Case Report

Acute disseminated encephalomyelitis in a child with enteric fever: A case report

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Abstract

The central nervous system is involved in 5-35% children suffering from typhoid fever and manifest as neuropsychiatric features such as confusion, encephalopathy, meningismus, convulsion and focal neurological deficit. However, only few cases of acute disseminated encephalomyelitis (ADEM) have been reported along with enteric fever. ADEM is a monophasic immune-mediated demyelination disorder, which commonly follows viral infections and vaccination. We here report a case of typhoid fever who later developed ADEM.

Key words: *Acute Disseminated Encephalomyelitis, Central Nervous System, Demyelination, Typhoid fever*

Enteric fever is endemic in developing countries where there is scarcity to access to portable water along with poor waste water and sewage disposable system. 5-35% the children suffering from typhoid fever may develop central nervous system (CNS) manifestations such as confusion, encephalopathy, meningismus, convulsion, and focal neurological deficit [1-3]. However, acute disseminated encephalomyelitis (ADEM) is rarely reported following typhoid fever, especially in the pediatric population, and we found only few reports after thorough literature search [4,5]. Therefore, we report a case of an adolescent boy who developed ADEM following the course of typhoid fever.

CASE REPORT

A 14-year-old male child was brought to the casualty with the complaints of fever with chills for 14 days, headache for 3 days, nausea and not accepting orally since last 2 days. There was a history of the passage of loose stools black in color since last 3 days. On examination, his Glossgow coma scale was 9/15, had fever 103°F, pulse rate 150/min, respiratory rate 25/min, capillary refill time 3 s, blood pressure 100/70 mmHg in right upper arm with signs of moderate dehydration. His pupils were of normal size reacting normally to light, pallor was present. On CNS examination, he had generalized the hypertonia with brisk deep tendon reflexes (DTRs) with clonus and bilaterally extensor planter reflexes. His speech was slurred and was talking incoherently. Signs of meningism were present. Rest of the systems was normal. Provisional diagnosis of meningoencephalitis with moderate dehydration was kept, and child was admitted in post intensive care unit. Fluid resuscitation was given and treatment in the form of intravenous antibiotics (ceftriaxone and amikacin), paracetamol, ranitidine was started.

On investigation, his hemoglobin was 7.3 g%, packed cell volume 22.4%, total leukocyte count 1800/mm^3, P 55%, L 41%, E 2%, M 2%, and platelets 1.07 lakhs. Peripheral smear showed microcytic hypochromic picture with few macrocytes. Liver function tests showed normal serum bilirubin (total 1.0 mg/dl, direct 0.4 mg/dl, and indirect

0.6 mg/dl) raised serum glutamic oxaloacetic transaminase (839 IU), and serum glutamic pyruvic transaminase (162 IU) with serum albumin of 3.1 mg/dl, globulin 2.6 mg/dl, and A: G ratio of 1.19. His coagulation profile was also normal with normal serum electrolytes (sodium - 137 mEq/L and potassium - 3.8 mEq/L). Dengue serology and malarial antigen were negative. Stool for occult blood was positive with normal urine routine microscopy. CSF study revealed no significant abnormality. Widal test was positive 1/320 positive for O antigen and 1/320 for H antigen, although, blood culture for salmonella was negative. Computed tomography scan brain plain was normal. Diagnosis was revised as enteric encephalopathy and intravenous Dexamethasone 3 mg/kg stat followed by 1 mg/kg/kg/dose 6 hourly for 2 days was started.

However, the child continued to have slurred speech and hypertonia. DTR was brisk with clonus. In view of these persisting neurological signs, MRI brain was done, which revealed demyelination at the junction of gray and white matter at the parietal area bilaterally, which was suggestive of ADEM. Intravenous Methylprednisolone 30 mg/kg/day was given for 5 days. Gradually, the child started showing signs of improvement in the form of improvement in speech, as well as decrease in tone in all the limbs. The child was discharged on the 14th day after admission with normal speech and normal neurological examination findings.

DISCUSSION

ADEM is an immune-mediated inflammatory disorder of the CNS characterized by a widespread demyelination that predominantly involves the white matter of the brain and spinal cord. It usually results from viral infections such as chicken pox, mumps, cytomegalovirus, Ebstein-barr virus, Herpes simplex virus and measles and vaccination (MMR, DPT, and rabies) [6,7]. However, ADEM following typhoid fever is a rare phenomenon. Recently, two different cases have been reported where ADEM was associated with enteric fever.

The first case was from Nepal, where a 7-year-old girl with cerebellar signs suffered from enteric fever about one month before the onset of presenting symptoms. Her MRI brain revealed diffuse cerebral and cerebellar involvement suggestive of ADEM [4]. In the second report, 2 cases of ADEM were reported from Nigeria following enteric fever. First one of these cases was a 7-year-old girl who was operated for the enteric perforation and during post-operative recovery period, she developed cerebellar signs along with aphasia and visual disturbances. Her MRI revealed large globular lesions in subcortical white matter. The second case was a 12-year-old boy who was receiving treatment for enteric fever and during the course of treatment developed generalized convulsions with features of raised intracranial tension and expired during treatment. His postmortem findings were suggestive of widespread demyelination in midbrain, pons, and cerebellum [5].

Our case also had slurred speech, hypertonia, clonus and meningism. His MRI brain revealed the demyelination at the junction of white and gray matter in the parietal area on both the sides. Our case did not respond to initial high doses of dexamethasone, but showed a rapid response to methylprednisone thus favoring the diagnosis of ADEM. The exact pathogenesis of CNS involvement in typhoid fever is not clear. Factors that are implicated include hyperpyrexia, hypovitaminosis, metabolic derangements, toxemia, and cerebral edema, and non-specific inflammatory changes in cerebral vessels such as capillary thrombosis, hemorrhage, edema, and perivascular infiltration associated with widespread demyelination [2,3]. In our case, there was no metabolic derangement, and anemia was due to poor nutritional intake.

ADEM can present with a varied spectrum of features, irritability and lethargy being common first signs and fever and headache in about half of the patients [8]. The common neurologic symptoms are visual field deficits, language disturbances, and mental status abnormalities ranging from irritability, lethargy to coma, and psychiatric changes including depression, personality changes and pyschosis. Meningeal signs are reported in children with severe disease. Weakness that may be hemiparetic or generalized and symmetric is more commonly detected than sensory defects. Other reported symptoms are cranial nerve palsies, generalized or focal seizures and ataxia [6-8]. These clinical features overlap with enteric encephalopathy. Therefore, MRI brain is essential to confirm the diagnosis of ADEM.

MRI findings in typhoid encephalopathy include bilateral symmetrical hypodensity with slight hypertrophy of gyri and diffuse cerebral edema without any focal lesions [9]. Use of intravenous methypredisone or dexamethasone in ADEM usually leads to improved recovery and reduced disability. Patients not responding to steroids are benefitted by the use of immunoglobulins and plasmapheresis [10]. The survival rates in patients with ADEM is usually high, especially if they are able to overcome the initial acute complications like raised intracranial tension and metabolic derangements.

CONCLUSION

ADEM though a rare complication of typhoid fever should be thought of in children suffering from enteric fever with mental status abnormalities, language disturbances and visual field defects.

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