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.1 Central nervous system involvement usually comprises of meningitis and encephalitis.1 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.
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. Other manifestations include cerebral hemorrhage, acute disseminated encephalomyelitis, transient parkinsonism, 6th and 7th cranial nerve palsy, hearing loss, opcosclonus, trigeminal neuralgia, myoclonus, myelitis, Guillain-Barre syndrome, brachial plexopathy, polynepropathy etc.

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.

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.

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. 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. 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. In cases not responding to doxycycline, rifampicin is used. 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. In our case the child improved without using any corticosteroid.

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