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CASE STUDY

A CASE OF MULTIPLE SCLEROSIS (MS) WITH FULMINANT ACUTE DISSEMINATED ENCEPHALOMYELITIS (ADEM) LIKE PRESENTATION

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ABSTRACT

Background: Multiple Sclerosis is the commonest immune mediated inflammatory disorder of the central nervous system. It is usually relapsing remitting and presents with focal neurological deficit. On the other hand ADEM (Acute Disseminated Encephalomyelitis) also known as postinfectous encephalomyelitis is a rare CNS demyelinating disorder which is typically monophisic and presents with encephalopathy.
Objective: We would like to report a rare case of MS that present with ADEM like clinical and radiological picture.
Material and Methods: A 16 year female presented to the emergency room with progressive deterioration in consciousness over five days.
Results and conclusion: MRI Brain and Cervical spine was suggestive of ADEM while CSF examination and brain biopsy was suggestive of MS.

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INTRODUCTION

Both multiple sclerosis and acute disseminated encephalomyelitis are considered as immune mediated inflammatory diseases of the CNS but each of them has unique features. MS is a relapsing remitting disease in 85 % of patients though other forms of MS include secondary progressive, primary progressive and relapsing progressive MS. On the contrary, ADEM which often occurs postinfectously with predilection to early childhood is usually monophisic. Nevertheless, multiphasic ADEM has been reported (Haren *et al*., 2016). The diagnosis of MS is based on the revised McDonald criteria 2010 (Polman *et al*., 2011) which demonstrate distribution in space and time as follows:

- Two or more attacks, objective clinical evidence of two or more lesions or one objective clinical evidence of one lesion with historical evidence of prior attack.
- Two or more attacks with objective clinical evidence of one lesion.

The distribution in space is showed by one or more T2 lesion in at least 2 of 4 areas of the CNS (Periventricular, Juxtacortical, Infratentorial and spinal cord) C- One attack, objective clinical evidence of two or more lesions and the distribution in time is showed by:

- A new T2 and or gadolinium enhancing lesions on follow up MRI with reference to a baseline scan, irrespective of the timing of the base line MRI.
- 2-Semultaneous presence of asymptomatic gadolinum enhancing and nonenhancing lesions at any time.
- D- 1 attack, objective clinical evidence of one lesion (clinically isolated syndrome CIS) with radiological evidence of distribution in space and time.
- E- For primary progressive MS the criteria are :
- One year of disease progression plus 2 of 3 criteria
- Evidence for DIS in the Brain
- B-Evidence for DIS in the spinal cord based on 2 or more T2 lesions.
- Positive CSF (evidence of oligoclonal clonal bands and or elevated IgG Index)
- For the Diagnosis of ADEM the new criteria are :
- A first polyfocal clinical CNS event with presumed inflammatory demyelinating cause
- 2-Encephalopathy unexplained by fever,systemic illness or posticta l symptoms)
- Brain MRI abnormalities consistent with demyelination during the acute (3 months) phase.
- No new clinical or MRI findings 3 months or more after the clinical onset Rare varint of multiple sclerosis

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known as Murberg type which usually has a monophasic sever fulminant course has been reported before (Jasperson, 1998; Johnson *et al*., 1990).

CASE REPORT

A 16 year female presented to the emergency room with progressive drowsiness over five days no history of fever or recent febrile illness.

On examination: Patient was lethargic confused disoriented inattentive obey simple commands she has intermittent episodes of up rolling of eyes with upper limbs shaking but no tongue biting or sphincter disturbance during the episodes.

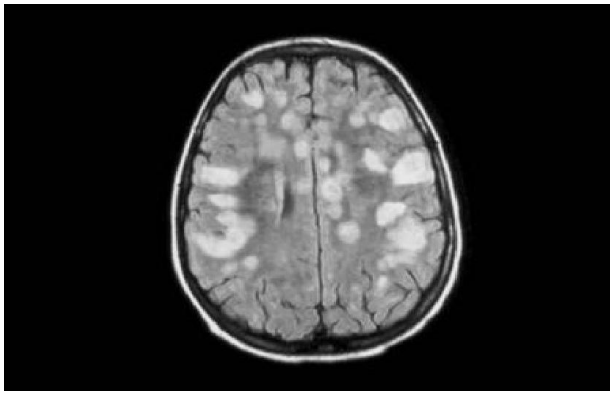

Image (A)

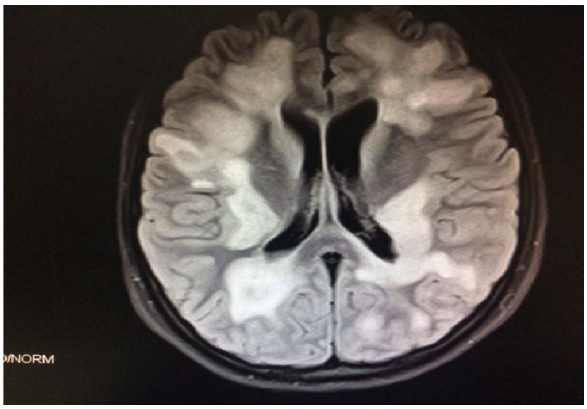

Image (B)

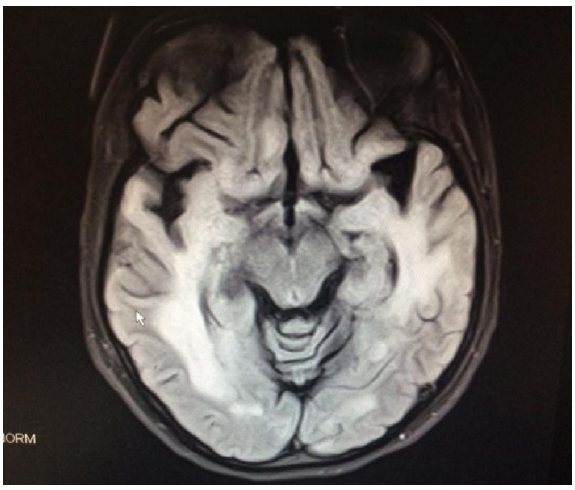

Image C

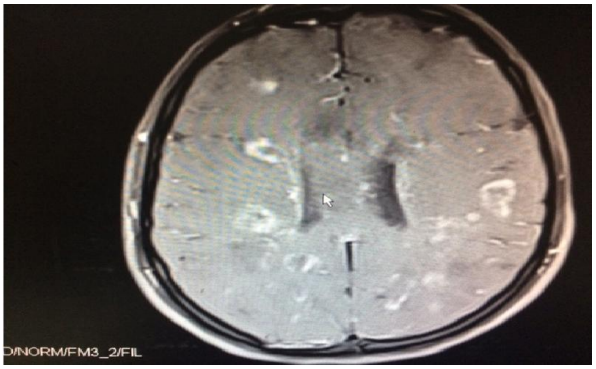

Image D

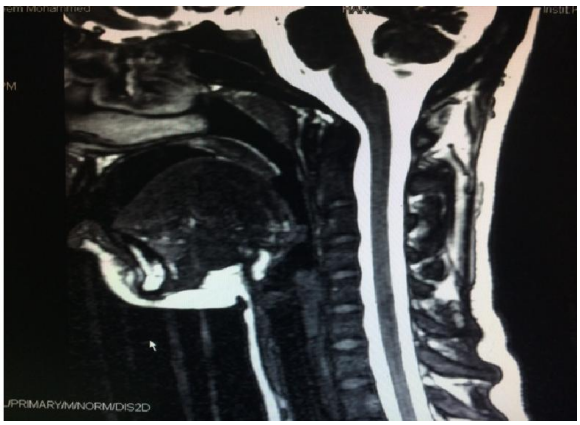

Image E

She has no meningeal signs or focal weakness but has up going planter reflex. Investigations revealed unremarkable cell count as well as normal renal and hepatic profile, negative HIV and toxoplasma serology. MRI Brain showed neumerous well defined variable size and shape from rounded to oval abnormal lesions involve both cerebral hemispheres and cerebellum .The lesions predominantly affect the jaxtacortical, per ventricular white matter as well as thalami and callososeptal regions with some restriction diffusion Image (A) (B) (C)The lesions showed variable pattern of enhancement (D). MRI Cervical spine showed focal anterior abnormal signal alterations seen at the medullary cervical junction at the level of C1 to the left side (E).

Carotid angiogram was unremarkable

CSF Examination: WBC =1 RBC =3 Protein = 0.44 g/L Glucose =5.22mmol/L CSF Culture showed no growth IgG Index (CSF/Serum) =6.2600. Albumin Index (CSF/Serum) = 6.7631. Presence of two Oligoclonal Bands EEG showed diffuse slowing. Brain Biopsy (Rt frontal lobe) showed increased number of microglial cells in the gray matter no neoplastic cells and there is loss of myelin Patient received methylprednislon 1 g IV for five days and started on levetiracetam 500 mg PO twice daily her level of consciousness detriorated patient intubated received IV Immunoglobuline 2g/kg. Followed by plasma exchange as patient did not improve initially. Follow up MRI Brain after one month showed significant reduction in the previously noted enhancement and edema. No new lesions detected Patient started on Interferon beta-1a injection remained in vegetative state until she died several weeks later.

DISCUSSION

This case was initially suspected to have ADEM given the fulminant presentation with encephalopathy, seizer and multiple hyperintense bilateral Asymmetric enhancing patchy demylinating lesions of different sizes which Envolve cortical gray-white matter junction, centeral white matter, basal ganglia, cerebellum in addition to spinal cord. This presentation is characteristic of ADEM But the presence of two oligoclonal bands in the CSF and brain biopsy weave suggestive of MS. This fulminant MS presentation though extremely rare, should be considered in patient with suspected ADEM. Acute cases of rare monophasic MS (Marburg type) and Balò's concentric sclerosis have been reported. They representrare variants of multiple sclerosis, clinically characterized by a rapid deterioration and grave prognosis and by unusual demylinating lesions, often large tumor-like demyelinating plaques in Marburg type and concentric layers of partial demyelination alternating with demyelinating bands in Balò's concentric sclerosis (Capello *et al*., 2004).

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