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RESEARCH ARTICLE

"ACUTE DISSEMINATED ENCEPHALOMYELITIS IN YOUNG FEMALE - A DIAGNOSIS OF EXCLUSION"

Dr. Manas Mitra[1], Dr. Priya Govil[2] and Dr. Kishalay Datta[3]

1. Resident, Emergency Medicine, Max Super Speciality Hospital, Shalimar Bagh, Delhi.
2. Senior Consultant, Emergency Medicine, Max Super Speciality Hospital, Shalimar Bagh, Delhi.
3. HOD, Emergency Medicine, Max Super Speciality Hospital, Shalimar Bagh, Delhi.

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Abstract

ADEM, also known as acute disseminated encephalomyelitis, is a rare neurological condition which has myriads of presentation. It is generally a diagnosis of exclusion in the department of emergency medicine. The patient presentation is varied and often missed on the initial neuroimaging. In this case report, we present to you a case of ADEM in a young female patient, the diagnosis challenge and the treatment in ADEM.

Introduction:-

Acute disseminated encephalomyelitis (ADEM) is a rare neurological condition affecting the central nervous system of the patient. The pathogenesis of the disease is still unclear, but reports suggest it to be an autoimmune disease, usually predisposed by infection or immunization [1]. The diagnosis is usually very challenging and it is often missed in the emergency medicine department. Because of its rare nature, it is a diagnosis of exclusion in patients with neurological symptoms. The diagnosis is made by magnetic resonance imaging of the brain. There is no specific blood marker for the diagnosis of ADEM in patients, thus, the diagnosis is made only after ruling out all other neurological diseases by imaging and laboratory findings.

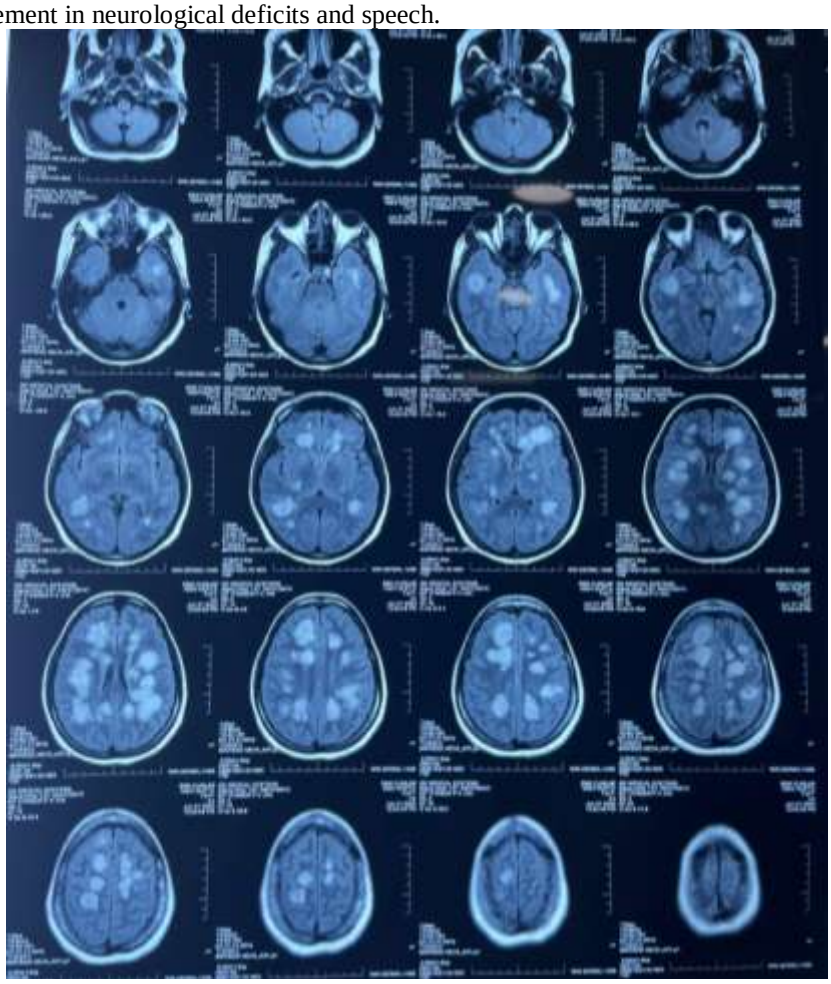
Case:
A 16 year old young female was brought to the emergency medicine department of our hospital after she complained of sudden weakness in all 4 limbs, associated with headache. The initial assessment in the emergency medicine department was normal. Neurological examination was normal. MRI brain was done which came out to be normal. Neurologist opinion was taken, psychiatric evaluation was requested by the child parents, hence taken. The child was initially discharged with a diagnosis of "attention disorder".

After 4 days, the child again presented to the ED, this time with paresis of all 4 limbs associated with altered sensorium with slurring of speech. The neurological examination showed increased tone in bilateral upper and lower limbs with brisk deep tendon reflexes, nystagmus towards the right, no meningeal signs and sensory examination was normal. MRI brain with spine scan done, which showed "hyperintensity in bilateral white matter". Lumbar puncture was done in ED, the reports showing cell count of 4 cells (100% lymphocytes), Protein - 36 mg/dl and Glucose - 51 mg/dl (capillary blood glucose was 88 mg/dl), LDH - 10 U/L. CSF analysis for microbial antigen test causing encephalitis (S. pneumoniae, H. influenzae b, Streptococcus Group B, E. coli, N. meningitidis), herpes IgM test and herpes PCR were inconclusive. Electroencephalogram revealed generalized cerebral dysfunction with no definitive irritable foci.

Corresponding Author:- Dr. Manas Mitra
Address:- Resident, Emergency Medicine, Max Super Speciality Hospital, Shalimar Bagh, Delhi.

After excluding all the possible neurological diagnosis, diagnosis of acute disseminated encephalomyelitis was made, the patient started on IV methylprednisolone (250mg) pulse therapy for 5 days, then transferred onto oral steroids. She responded well to speech therapy and the sensorium started improving from day 3 of initiation of the steroid therapy. Physiotherapy was given for limb weakness, which she responded well to. The patient was discharged after 7 days of hospitalization (3 days in ICU, 4 days in ward).

The neurological follow-up after 14 days revealed a healthy young female, walking into the emergency department for her workup, having improvement in neurological deficits and speech.



Discussion:-

The patient in our case study was diagnosed with acute disseminated encephalomyelitis (ADEM), after all other possible diagnosis was excluded by imaging and blood markers. The incidence of ADEM is around 0.1/1000 population, mostly occurring in the months of winter, largely affecting young children and female patients [2]. Majorly, cases occur post viral infection and post immunization [2, 3].

The etiopathogenesis of the disease is not known till date, but it has been postulated that ADEM generally occurs after an antigenic exposure. Autoantibodies are directed against the myelin sheath of the neuron, thus, damaging the myelin sheath [4]. The patient has various presentations involving the central nervous system like ataxia, movement disorder, visual defect, aphasia or seizure. As in our case, the child was having altered sensorium with paraparesis of all four limbs with speech difficulty.

There is no specific blood marker for the diagnosis of ADEM in patients. CSF studies may sometimes show raised protein and lymphocyte predominance. Specific markers like myelin basic protein, oligoclonal immunoglobulin bands, IgG, may be present, but not diagnostic [5]. The only current method for diagnosis of ADEM is MRI scan of the brain, showing hyperintense lesions in the white matter in T2 weighted images of MRI.

The treatment for ADEM includes supportive and immunomodulatory therapy [6]. Many literature teaching suggests use of high-dose intravenous methylprednisolone, intravenous immunoglobulin (IVIg), and plasmapheresis as treatment modality. Intravenous methylprednisolone is the first-line drug (10–30 mg/kg/day, up to a maximum of 1 g/day) for 3–5 days followed by oral corticosteroid treatment continued with gradual tapering over 6 weeks to reduce the risk of relapses. Intravenous immunoglobulin (IVIg) (0.4 gm/kg/day for 5 days) can also be considered. The use of plasmapheresis or IVIg could be done when corticosteroids fail.

The outcome in ADEM is fruitful with 57-90% of the recovery process [7]. The disease is monophasic, meaning, it occurs only once in a lifetime, but if it presents in successive years of life due to immune response, it is termed as "multiphasic disseminated encephalomyelitis" (MDEM).

Conclusion:-

Acute disseminated encephalomyelitis is a diagnosis of exclusion, where patients can present with various neurological symptoms. The diagnosis of choice is MRI brain T2 weighted images. The treatment outcome is good and prognosis is also satisfactory, provided it is diagnosed and treated correctly. The treatment is immunotherapy and supportive management. Our patient was timely diagnosed and intervened, hence, she recovered well with good end results.

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