Case Report

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Early Diagnosis and Treatment not Always a Key to Favorable Outcome: A Case Report of ADEM Correctly Diagnosed and Treated Still Surviving for Better Life

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Abstract

Acute disseminated encephalomyelitis (ADEM) is a rare disease of central nervous system with a spectrum of presentation. It is a diagnosis of exclusion and relies on neuroimaging which may be normal at the onset. It is a diagnostic challenge at its first attack. The disease is although more common in children it can invariably be present in adults. Here we present a case report of ADEM in a 30 year old female who presented to ER with history of multiple episodes of vomiting followed by sudden onset of dysphasia and other neurological complaints. The patient had a history of recent travel to a pilgrimage where she had enteric fever around 15 days ago. It was our neurology team which correctly recognised and treated it as ADEM. The patient responded well to the treatment and discharged in stable condition after 5days. Sadly the disease had a relapse which now showed no response to iv immunoglobulins, steroids or plasmapheresis. The patient was in the hospital for symptomatic management and is still surviving in the hope of a normal well being.

Keywords: Acute Disseminated Encephalomyelitis; Central Nervous System; Neuroimaging.

Introduction

Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating disease of the central nervous system. Its onset is acute and often rapidly progressive. It is traditionally mono phasic but some patients may have recurrences.

ADEM typically presents with multifocal neurological signs, including motor, sensory, cranial nerve, brainstem deficits as well as nonspecific symptoms such as headache, malaise and altered mental status.

The diagnosis is supported by the presence of one or more supratentorial or infratentorial demyelinating lesions in the brain on magnetic resonance imaging (MRI) and the absence of destructive black hole lesions on T1-weighted MRI. Abnormal cerebrospinal fluid findings such as mild lymphocytic pleocytosis and slightly elevated protein level are suggestive of ADEM.

More than half of patients have an illness, usually an infection, two to four weeks before developing ADEM. Most of these illnesses are viral or bacterial. In children with ADEM, prolonged and severe headaches occur. In addition the patient develops fevers during the ADEM course.

Along with this pattern, the patients usually get neurological symptoms which may include:

- Confusion, drowsiness and even coma
- Unsteadiness and falling
- Visual blurring or double vision
- Trouble swallowing
- Weakness of the arms and legs

In adults with ADEM, motor (movement) and sensory (tingling, numbness) symptoms tend to be more common. Overall what triggers a diagnosis of ADEM is a rapidly developing illness with neurological symptoms often with fever and headache

usually following an upper respiratory tract infection and which has significant MRI and spinal fluid findings consistent with ADEM.

Case Report

30 year old female brought by attendants with history of multiple episodes of vomiting followed by sudden onset of dysphasia.

On examination in emergency her vitals were HR-88/m, BP-130/80mm Hg, RR-18/m, T-99 F, RBS-140mg/dl with patent airway and bilateral equal air entry.

Secondary examination was all normal except CNS which revealed GCS E4V1M6, planters bilateral mute, right sided neck dystonia and reflexes all limbs 2+.

The attendants gave a history of recent travel to some pilgrimage around 15 days ago where patient had complaints of loose watery stools and vomiting. She was diagnosed as enteric fever and managed symptomatically.

All routine investigations were sent from the ER and MRI brain planned. The blood reports revealed elevated TLC levels. MRI brain showed multiple demyelinating lesions in bilateral cerebral hemisphere. CSF was acellular with high protein. A diagnosis of ADEM was made.

Patient was admitted under Neurology team and treatment was started accordingly.

Patient received high dose of steroids, immunoglobulins, iv fluids, iv antibiotics. She gradually became better and discharged home in a stable conditions with advise for gradual ambulation.

After about one month, patient represented with complaints of mild remitting fever since 10 days, history of twisting of the tongue around 6 days ago. Weakness of right side of body since 1 day with decreased responsiveness since the day of readmission.

Again the vitals were normal, secondary examination was all normal except CNS which revealed GCS-E4V1M5, plantars bilateral extensor, hyper reflexia, power grade- Left side -5/5, Right side-1/5. Bilateral pupils mid dilated with sluggishly reaction. Repeat MRI revealed similar changes of severe ADEM with brain stem involvement.

Patient was again admitted under Neurology unit and was restarted on steroids, anti epileptics. Plasmapheresis was done but the patient's clinical condition gradually detoriated. The patient had decerebrate rigidity with severe hyperthermia for which she had been treated accordingly. The patient was sent home in the same state and advised symptomatic management.

It has been found that the family is still making all possible efforts but no response is noticed.

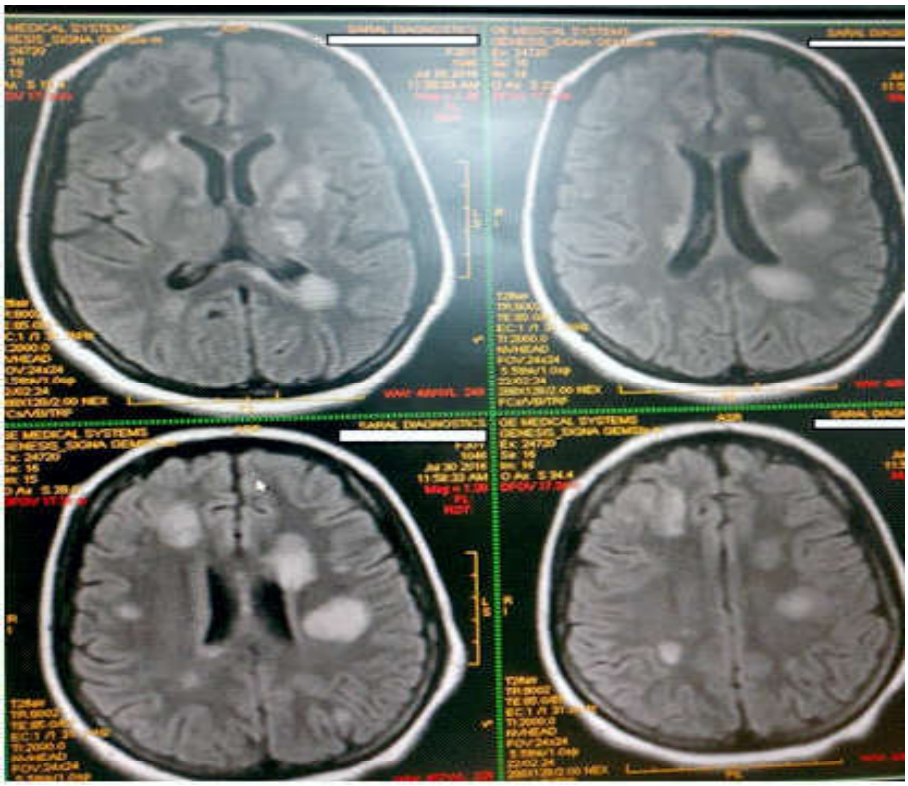

Fig. 1:

Discussion

Early diagnosis and management is definitely a key to every disease but the response it has on every individual is not unanimous. Here we had a 30 year old female who was correctly diagnosed and treated for ADEM. The results were favourable initially but the relapse showed no response to the appropriate management of the disease. It has been more than 8 months now that the patient is in a debilitated stage though every attempt is continued to make her live better.

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