Multiphasic Acute Disseminated Encephalomyelitis

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Abstract:-

We report a rare case of multiphasic ADEM. This patient had three episodes of different neurological deficit starting from the age of 12 yrs. His CSF for oligoclonal bands was negative. His MRI was suggestive of encephalitis.

Introduction:- Acute disseminated encepahalomyelitis shows acute encephalitic, myelitic or encehalomyelitic features. It is a nonvasculitic inflammatory demyelinating condition. It usually shows monophasic course. However it can be either recurrent or multiphasic also. Recurrent ADEM means that further episodes are stereotyped although the complete original syndrome is not necessarily present. Multiphasic ADEM indicates two or more separate episodes that differ in cliniical presentation(1). We report a case with three episodes with different clinical presentation each time.

Case report

19 yr old male patient, student by occupation presented to us with complaints of fever since 8 days, slurring of speech , tingling sensation on left half of body since 7 days, weakness in left half of body since 3 days, seizures 5-6 episodes left side of face since 3 days.

Past history revealed that, in 2004 (when he was 12 years old) he had h/o weakness in right half of body and face and focal seizures on right side of face for which he was admitted in private hospital and diagnosed as c/o ?encephalomyelitis with CVE with brainstem infarct however CT/MRI was not done that time. He was advised to take anticonvulsant drug therapy but he stopped taking medication after one year. In 2008(when he was 16 years old) patient again had h/o focal seizures and weakness in both lower limbs with retention of urine. He was admitted in GMCH Nagpur. MRI done(PHOTO NO 1) that time showed multiple patchy areas of altered signal intensity involving grey and white matter in both cerebral and cerebellar hemispheres including striato capsular area, thalamus and spinal cord" f/s/o Acute Disseminated Encephalomyelitis. He was started on T Dilantin 100mg tds. In June 2011 again had focal seizures and was admitted in private hospital and diagnosed as seizures disorder and started on T Dilantin and T Levotiracetam.

At the time of this admission,patient was apparently alright 8 days back. To start with he developed fever mild grade on and off in nature not associated with chills, slurring of speech since 7 days, weakness in left half of body which was gradual in onset since 3 days, focal seizures in left side of face in the form of twitching. There was no h/o burning micturition, cough, headache, vomiting, syncope, blurring of vision, bladder and bowel involvement or ear discharge. On examination he was febrile,was conscious,disoriented and had seizures on left side of face.His b.p.was 110/70mm of mercury.Respiratory and cardiovascular system examinationsdid not reveal any abnormality. . Neurological examination revealed supranuclear VII N palsy on left side with grade 3 power in left half of body..There was hypertonia in both upper and lower limbs on left side.Reflexes were brisk & left plantar was extensor.

His investigations done during this admission revealed the following.

CSF study showed proteinof 25 mg/dl, sugar 71 mg/dl, chloride 121 mg/dl, ADA was 1.9 , CSF was acellular . CSF with parallel serum sample was negative for oligoclonal band. ANA done by immunofluroscence was negative. Serum Lactate was 2.11mmol/lit . 2D ECHO did not reveal any cardiac abnormality. FUNDUS was normal. MRI BRAIN showed focal leptomeningeal involvement in right high parietal region and edema s/o focal meningitis(photo no.2).

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Diagnosis of Acute disseminated encephalomyelitis (muliphasic)was entertained though this time MRI did not reveal encephelitic changes as it was done after he received steroids.

Patient was put on T. DILANTIN 100mg TDS, T FOLIC ACID 5mg OD, T FRISIUM 200mg BD, T diamox 250mg BD, Inj Methyprednisolone 1gm OD x 5days. After seven days he recoverd completely and was discharged.

Discussion-

Classicaly ADEM follows a monophasic course that may occur after vaccination(small pox &certain rabies vaccines) or after viral infections like measales, rubella mumps, influenza.(1)

Poser has proposed that DEM with MS classified into two types, in first one An initial episode of ADEM from which there is complete or partial recovery is followed by reccurences that are characteristically stereotyped. this is called recurrent DEM (RDEM). In the other type two or more separate acute episodes that differ in clinical presentation this is called multiphasic DEM.(,2,3)

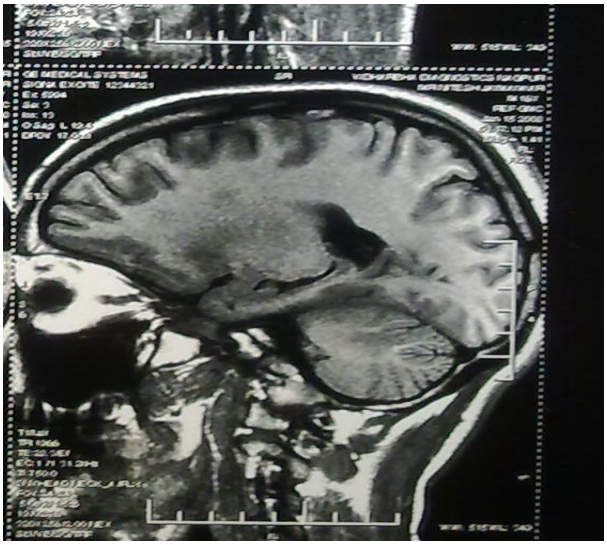
MDEM in extremely difficult to diffrentiate from MS on clinical ground but it is important to do so since the prognosis of MDEM is much better. In patients with DEM the CSF most commonly shows moderate pleocytosis and increase in total proteins. IgG is often raised but oligoclonal bands are rare. Oligoclonal bands never disapperes in MS but may do so in DEM and this is important in differentiating MS from both RDEM and MDEM.(4) MRI in DEM is characterstic , reveals that unlike images in MS, the lesion are extensive often following the outline of cortical ribbon or may consist of very large globular areas of increased signal intensity that donot produce mass effect. The cerebellum and cerebral cortex are often involved in DEM and occasionally Thalamus and Basal Ganglion, in contrast to fact that involvement of thalamus is exceedingly rare in MS. In DEM the corpus callosum is usually not affected and the periventricular distribution of the lesion is not as constant as in MS(3).

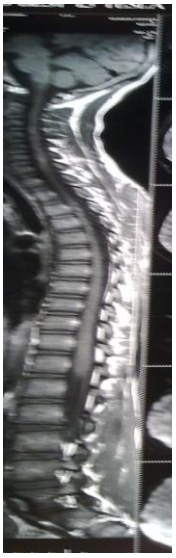
Initial treatment of ADEM with high dose of glucocorticoides depending upon the response treatment may need to be continued for four to eight weeks. Patients who failed to respond may benefit from plasma exchange or intravenous immunoglobulinsPrognosis of these patients is related to severity of underlying illness.Mortality of 5 to 20% is described with measales encephalomyelitisSurvival with seizures and behavioural disorder is common(1)

Our patient had three episodes of neurological deficit with different clinical presentationeach time and relatively asymptomatic period between these episodes. Differential diagnosis in our patient includes stroke or demyelination MRI done did not reveal any acute or chronic infract so the diagnosis of stroke was unlikely. Our patient had episodes of fever, headache , meningism and seizures. these features favoures the diagnosis of ADEM(1). CSF for oligoclonal bands was negative also MRI done in 2008 shows typical finding of ADEM. [fig no 1& 2]. Different clinical presentations each time favours diagnosis of Multiphasic DEM in contrast to recurrent DEM. So patient was kept as case of MDEM. Patient was treated with 5 day course of methyl prednisolone following which his power improved to grade five.Patient is under follow up &asymptomatic at present.

Schwartz et al(4) perfomed a follow up cohort study of 40 adult patients(28 women) diagnosed as ADEM. A final diagnosis of ADEM or clinically definite MS was established. On follow up examination after 8 to 137 months.clinical symptoms, MRI and CSF findings and response to standardized treatment during acute phase of disease were analyzed.Upon follow up,14 patients had developed clinically definite MS.Of the 26 patients with final diagnosis of ADEM,2 patients died,9 had minor deficits,3 had moderate deficits and 12 had no remaining symptoms.They concluded that many patients initially diagnosed as ADEM develop clinically definite MS.The authors found no useful diagnostic criteria for the differentiation of first episode of MS from monophasic ADEM.

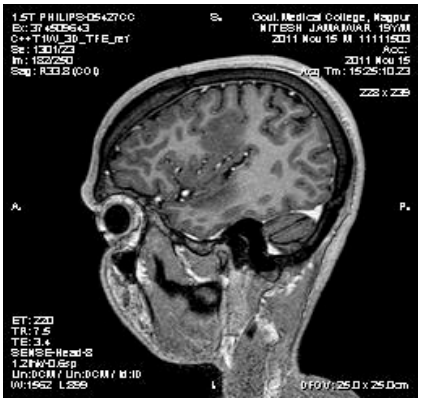
Photo No 1

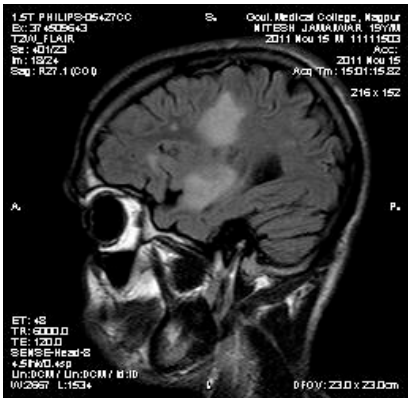




MRI done in 2008 showed multiple patchy areas of altered signal Intensity in grey and white matter in bilateral cerebral and cerebellar Hemispheres including striatocapsular region and spinal cord s/o ADEM

Photo No 2





T2 Image Post Contrast

MRI Brain (nov.2011) Plain&Contrast S/0Focal Leptomenengial Enhancement&Altered Signal Intensityin Parietal&temporal Region S/0focal Menengitis & edema

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