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An Unusual Etiology of Cerebellar Ataxia

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Abstract

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. Scrub typhus very rarely presents itself with CNS manifestations. Here, we present the case of a 7-year-old girl from the Hooghly district in West Bengal, who presented to us with the history of fever, cerebellar signs, and sudden onset of visual loss. She was ultimately diagnosed with scrub typhus cerebellitis.

Keywords

ataxia, scrub typhus, cerebellitis

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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.[1] The causes of Cerebellar Ataxia can be congenital, metabolic, toxin related, neoplastic, vascular, infectious/post-infectious or inflammatory (cerebellar abscess, acute labyrinthitis, acute cerebellar ataxia, acute demyelinating encephalomyelitis. Infectious causes include Coxsackie, Diphtheria, Echovirus, Epstein–Barr, mumps, mycoplasma, rubella).[2]

SCRUB TYPHUS is a Rickettsial disease caused by *Orientia tsutsugamushi* that generally presents with nonspecific symptoms of fever, headache, and myalgia with signs of generalized lymphadenopathy and hepatosplenomegaly and in some cases meningitis and encephalitis. However, cerebellar involvement by this organism is very rare. In India, such a presentation has been reported previously on very few occasions.[3-5]

Discussion

Scrub Typhus is transmitted to mammals including humans by bite of the Chigger larvae of trombiculide mite. When the Chigger larvae feeds on skin, there is regurgitation of infected saliva. 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. In India, epidemics have been reported from northern, eastern, and southern states and from both rural and urban areas.

According to unpublished data obtained from our hospital and other institutes in the city, recently there seems to be a reemergence of this infection in both urban and rural areas, possibly due to more widespread presence of the organism and vector in the environment, poor living conditions in terms of people walking barefoot in villages, and people living in close proximity to rats. Also, there is a higher index of suspicion among physicians leading to routine testing of patients presenting with typical features.

Orientia causes infection of vascular endothelium, leading to maculopapular lesions, petechia or palpable purpura, and immune-mediated inflammation leading to end-organ injury in severe cases. Orientia also infects the reticuloendothelial system. Clinical manifestations include fever of 9 to 11 days duration, regional or generalized lymphadenopathy and GI symptoms such as abdominal pain, vomiting, and diarrhea. Single painless eschar may be seen at the site of Chigger bite (in 7%-68% cases) along with a generalized rash. Complications include pneumonitis, meningoencephalitis, acute renal

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failure, disseminated intravascular coagulation and myocarditis.[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]

Laboratory findings include leukocytosis (in 40% cases) and thrombocytopenia (one quarter to one-third cases), elevated transaminases, and hypoalbuminemia. 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.[6]

For treatment, the recommended regimen is Doxycycline 4 mg/kg/d in 2 divided doses for up to 7 days. This also applies for children who are less than 8 years (as tooth discoloration is dose-dependent). Alternatives include azithromycin, clarithromycin, and chloramphenicol.[8]

Case Report

A 7-year-old girl, from the district of Hooghly district in West Bengal, with no significant past history presented to us with fever for 15 days, difficulty in walking for last 4 days, and impairment of vision for 1 day. The fever was high grade (102.4°F on initial presentation) and intermittent in nature without chills, rigor, or rash. It used to subside on taking antipyretics. The fever was associated with headache and few episodes of projectile vomiting. Four days before presentation, the patient started having difficulty in walking with a tendency to fall to any one side and over the course of 4 days, she was not able to walk. Visual impairment was suddenly onset, with the child having difficulty in reading books and was associated with diplopia. The child had no history of spectacle use for refractive errors. There was neither any history of seizures, loss of consciousness, altered sensorium, trauma, low back pain, or any weakness nor of tinnitus, vertigo, or ear discharge. There was also no history of eye pain or redness. Bladder and bowel habit was normal. There was no history of similar disease in the family or of any developmental delay or regression. All immunizations were up-to-date, with no recent history of vaccination.

On further evaluation, child was febrile, irritable, and sick looking. There was no other abnormal finding on general survey, no apparent rash or neurocutaneous marker noted. No eschar was noted after thorough examination of entire body surface, including flexural areas, external genitalia, and scalp. Pulse was 120/min, with no special character. Blood pressure was 117/80, which is just above the 95th percentile for age, sex, and height. Respiratory rate was 24/min.

Neurological examination: Higher functions were normal with no impairment in memory, speech, or language. 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. Visual acuity testing could not be done initially as the child was not cooperative. On reexamination after stabilization, visual acuity testing was 6/6 in both unaided eyes. Cranial nerve VI was affected, with latent squint of right eye manifested on asking the child to look toward the right. We suspected that the apparent impairment of vision and abducens nerve involvement were due to raised intracranial tension. All other cranial nerves were within normal limit. Sensory system examination revealed no impairment in fine touch, pain, temperature, position, and vibration sense. Romberg test was within normal limit. 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 papilledema. There was no organomegaly on abdominal examination. Cardiovascular and respiratory systems were within normal limit.

Investigations: There was moderate anemia with Hb—7.2 g/dl with microcytosis and hypochromia, probably due to nutritional iron deficiency. Leukocytosis was present (TLC– 15010/cmm), with neutrophilic predominance (N62 L32 M4 E2 B0). Malarial parasite was negative on peripheral blood smear; malarial parasite dual antigen testing was negative. Liver function and kidney function tests and electrolytes were also normal for age and sex.

Tests for dengue and typhoid, Japanese encephalitis, Herpes simplex, and human immunodeficiency virus 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. Blood for Scrub typhus was IgM-Positive.

Cerebrospinal fluid– Cell type and count– 102 cells/cmm, 92% being mononuclear, 8% being polymorphs. Glucose -59 mg/dL. Cerebrospinal fluid protein was elevated to 119 mg/dL.

Cerebrospinal fluid Gram' stain showed no white blood cells, no microorganisms, and no growth after 48 hours incubation. Cerebrospinal fluid serology revealed Scrub typhus IgM/immunoglobulin G Positive. Magnetic resonance imaging brain showed no abnormality.

The patient was initially given intravenous fluid and antibiotic injection, Ceftriaxone as per standard recommendations on an empirical basis, after blood samples were drawn and lumbar puncture was done. We also started management of raised intracranial tension with head end elevation, control of fever with antipyretics, and hypertonic saline infusion at 0.5 mL/kg/h. This was continued for 24 hours till there was resolution of irritability, hypertension, improvement of vision, and subsidence of squint. On receiving the Scrub typhus reports, we started oral doxycycline at 5 mg/kg/d which led to dramatic improvement in 48 hours, with steadying of gait. Doxycycline was continued for a total of 7 days, following which the general condition improved and child became afebrile. She now had a visual acuity of 6/6, no sensorimotor or cognitive deficit and no problem with balance and coordination. We then discharged the child after a stay of 8 days after documenting the favorable neurological outcome.

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Ethical Approval

As this is a case report, ethical approval was not taken. Care has been taken to avoid any breach of privacy of the patient.

References

1. Morrison P, Mink J. Ataxias. In: *Nelson Textbook of Pediatrics*. 21st ed. Vol 2; 2019:3150.
2. Morrison P, Mink J. Ataxias. In: *Nelson Textbook of Pediatrics*. 21st ed. Vol 2; 2019:3151. Table 615.2.
3. Ghosh A, Sharma S, Choudhury J. Acute cerebellar ataxia in a 3-year-old Bengali girl: a novel presentation of scrub typhus in pediatric age group. *Int J Contemp Pediatr*. 2017;4(7):652-654.
4. Bhoil R, Kumar S, Sood RG, Bhoil S, Verma R, Thakur R. Cerebellitis as an atypical manifestation of scrub typhus. *Neurology*. 2016;86(22):2113-2114. doi:10.1212/WNL.0000000000002717.
5. Mahajan SK, Sharma S, Kaushik M, et al. Scrub typhus presenting as acute cerebellitis. *J Assoc Physicians India*. 2016;64(2): 69-70.
6. Reller M, Dumler J. Scrub typhus (*Orientia tsutsugamushi*). In: *Nelson Textbook of Pediatrics*. 21st ed. Vol. 1; 2019:1628.
7. Didel S, Basha MA, Biswal M, Suthar R, Sankhyan N. Acute cerebellitis in a child with scrub typhus. *Pediatr Infect Dis J*. 2017;36(7):696-697. doi:10.1097/INF.0000000000001524.
8. Chanta C, Phloenchaiwanit P. Randomized controlled trial of azithromycin versus doxycycline or chloramphenicol for treatment of uncomplicated pediatric scrub typhus. *J Med Assoc Thai*. 2015; 98(8):756-760.