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Acute Disseminated Encephalomyelitis In A Middle Age Female

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ABSTRACT:

Acute disseminated encephalomyelitis (ADEM) is an autoimmune condition that predominately involve demyelination of the white matter of the brain and spinal cord. It resembles other acute demyelinating syndromes like multiple sclerosis and most often presents in childhood. This is a case of a 52-year woman, presented with lower limb weakness, confusion and spasticity throughout the body with no preceding illness. CSF showed increased proteins with no oligoclonal bands. MRI Brain showed abnormal high intensity areas in bilateral parietal, periventricular region, with no contrast enhancement. She was given 1gram iv methylprednisolone based on the diagnosis of ADEM and responded well.

KEYWORDS:

Acute disseminated encephalomyelitis, demyelination, multiple sclerosis, methylprednisolone

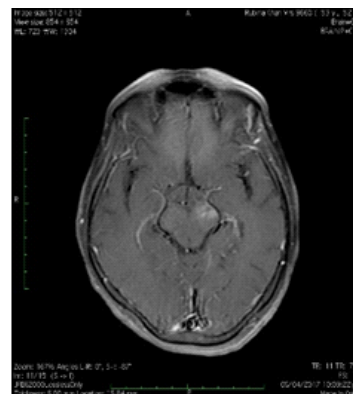
INTRODUCTION:

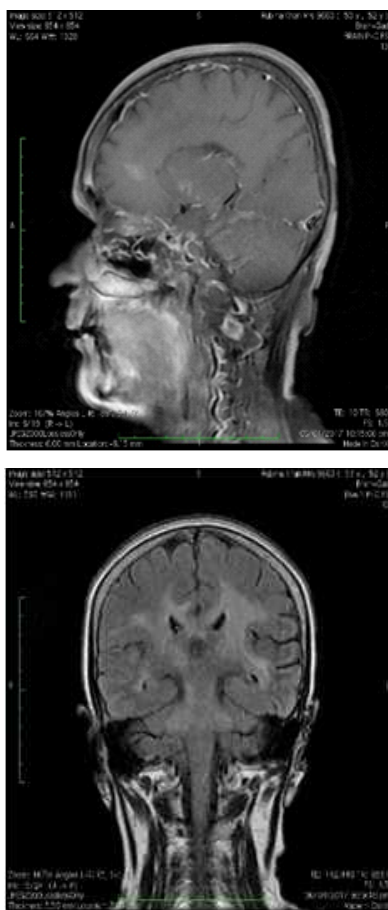
Acute disseminated encephalomyelitis (ADEM) is a monophasic autoimmune inflammatory demyelinating disease of brain and spinal cord. Mostly young and adolescent children are affected and the documented incidence is 0.8 per 100 000 population per year (1). The autoimmune response to myelin basic protein that is T-cell mediated, triggered by an infection or vaccination, underlies its pathogenesis (2). The differentiation of ADEM from a first attack of multiple sclerosis is crucial as it has both prognostic and therapeutic implications. MRI findings can be a helpful tool to label ADEM and high dose steroids is the treatment of choice for ADEM. Other options like immunosuppressants, plasmapheresis and iv immunoglobulins are given in refractory cases.

CASE REPORT:

This is a case report of a 52 years old diabetic female presented with complaints of inability to walk secondary to progressive lower limb weakness and dysphagia for 1 month and generalized body stiffness for 1 week. She was unable to walk for 1 month neither she could perform her daily chores. There was no history of paresthesia or pain in legs, no history of preceding viral illness or vaccination. On examination; she was vitally stable, but confused. Her pupils were bilateral equally reactive to light, extraocular movements were normal, uvula centrally placed but gag reflex was poor. All sensory modalities of both upper and lower limbs were intact, with no

fasciculations. Muscle bulk was normal in all four limbs, with a power of 3/5, hypertonia and brisk reflexes with no clonus in all four limbs, and the plantars were bilaterally down going. Cerebellar examination couldn't be done because of stiffness of limbs. Signs of meningeal irritation were absent. Her lab workup showed Hb=12.9g/dl, Wbc=12.5x10⁹/l, Platelets=310x10⁹/l, UCE, Calcium levels, LFTs, TSH and serum B12 levels were normal. Vasculitis work up came out to be normal. CSF DR showed; RBC=50cells/ul, WBC=5cells/ul, sugar=69mg/dl, (BSR=110mg/dl) and protein=50mg/dl. CSF was negative for oligoclonal bands. EEG and VEP were normal. MRI brain was done that showed abnormal high intensity areas seen in bilateral parietal, periventricular region, these show high signals on ADC image. On subsequent post contrast T1 weighted images, no significant enhancement is noted in such areas, findings are suggestive of ACUTE DISSEMINATED ENCEPHALOMYELITIS. (fig a-c)





Mri Showing High Intensity Areas In B/I Parietal, Periventricular Regions

During hospital stay initial suspicion was Parkinson's disease due to increased rigidity and mask like face. Initially patient was given supportive treatment, physiotherapy and anti-Parkinson drugs. As MRI BRAIN revealed the diagnosis of Acute Disseminated Encephalomyelitis (ADEM), the patient was started on intravenous methylprednisolone 1gram per day. Patient was improved on third day of intravenous steroids and her rigidity improved and she started responding to commands. She was discharged on oral steroids and was called for follow-up. Her symptoms did not recur over a follow up duration for 6 months. Keeping this in view, the diagnosis of ADEM is much strengthened as compared to other diseases mimicking ADEM on radiological basis like Adult onset leukodystrophies, CNS lymphoma and PML.

DISCUSSION:

Acute disseminated encephalomyelitis (ADEM) is an autoimmune disease that predominantly involve the white matter of brain and spinal cord [3]. The triggering factors are bacterial, viral or parasitic infections, after immunization or after taking some

medicines[4]. Therefore it can also be named as post-infectious encephalomyelitis and immune-mediated encephalomyelitis. Acute disseminated encephalomyelitis has an annual reported incidence of 0.4–0.8 per 100,000 [5,6]. Children and young adults are commonly affected in winter and spring season[5,6]. There is no gender predominance and the mean age at the onset of symptoms is 6-8 years [7, 8]. The survival rate with minor residual disability is 70 to 90%, while complete recovery is seen in 50 to 75% cases, with the mortality rate of 5% [6].

ADEM is a monophasic disease. It manifests clinically as sudden onset multifocal neurologic deficit including seizure disorder, motor, sensory and visual field defects, aphasia, movement disorders, impaired sensorium, and psychosis [9]. Our patient presented with both lower limbs weakness, dysphagia, rigidity and confusion. But the diagnostic dilemma in our case is that there is no clinical evidence of any previous illness or immunization, as the acute phase of ADEM presents about 4–21 days after the preceding infection [9]. The diagnosis is made on the basis of exclusion, as it is difficult to differentiate clinically between ADEM and viral encephalomyelitis. MRI is a very useful tool in diagnosing ADEM. The MRI, T2 images shows widespread lesions consistent with inflammation, demyelination and edema in the white matter, basal ganglia, thalamus, and brainstem [7]. CSF protein can be mildly elevated, sometimes with lymphocytic predominance. The presence of oligoclonal immunoglobulin bands, IgG or myelin basic protein (MBP) does not add to the diagnosis of ADEM [10]. There are non-specific features on EEG, and VEP shows delayed responses. Our patient's brain MRI findings are very suggestive of ADEM, while the CSF studies are normal, and oligoclonal bands not detected along with normal VEP results are adding to the diagnosis of ADEM rather than MS.

The treatment of choice is high doses of intravenous corticosteroids [11] like methylprednisolone (20–30 mg/kg/day) for 3–5 days followed by oral prednisolone tapered over 4–6 weeks. There are high chances of relapse if steroids are tapered before three weeks[12, 13]. The second line treatment is intravenous immunoglobulin, if high-dose corticosteroids use is contraindicated or the disease is refractory to steroids. However, plasmapheresis is recommended in severe cases [11, 14]. The immunosuppressive drugs like cyclophosphamide and mitoxantrone are the alternate treatment options in refractory cases [6]. Decompressive craniotomy is done in cases of intracranial hypertension [15]. Our patient responded well to high dose steroid therapy and clinically improved.

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Hina Mushtaq; concept, data collection, data analysis, manuscript writing, manuscript review
Maria Qureshi; concept, data collection, data analysis, manuscript writing, manuscript review
Irfan Ahsan; concept, data collection, data analysis, manuscript writing, manuscript review