intravenous ceftriaxone (100 mg/kg/day) but was shifted to intravenous doxycycline (5 mg/kg/day) after obtaining the scrub typhus reports. Clinical response was observed within 48 hours. Fever came down, photophobia also resolved. Doxycycline (first intravenous, then oral) was continued for a total duration of 10 days. Child regained ability to sit at the time of discharge but mild ataxia persisted which after 1 week of follow up subsided.

## DISCUSSION

Scrub typhus, a disease which is transmitted by the bite of larva (chiggers) of trombiculid mites usually presents with fever, headache, muscle pain, sore throat, abdominal pain and sometimes eschar.<sup>1</sup> Among the various clinical manifestations, myocarditis and encephalitis are most life-threatening complications.<sup>2</sup> South East Asia and the Pacific Rim are the regions where this infection is most prevalent with about 1 million new cases per year.<sup>3</sup>

Among the rickettsial diseases epidemic typhus and Rocky Mountain spotted fever are the main culprits causing neurological manifestations followed by scrub typhus, meningitis and meningoencephalitis being the most common presentations.<sup>1</sup> Other manifestations include cerebral hemorrhage, acute disseminated encephalomyelitis, transient parkinsonism, 6th and 7th cranial nerve palsy, hearing loss, opsoclonus, trigeminal neuralgia, myoclonus, myelitis, Guillain-Barre syndrome, brachial plexopathy, polyneuropathy etc.<sup>4</sup>

Rickettsial organisms attack endothelial cells by a mechanism of oxidative stress leading to inflammation causing local and systemic vasculitis. In case of scrub typhus, involvement of central nervous system is considered to be due to proliferation of *O. tsutsugamushi* in the endothelium of small blood vessels. This invasion leads to immune response in the form of release of cytokines which damages the endothelium causing platelet aggregation, fluid leakage and focal microinfarction along with proliferation of polymorphs and monocytes. Other organs like skin, kidney, skeletal muscle and cardiac muscle can also be involved by this process.<sup>5</sup>

Pathological findings seen in central nervous system in cases of scrub typhus includes diffuse or focal mononuclear cell exudates in leptomeninges, hemorrhages in brain substance and classically presence of typhus nodules (which are clusters of microglial cells) distributed throughout the brain parenchyma.<sup>6</sup>

Diagnosing this infection early is important considering its complications and easily available treatment modality. Diagnosis can be difficult without advanced laboratory techniques.

Serological tests such as: Microimmunofluorescence, latex agglutination, indirect hemagglutination, immunoperoxidase assay, and enzyme-linked immunosorbent assay (ELISA) remain the most important tests for the diagnosis.<sup>7</sup> Immunofluorescence assay (IFA) is the "gold standard" test. In our case we used ELISA as diagnostic method. Routine CSF study in case of scrub typhus usually shows lymphocytic pleocytosis with normal glucose and protein level, similar to that of viral meningitis.<sup>8</sup> Imaging like CT/MRI scans are often useful in diagnosing neurological involvements like hemorrhage, ADEM or cerebral edema. In our case the MRI finding of signal changes and visualization of loss of outlines of folia corroborated with the clinical diagnosis of acute cerebellar involvement.

Though from some parts of the Asian countries drug resistant is being reported still doxycycline is the drug of choice.<sup>5</sup> In cases not responding to doxycycline, rifampicin is used.<sup>9</sup> Chloramphenicol is another option.

Though Choi et al used steroids in a patient of scrub typhus with acutely progressive local neurologic

symptoms and subsequently the presenting neurologic symptoms such as restlessness, irritability, abnormal lateral gaze and paralysis in upper extremities resolved, there is still no consensus of using corticosteroids in scrub typhus.<sup>10</sup> In our case the child improved without using any corticosteroid.

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