Acute cerebellar ataxia associated with enteric fever in a child: a case report

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SUMMARY: İncecik F, Hergüner MÖ, Mert G, Alabaz D, Altunbaşak Ş. Acute cerebellar ataxia associated with enteric fever in a child: a case report. Turk J Pediatr 2013; 55: 441-442.

Enteric fever is a common infectious disease of the tropical world. Characteristic presenting features include fever, relative bradycardia, diarrhea or constipation, and abdominal pain. Central nervous system involvement is not rare and has a wide spectrum of presentation in enteric fever. Complications such as meningism, delirium, coma, and convulsions have been reported often. However, isolated acute cerebellar ataxia associated with enteric fever is rare.

Here, we report a seven-year-old boy with enteric fever who presented with acute cerebellar ataxia. Following treatment with appropriate antibiotics, the patient showed complete recovery over the next four weeks.

Key words: enteric fever, ataxia, children.

Enteric fever is caused by Salmonella group organisms. It is a common infectious disease of the tropical world, with about 80% of cases occurring in Asian countries¹. Fever, anorexia and headache are common symptoms of the disease. Abdominal pain, diarrhea, constipation, and myalgia are other known symptoms.

Central nervous system involvement is a recognized complication of enteric fever, with the incidence varying from 5-35% in adults and 2.1-2.7% in children². Various well-known neuro-psychiatric manifestations include confusional state, encephalopathy, meningism, convulsions, and focal neurological deficits³. Cerebellar involvement is rare. In the literature, isolated acute cerebellar ataxia associated with enteric fever has been reported in only a few case reports^{4,5}.

Here, we report a case of enteric fever who presented with acute cerebellar ataxia.

Case Report

A seven-year-old boy admitted to our hospital with a complaint of instability of gait and frequent falls while walking for five days before admission. For the previous 12 days, he had complained of fever, headache, diarrhea, and abdominal pain.

On his physical examination, the vital signs including pulse rate and blood pressure were normal. He had bilateral cerebellar signs in the form of hypotonia of all the limbs, dysarthria, nystagmus, bilateral dysdiadochokinesia, and bilateral finger-nose and heel-knee incoordination. He also had marked ataxia (trunk, limbs). The deep tendon reflexes were normal and plantars were flexor. Cranial nerves were intact, and there were no signs of meningeal irritation. The remainder of the physical and neurological examination was unremarkable.

In the laboratory studies, hemoglobin, total and differential white cell counts, erythrocyte sedimentation rate, serum urea, creatinine, and urinary sediment analyses were in normal limits. His cerebral magnetic resonance imaging was normal. Lumbar puncture was performed, and cerebrospinal fluid (CSF) was acellular, and on culture it was sterile. Abdominal ultrasonography was also normal. A Gruber-Widal test done at the time of admission showed antibody titer of 1:1320 for both O and H antigens.

The diagnosis of enteric fever complicated by cerebellitis was made on the basis of the clinical findings and the laboratory studies. The patient was treated with intravenous ceftriaxone 100

Discussion

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Central nervous system involvement is not uncommon in patients with enteric fever. Neurological involvement has a wide spectrum of presentation. Meningism, delirium, coma, and convulsions have been reported often. Other neurological manifestations such as acute transverse myelitis, isolated cranial nerve palsies, focal neurological deficits, optic neuritis, Guillain-Barré syndrome, and Bickerstaff's brainstem encephalitis are found rarely in the literature^{3,6}.

Acute cerebellar ataxia as an isolated neurological complication of enteric fever is very rare and limited to only a few case reports^{4,5}. Most of the neurologic complications occur during the second week, but it may manifest within the first few days of illness⁷.

In the literature, Kalra et al.⁴ reported a patient with enteric fever who developed acute cerebellar ataxia during the second week of illness. In another report, Dewan et al.⁸ reported a seven-year-old boy with enteric fever who presented with cerebellar ataxia on the 2nd day of fever. Sawhney et al.⁹ reported cerebellar involvement in three patients on the 2nd or 3rd day of enteric fever in adult patients. In our case, ataxia developed in the first week of the illness. Cerebellar signs are most likely to appear during the second week of illness, but as in our case, it may appear earlier.

The exact pathogenesis of this complication is unknown. Metabolic disturbances, toxemia, hyperpyrexia, and non-specific central nervous system changes such as edema and hemorrhage have been hypothesized. Because of its acute onset, self-limiting course and CSF pleocytosis, a para- or post-infectious demyelinating process has been postulated as the mechanism for ataxia¹⁰.

Management of enteric fever with ataxia requires only appropriate antibiotics. The cerebellar symptoms do not warrant any specific treatment (including corticosteroids). However, some have also recommended use of dexamethasone as an adjunct to antibacterial therapy in patients with neurological complications¹¹. Kang et

al.¹² reported the success of high doses of intravenous dexamethasone together with antibiotics in the treatment of a patient presenting with cerebellar ataxia. We used only ceftriaxone, and his symptoms recovered successfully.

This case highlights the fact that enteric fever with cerebellar ataxia can be very important and the diagnosis should be suspected in febrile children presenting with ataxia and/or acute neurological symptoms.

REFERENCES

- Park K. Typhoid fever. In: Park's Textbook of Preventive and Social Medicine (19th ed). Jabalpur, India: Banarsidas Bhanot; 2007: 195-198.
- Biswal N, Mathai B, Bhatia BD, Srinivasan S, Nalini P. Enteric fever: a changing perspective. Indian Pediatr 1994; 31: 813–819.
- 3. Osuntokun BD, Bademosi O, Ogunremi K, Wright SG. Neuropsychiatric manifestations of typhoid fever in 959 patients. Arch Neurol 1972; 27: 7-13.
- Kalra OP, Agrawal NK, Agarwal S. Acute reversible cerebellar ataxia in typhoid fever. J Indian Acad Clin Med 2002; 3: 96–97.
- 5. Thapa R, Banerjee P, Akhtar N, Jain TS. Transient dysautonomia and cerebellitis in childhood enteric fever. J Child Neurol 2008; 23: 1081-1082.
- 6. May W, Senitiri I. Guillain-Barré syndrome associated with typhoid fever. A case study in the Fiji Islands. Pac Health Dialog 2010; 16: 85-88.
- Lakhotia M, Gehlot RS, Jain P, Sharma S, Bhargava A. Neurological manifestations of enteric fever. J Indian Acad Clin Med 2003; 4: 196–199.
- 8. Dewan P, Pooniya V, Kaushik JS, Gomber S, Singhal S. Isolated cerebellar ataxia: an early neurological complication of enteric fever. Ann Trop Paediatr 2009; 29: 217-219.
- 9. Sawhney IM, Prabhakar S, Dhand UK, Chopra JS. Acute cerebellar ataxia in enteric fever. Trans R Soc Trop Med Hyg 1986; 80: 85-86.
- Scragg J, Rubidge C, Wallace HL. Typhoid fever in African and Indian children in Durban. Arch Dis Child 1969; 44: 18–28.
- 11. Girgis NI, Farid Z, Sultan Y, Hibbs RG. Use of dexamethasone in treatment of enteric fever complicated by cerebellar involvement. Trans R Soc Trop Med Hyg 1993; 87: 690–691.
- 12. Kang JK, Hwang YM, Lee MC, Song JH, Lee YS. Two cases of suspected typhoid encephalitis treated with high dose dexamethasone. J Korean Neurol Assoc 1993; 11: 229-234.