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Case Report

Acute cerebellar ataxia: a rare 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. Scrub typhus very rarely presents itself with CNS manifestations. In central nervous system it most commonly causes meningitis and encephalitis although several other atypical presentations have been documented. Cerebellar ataxia, which is the lack of coordination, has a number of causes none of which are as uncommon or unheard of as Scrub Typhus. We report a case of a 15 years old child presenting with fever and isolated acute cerebellitis. Scrub Typhus was diagnosed by serum IgM ELISA. Patient showed rapid response to doxycycline therapy.

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1. Introduction

Scrub typhus is a rickettsial disease caused by *Orientia tsutsugamushi*. The disease is typically characterized by fever, myalgia, sore throat, rash, eschar formation, abdominal pain and headache along with signs of generalized lymphadenopathy and hepatosplenomegaly. Serious complications include interstitial pneumonia, gastrointestinal bleeding, acute renal failure, and multiple organ failures. Neurological involvement has been commonly reported in rickettsial diseases in the form of meningitis and encephalitis.¹ However, cerebellar involvement by this organism is very rare. Febrile illness due to scrub typhus presenting with acute cerebellar ataxia is rare.

Ataxia is defined as the inability to make smooth accurate and coordinated movements because of a dysfunction of the cerebellum, its inputs or outputs, its sensory pathways in the posterior columns of the spinal cord, or a combination of these.²

We report a rare case of A 15-year-old boy from the Udaipur district in Rajasthan, with no significant past history presented with fever for 5 days, along with difficulty in walking and truncal ataxia for last 2 days, slurring of speech last 1 day. The fever was high grade (102.4F on initial presentation) and intermittent in nature without chills, rigor, or rash. It used to subside on taking antipyretics. The patient started having difficulty in walking with a tendency to fall to any one side and over the course of 2 days, she was not able to walk. There was neither any history of seizures, loss of consciousness, altered sensorium, trauma, low back pain, nor any weakness nor of tinnitus, vertigo, or ear discharge. Bladder and bowel habit was normal.

On general examination - child was febrile, drowsy. There was no other abnormal finding on general survey, no apparent rash or neurocutaneous marker noted. A local examination revealed an eschar on the right thigh. Pulse was 120/min. Blood pressure was 110/80, which is just above the 95th percentile for age, sex, and height. Respiratory rate was 24/min.

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1.1. Neurological examination

Higher functions were normal with no impairment in memory. Patient was well oriented to time, place, and person. There was no muscle weakness or hyper/hypotonia. Tendon reflexes were normal as were the superficial reflexes. There was no impairment in smell or taste.

All other cranial nerves were within normal limit. Sensory system examination revealed no impairment in fine touch, pain, temperature, position, and vibration sense. Cerebellar examination revealed unsteady gait with inability to walk in a straight line and tendency to fall to one side along with past pointing on finger nose test and dysdiadochokinesia. Neck rigidity was present, but Kernig's and Brudzinski's signs were negative. Fundoscopic examination did not reveal any signs of papilloedema. There was no organomegaly on abdominal examination. Cardiovascular and respiratory systems were within normal limit.

Initial investigations revealed: Hb-9.8gm%, Total leukocyte count- 4,500/cmm (Neutrophil-71%, Lymphocyte-21.40%), Platelet - 53000, urea-49, creatinine-.81, Albumin- 2.5gm/dl, alanine aminotransferase (ALT)-53 U/L, aspartate aminotransferase(AST)-152 U/L, Na-122 mEq/L, K- 4.3mEq/L.

Tests for Malaria, Dengue and typhoid were performed as per routine investigations for fever, all of which were negative. Blood culture revealed no growth. Urinalysis was normal and urine culture revealed no growth.

Lumbar puncture report was suggestive of aseptic meningitis: clear CSF with 32 cells (70 lymphocytic), glucose and protein were normal range. A CSF analysis for *O.tsutsumugamushi* could not be performed as it was not available in our hospital. Blood for Scrub typhus was IgM-ELISA Positive. Magnetic resonance imaging brain showed no abnormality.

The patient was initially given intravenous fluid and empirical antibiotic as per standard recommendations, after blood samples were drawn and lumbar puncture was done. On receiving the Scrub typhus reports, we started oral doxycycline 4 mg/kg/day which led to dramatic improvement in 48 hours, with steady improvement of gait. Doxycycline was continued for a total of 7 days, following which the general condition improved and child became afebrile. We then discharged the child after a stay of 8 days after documenting the favorable neurological outcome.

2. Discussion

Scrub typhus, a disease which is transmitted by the bite of larva (chiggers) of trombiculid mites. Man is an accidental host, as these mites generally bite rats and other small mammals. Transovarial transmission among mites is the major mechanism of maintenance in nature. Scrub typhus usually presents with fever, headache, muscle pain, sore

throat, abdominal pain and sometimes Escher.¹ Eschar is pathognomonic and is present in 40%–50% of cases. It is usually located at site of Chigger bite such as the axilla, groin, waist and neck where clothes bind or skin folds.³ The presentation can be mild with a self-limiting clinical course with recovery in a few days or, rarely, severe disease with a protracted and fatal course.¹ Clinical manifestations include fever of 9 to 11 days duration, regional or generalized lymphadenopathy and GI symptoms such as abdominal pain, vomiting, and diarrhea. Complications include pneumonitis, meningoencephalitis, acute renal failure, disseminated intravascular coagulation and myocarditis.⁴

Among the various clinical manifestations, myocarditis and encephalitis are most life-threatening complications.⁵ 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. tsutsumugamushi* 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.⁶

Neurological manifestations such as Cerebellitis may be due to invasion of vascular endothelial cells by the organism itself. Autopsy of patients with scrub typhus have revealed focal vasculitis and lymphocytic infiltration of blood vessels.⁷

Most commonly used test in indirect immunofluorescence for detecting scrub typhus immunoglobulin M (IgM). It has 90% sensitivity when combined with history of 11 days plus fever.⁴

Microimmunofluorescence, latex agglutination, indirect hemagglutination, immunoperoxidase assay, and enzyme-linked immunosorbent assay (ELISA) remain the most important Serological tests for the diagnosis.⁸ 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.⁹

For treatment, the recommended regimen is Doxycycline 4 mg/kg/d in 2 divided doses for up to 7 days. Alternatives include azithromycin, clarithromycin, and chloramphenicol.¹⁰

Choi et al used steroids in a patient of scrub typhus with acutely progressive local neurologic symptoms and subsequently neurologic symptoms resolved, there is still no consensus of using corticosteroids in scrub typhus.¹¹ In our case the child improved without using of any corticosteroid.

Our findings are similar to the findings noted by Ghosh A et al.¹² and Kaiser et al.¹³

Early diagnosis of infection is important considering its complications and easily available treatment modality.

3. Conflict of Interest

The author declares no potential conflicts of interest with respect to research, authorship, and/or publication of this article.

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