Case Report:

Acute Disseminated Encephalomyelitis Presenting with Ptosis and Facial Palsy

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Abstract

Background: Acute disseminated encephalomyelitis (ADEM) is an acute demyelinating infection of the central nervous system (CNS) that is characterized by multifocal white matter involvement often following an infection or vaccination.

Case: An 11-year-old girl was hospitalized with complaints of acute abdominal pain for three days followed by ptosis and facial palsy. The child was diagnosed with ADEM after clinical, laboratory, and cranial MRI findings. An initial 5-days therapy with pulsed methylprednisolone, the child showed obvious clinical improvement, the treatment was continued and significant improvement was achieved. We report such case as a rare association with cranial nerves involvements with clinical presentation of ptosis and facial palsy.

Conclusions: ADEM is an inflammatory demyelinating condition that bears a clinical and pathological resemblance with MS. MRI is the premier modality of choice in its diagnosis.

Key Words: Acute disseminated encephalomyelitis – Ptosis – Facial palsy – Child – KSA.

Introduction

ACUTE disseminated encephalomyelitis (ADEM) is an inflammatory immune-mediated disorder of the CNS, characterized by diffuse demyelination that specially involves white matter, grey matter and spinal cord [1]. Its incidence is 0.2-0.8/100,000 children in USA and Canada and 0.7/100,000 children in Germany [2]. Although the disease has generally an acute and monophasic course, relapses have been reported and this represents a problem in differentiation between ADEM and other demyelinating immune-mediated neurological diseases such as multiple sclerosis (MS) [3-6]. Cerebrospinal fluid (CSF) analysis may show moderate mononuclear pleocytosis and moderate increase in protein [7].

We report this patient affected by ADEM seen in our hospital, Aseer central hospital.

Case Report

In December 2012, an 11-year-old Saudi girl from Abha, who was the fourth-born of both healthy and non-consanguineous parents, and not known to have any chronic medical illnesses presented to the Emergency Department of Aseer Central Hospital, Abha City. She had a history of severe epigastric pain for three days, which was acute and colicky in nature for 20-30 minutes in duration. The pain was severe enough to disturb her sleep, with no radiation, aggravating or relieving factors. It was associated with non-projectile vomiting and constipation for 3 days. There was no history of jaundice or abdominal distension, no history of respiratory symptoms, joint involvement, skin rash, or convulsion.

Physical examination revealed that the patient was looking unwell, in pain, conscious, alert, well hydrated and not in respiratory distress. Vital signs were normal including oxygen saturation. Her throat was mildly congested with no lymphadenopathy. Cardiovascular and respiratory systems examinations were unremarkable. The abdomen was mildly distended, with epigastric tenderness but no organomegaly. Bowel sounds were normal. Initial central nervous system examination, including cranial nerves was normal.

Laboratory investigations revealed normal complete blood count and erythrocyte sedimentation rate. Renal function tests, liver function tests and
amylase were within their normal ranges. Urine and blood cultures were negative. Other investigations including cytomegalovirus (CMV), Epstein-Barr virus (EBV), herpes simplex virus and HIV serologies came normal. Anti-nuclear antibodies, Anti-ds-DNA, C3 and C4 were normal.

Abdominal ultrasound was unremarkable. Abdominal computed tomography scan had been done to explore further underlying causes of abdominal pain and it showed normal study. At this stage, surgical causes had been excluded. The patient was admitted to the general pediatric ward for observation. She was started on proton pump inhibitor, laxatives and fleet enema with possibility of acute gastritis. Twelve hours later, the patient developed low grade fever and sudden frontal headache associated with left eyelid ptosis and left dilated pupil, no squint or nystagmus. Extra-ocular movement and fundus examination were normal. Visual acuity, fields and color vision were unremarkable. The meningeal signs were negative and other cranial nerves were normal. Motor and sensory examination of both upper and lower limbs, gait and coordination were normal. CT scan of brain showed normal findings. At that stage, lumbar puncture had been done and showed high oligoclonal band, high CSF protein (67mg/dl) with normal glucose and cell count.

The patient’s condition deteriorated on the 2nd day of admission when she developed frequent vomiting, progressive frontal headache and left eye ptosis as well as left facial nerve palsy. She had progressive difficulty of walking and speech. Repeated examination at that time showed exaggerated deep tendon reflexes and hypertonia.

MRI brain was done and showed multiple, widespread areas of abnormal signal intensity involving the white matter tracts at the frontoparietal region bilaterally. Further involvement of the body of corpus callosum was seen. Another focus was seen at the right side of the lower medulla oblongata. All these lesions were having poorly defined outline and presented a high signal intensity in T2-weighted images and FLAIR sequences and low signal intensity in T1-weighted images. No evidence of associated mass effect or any enhancement could be identified. Otherwise, no obvious mass lesions or extra-axial fluid collection were detected. Those findings were consistent with white matter demyelinating disease (Fig. 1).

The patient was started on methylprednisolone 30mg/kg/dose for 5 days. Thereafter, she started to improve gradually over 2 weeks period and continued on tapering dose of prednisolone for four weeks.

At time of discharge, all neurological sequelae, including ptosis, facial palsy, and difficulty in speech and walking had resolved. One month later, she was seen in the clinic with no complaint and normal neurological examination. Follow-up MRI brain showed resolved previous lesions in frontoparietal region, corpus callosum and medulla oblongata. No new lesions could be seen (Fig. 2).

Fig. (1): MRI Brain with multiple, widespread areas of abnormal signal intensity involving the white matter tracts at the frontoparietal region bilaterally (A). Further involvement of the body of corpus callosum is seen (B). Another focus is seen at the left side of the lower medulla oblongata (C).

Fig. (2): Follow-up MRI Brain showed resolved previous lesions in frontoparietal region, corpus callosum and medulla oblongata. No new lesions could be seen.
Discussion

This case presented during winter. This timing may reflect an infection mediated syndrome. ADEM most commonly presents during the months of winter and spring [3]. More than half of ADEM patients have an incidence of precipitating infection with a mean latency of approximately 2 weeks [4-5].

A large list of infections may precipitate ADEM, though isolation of a specific agent is frequently uncommon. The classic infection precipitants include measles, Epstein Barr virus, mycoplasma and Group A streptococcus. Multiple sclerosis is the main differential diagnosis. Although infection may precipitate multiple sclerosis relapse, the association with infection and seasonality is less pronounced. In contrast to multiple sclerosis, ADEM is usually a monophasic disorder with favorable long-term prognosis. Monophasic ADEM is more common in children whereas multiple sclerosis is more common in adults [8].

The pathogenesis of ADEM is thought to result from a transient autoimmune response towards myelin or other self antigens via molecular mimicry, or by non-specific activation of auto reactive T-cell clones. The lesions are of similar histological age and more numerous in white matter, but often involve the deep cortical lamina, thalami, hypothalamus and other gray matter structures [8].

This case is of an 11-year old girl. However, most series of ADEM have failed to show a sex predominance although some series show a mild male predominance [9].

The onset of symptoms in our case included severe epigastric colicky pain associated with non-projectile vomiting and constipation. Twelve hours after admission, the patient developed low grade fever and sudden frontal headache associated with left eyelid ptosis and left dilated pupil. The patient was conscious, meningeal signs were negative and neurological signs were normal.

However, Yadav and Agarwal [8] noted that, cases of ADEM are usually preceded by a prodromal phase of several days of fever, malaise and myalgia. Clinical features include multifocal neurological disturbances such as bilateral optic neuritis, visual field defects, aphasia, sensory and motor deficits, ataxia, movement disorders and signs of an acute meningo-encephalopathy with meningism such as depressed level of consciousness, focal or generalized seizures and psychosis.

Lumbar puncture of our case showed high oligoclonal band and high CSF protein with normal glucose and cell count. Murthy et al., [3] stated that in cases of ADEM, lab evaluation reveals that CSF is abnormal in 67% of patients and usually demonstrates increased pressure, proteins and white and red cells. However, Schwaz et al., [10] noted that in cases of ADEM, CSF only rarely shows intrathecal oligoclonal immunoglobulin (IgG) which invariably ceases as patients improve.

CT scan of the brain of our case showed normal findings. This finding is in agreement with that stated by Yadav and Agarwal [8], who noted that CT is usually normal at onset and later shows low attenuation multifocal lesions in subcortical white matter.

The diagnosis of our cases was mainly settled through MRI. MRI is usually the premier modality of choice in the diagnosis of ADEM. Long T1 and T2 values are associated with presence of inflammation, demyelination and hemorrhagic necrosis. The lesions in ADEM often have poorly defined margins, are usually located in the peripheral subcortical cerebral white matter [3,5,9].

Our case was managed by steroids for 5 days, after which she started to improve gradually over 2 weeks. Schwaz et al., [10] stated that spontaneous improvement has been repeatedly noted in ADEM. However complete recovery is less frequently seen in patients not receiving some form of treatment. High dose steroids have been found to be effective.

Conclusions:

ADEM is an inflammatory demyelinating condition that bears a clinical and pathological resemblance with MS. MRI is the premier modality of choice in its diagnosis.

References


