Acute Disseminated Encephalomyelitis Following Typhoid Fever: A Case Report

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ABSTRACT
The involvement of central nervous system in children with typhoid fever is common. Acute disseminated encephalomyelitis is a rare immune mediated and demyelinating disease of the central nervous system that usually affects children. We report a 7-year-old child with typhoid fever who developed acute cerebellar syndrome due to acute disseminated encephalomyelitis.

Keywords: acute disseminated encephalomyelitis; acute cerebellar syndrome; typhoid fever

INTRODUCTION
Typhoid fever still remains a major public health problem, with global annual incidence of about 21 million cases.1 Affected children typically present with fever, anorexia, headache, abdominal pains, diarrhoea, constipation and myalgia. However, central nervous system (CNS) involvement in typhoid fever is an important atypical presentation in childhood. Acute disseminated encephalomyelitis (ADEM) is a rare immune mediated and demyelinating disease of the brain and spinal cord and can be defined as scattered focal or multifocal inflammation of CNS.2 ADEM is a monophasic disease that usually develops following acute viral infection, vaccination or organ transplantation.3,4 This disease entity as a complication of typhoid fever is very rare and only few cases had been reported.5 Here we report a case of ADEM in a 7-year-old child with diagnosed typhoid fever, acute cerebellar syndrome and magnetic resonance imaging (MRI) evidence of ADEM.

CASE REPORT
A 7-year-old female child was admitted with acute onset difficulty in walking, and dysarthria. Symptoms got progressively worse over a period of 2 days. She has had a febrile illness four weeks prior to the present illness. That time a Widal test was positive with a Salmonella typhi H titer of more than 1:160 and she was diagnosed to have typhoid fever. On admission she had nystagmus, cerebellar and pyramidal tract signs and gait ataxia. The physical examination showed the patient’s vital signs stable. Her past medical history was unremarkable. A lumbar puncture was performed. Cerebrospinal fluid (CSF) revealed predominant lymphocytosis, adenosine deaminase (ADA) of 2.4 U/L (normal <10 U/L), a protein content of 33.8 mg/dL, and a glucose content of 55 mg/dL. The CSF culture showed no growth of bacteria.

Computed tomography (CT) scan brain revealed ill-defined areas of hypodensity in the right
cerebellar hemisphere. MRI brain scan revealed hyperintense areas involving bilateral middle cerebellar peduncle, right cerebellar hemisphere, pons, right side of mid brain, bilateral periventricular and subcortical white matter in T2 weighted (T2WI) and Fluid-attenuated inversion recovery (FLAIR) showing open ring enhancement in T1 weighted post contrast images (Figure. 1, Figure. 2, and Figure. 3). Corpus callosum was intact.

The clinical picture and the MRI scan findings were suggestive of acute disseminated encephalomyelitis (ADEM). She was started on high dose IV steroids. She made a dramatic improvement over the next few weeks and was able to walk well at the end of four weeks. On discharge, the patient was fully relieved from all previous symptoms.

DISCUSSION

ADEM is a rare acute autoimmune mediated disease characterized by a widespread demyelization that predominantly involves the white matter of the brain and spinal cord. It is an inflammatory process manifested by rapid onset of multifocal neurological impairment. ADEM usually develops following acute viral infection, vaccination or organ transplantation over days, weeks or months, punctuated by an acute worsening
and is more frequent in children [3,4]. However, the present case report like few others showed that it can also be caused by bacterial infections such as typhoid fever.

The true incidence of ADEM is unknown. In one study, ADEM almost involves children with the incidence rate of 0.4/100000 yearly in patients less than 20 years-old. The peak incidence of ADEM is among 5 to 8 years of age. Male to female ratio is the same.

Imaging is a valuable diagnostic tool. CT and MRI are worthwhile tools in establishing the diagnosis of ADEM. CT is often normal at first, but changes during 5-14 days later. CT changes include: multifocal lesions in subcortical zone of white matter with low attenuation. Of course MRI is the most extremely diagnostic tool because demyelinating lesions are better seen in MRI. Cerebellum and brain stem involvement are common. MRI characteristics include patchy lesions with severe increase of signals in conventional T2-WI and FLAIR. Although, white matter is the most common place of involvement, gray- matter especially basal ganglia, thalamus and brain stem can be involved. Corpus callosum is usually intact and its involvement is suggestive and has the characteristic of Multiple Sclerosis (MS). The other variables are cerebrospinal fluid (CSF) changes; often include increased CSF pressure, raised protein and lymphocytic pleocytosis. Glucose is usually in a normal range.

Multiple sclerosis (MS) is the main differential diagnosis of ADEM. In contrast to MS, ADEM is usually a monophasic disorder with favorable long term prognosis. Monophasic ADEM is more common in children whereas MS is more common in adults. It has been shown that the mean age was 7.1 years and 12 years in ADEM and MS patients respectively. However, history of typhoid fever, high load of lesions in MRI, early onset ataxia, deep cortical gray matter involvement and intact corpus callosum in our case highly suggested ADEM. This patient was definitely suffering from typhoid fever in the early stage, but subsequently it was complicated by the development of ADEM. After the MRI was scanned, the diagnosis of ADEM was made. Moreover, the diagnosis was also supported by the CSF findings (lymphocytic pleocytosis).

**CONCLUSION**

Typhoid fever is associated with several neurological complications including post-infectious demyelination as the underlying pathological process. It is usually reversible; however, it may be lethal if left untreated. Since clinical diagnosis of ADEM may not be clear initially and in the absence of brain MRI scan, pediatricians, particularly those who work in low resource countries like ours, should consider this diagnosis in children with typhoid fever associated with cerebellar dysfunction.

**REFERENCES**

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