CASE REPORT

ENTERIC FEVER PRESENTING AS PALATAL PALSY, CEREBELLAR ATAXIA AND SEVERE THROMBOCYTOPENIA: A CASE REPORT

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ABSTRACT

Enteric fever, being a systemic infection is associated with a variety of clinical presentations, multisystem involvement and complications. Although central nervous system involvement is not uncommon, acute cerebellar ataxia and palatal palsy as a presenting feature are rare. A 13 year old girl with enteric fever who presented with acute cerebellar ataxia and palatal palsy with marked thrombocytopenia in early phase of illness is being reported. This atypical presentation is not common in enteric fever in early course of the disease and can lead to misdiagnosis as well as a delay in the initiation of appropriate therapy. Prompt clinical improvement and the return of platelet counts to normal were noted after the patient was started on intravenous ceftriaxone.

Key Words: Enteric fever; cerebellar ataxia; thrombocytopenia; ceftriaxone; palatal palsy.

INTRODUCTION

Enteric fever is a systemic infection which can present in a number of ways. Neurological involvement is not uncommon, manifesting as meningism, delirium, ataxia, coma or convulsions and occurs mainly in the second to third week. Acute abdomen, intestinal perforation, pneumonia are likely to develop in the third to fourth week. We report a case presenting with acute ataxia, palatal palsy and marked thrombocytopenia in the first week of illness.

CASE REPORT

A 13 year old girl presented with fever for six days with vomiting and watery diarrhea. Since the past four days she developed unsteady gait, difficulty in walking, maintaining balance and holding objects. She had difficulty in swallowing with slurring of speech followed by complete aphasia. There was no history of headache, seizures or posturing. Bladder habits were normal. On initial examination, she was febrile with stable vitals. There was no rash or rose spots. On central nervous system examination (CNS), she was confused. There was palatal palsy, gag reflex was absent. There was generalized hypotonia. Power was 4/5 in all four limbs. There was no focal neurological deficit, reflexes were normal with pendular knee jerk. Plantar reflex was flexor. Cerebellar signs were present. Sensory system was normal. Neck rigidity was present. Fundus examination was normal. Rest systemic examination was normal. She had received some injectables for two days before being referred to our college. A differential diagnosis of acute viral meningoencephalitis and enteric fever with complications was kept. She was started on injections Acyclovir and Ceftriaxone. On investigating, the hemogram revealed pancytopenia with a hemoglobin of 7.5 g/dl, total leucocyte count of 2,200/ul and platelet count of 9000/ul. Peripheral smear revealed microytic anemia, no malarial parasites. Liver function tests showed an albumin of 3.3 g/dl, total bilirubin 0.6mg/dl, ALP 47 iu/l, SGOT 1444 iu/l, SGPT 395 iu/l). Cerebrospinal fluid analysis revealed proteins 56 mg%, there were 5 cells, all lymphocytes. Blood sugar, renal functions and electrolytes were normal. Weil felix test for Scrub typhus was negative. Widal test showed high titres of TO> 360 and TH > 360. CT scan and MRI were normal. Acyclovir was stopped after two days while ceftriaxone was continued for 14 days. The patient was completely oriented within 48 hours, became afebrile after 72 hours. She was started on tube feeds. By day six, she started speaking which was scanning with nasal twang. Meningeal signs disappeared, palatal palsy improved and she started swallowing semisolids. She started walking on the 8th day with support and without support by the 10th day. Her platelet and total leucocyte counts returned to normal after three days. Her blood culture was sterile. After two weeks, there was complete recovery of palatal palsy and speech improved and she was eating well orally. Cerebellar signs and nasal twang improved but persisted till discharge. On followup visit at four weeks, she was neurologically normal. A repeat widal test done at this time showed TO < 160 and TH = 80.

DISCUSSION

Complications in enteric fever occur mostly during the second week. The predilection of typhoid toxin for central nervous system is known. Any part of the central nervous system may be affected . Enteric fever presenting as acute cerebellar ataxia and palatal palsy in early course of illness is rare and only few cases have been reported. Recovery with appropriate treatment is often complete but may take as long as two weeks.[1] Our patient took about 4 weeks to recover. Op Kalra, etal [2] reported a similar case in a 19 year old Indian patient. In a large study, Scragg *et al*[3] examined 316 African and Indian children with enteric fever, but did not encounter a single case of palatal paralysis. Systematic literature review carried out by G Jombo et al [4] compiling all relevant articles over the past 30 years, showed only 0.1% incidence of palatal palsy. Widal test was significant in 17.3% and blood culture was positive in only 10.8% patients. Complete recovery is expected following successful treatment of the underlying infection. Our case also presented with anemia, thrombocytopenia and leucopenia. Toxic marrow suppression is believed to be a cause for this. There are no clear recommendations on the duration of therapy in cases presenting with cerebellar ataxia or palatal palsy. We treated our patient for 14 days with good recovery. The above case emphasizes the need to re-look at enteric fever as an entity as it mimics a number of other possible diseases. A high index of suspicion and wide knowledge of the atypical presentations is required, especially in endemic areas. The "Gold Standard" for diagnosis of enteric fever is the isolation of bacteria from blood or bone marrow. However for diagnosing enteric fever, widal test is the second most widely used.[5] The widespread and uncontrolled use of antibiotics leads to negative results, [5] which may have been the cause of negative culture in our patient too. This patient showed a strongly positive widal test at admission which normalized after successful treatment. Sensitivity of single tube widal test is not very high. But the test is still valuable when a high cut-off titer is taken in developing countries like India. Titres of 160 for anti-O and 320 for anti-H were considered diagnostic for enteric fever in Chandigarh in North India.[6] Similar diagnostic 'O' and 'H' titres have been reported recently by others.[7] The high titers (>1/160) for Salmonella typhi "O" and (>1/320) for Salmonella typhi "H" antigens performed on single acute-phase sera must be considered as significant and diagnostic. There were three cases of enteric fever with blood culture positive for salmonella typhi during the same month, which suggests endemicity of the disease in our area.

CONCLUSION

With emergence of multidrug resistant enteric fever and high risk of complications, we suggest that enteric fever should be considered in the differential diagnosis of cerebellar ataxia, palatal palsy in a febrile child.

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