Case Report

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Recurrent ADEM Mimicking Young Stroke

Sureka RK[1]*, Gupta M[2], Agarwal A[3] and Chaturvedi S[3]

[1]*Prof & Head, Department of Neurology, Mahatma Gandhi Medical College, Jaipur, India*

[2]*Associate professor, Department of Neurology, Mahatma Gandhi Medical college and hospital, Jaipur, India*

[3]*Senior resident, Department of Neurology, Mahatma Gandhi Medical College, Jaipur, India*

***Corresponding author:** Sureka RK, Prof & Head, Department of Neurology, Mahatma Gandhi Medical College, Jaipur, India

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) usually is a monophasic demyelinating disorder, often post infectious predominantly observed in children. Relapses are rare, and patients with recurrent episodes are eventually diagnosed as having multiple sclerosis. There were no definite diagnostic criteria or established treatment for ADEM. International Pediatric MS Study Group laid down the first consensus definition. In this case report, we present an adult case of recurrent disseminated encephalomyelitis with recurrent episodes of fever, hemiparesis, seizure, and unconsciousness, who responded dramatically in both episodes with intravenous steroid treatment with no CNS deficit on follow up.

Abbreviations: ADEM: Acute Disseminated Encephalomyelitis; CSF: Cerebrospinal Fluid; MRI: Magnetic Resonance Imaging; IVIG: Intravenous Immunoglobulin; RCVS: Reversible Cerebral Vasoconstriction Syndrome; MS: Multiple Sclerosis

Introduction

ADEM, also known as post-infectious encephalomyelitis, is a demyelinating CNS disorder that usually follows the occurrence of infection, or more infrequently, after the administration of a vaccination [1-3]. ADEM should be suspected when one or more of the following features are present such as a multifocal and polysymptomatic initial presentation, the presence of signs and symptoms suggestive of meningoencephalitis, encephalopathy, bilateral optic neuritis, cerebrospinal fluid (CSF) fluid pleocytosis along with the typical magnetic resonance imaging (MRI) picture [4,5]. It affects children more than adults but can affect anyone. Although it is monophasic by definition, relapsing forms of ADEM have also been recognized.

Case Report

A 30-year male, right-handed, presented with a history of fever for 7 days, holocranial episodic acute severe headache for 3 days, which was associated with nausea, photophobia, and phonophobia. Headache was short-lived but was recurrent, it persisted for around 2-3 hours and get relieved. No triggering factor for this exacerbation. No associated neck pain. The patient also had 1 episode of involuntary body movements, frothing from the mouth, urinary incontinence with loss of consciousness for 1 day. H/O altered sensorium with the irrelevant talk was present. A few hours later, the Patient complaint of Weakness of the left upper and lower limb with facial deviation to the right side. Weakness was to the extent he was unable to lift hand and legs above the bed. On examination, patient was conscious but altered sensorium, left UMN facial nerve palsy, left hemiparesis (Grade- 3/5), brisk deep tendon reflexes, bilateral Extensor plantar reflex, no meningeal signs, no involvement of bowel/bladder, and the sensory deficit was there. Systemic examinations were normal. Blood picture and biochemical investigations were within normal limits. Magnetic resonance imaging (MRI) brain showed focal hyperintense lesions (FLAIR and T2) in the subcortical white matter of right basal ganglia, right corona radiate, left parietal- temporal- occipital region [(Figure 1)-MRI July 2018], and bilateral cerebellar hemisphere and no restriction in DWI &meningeal enhancement in a gadolinium-based MRI contrast study.

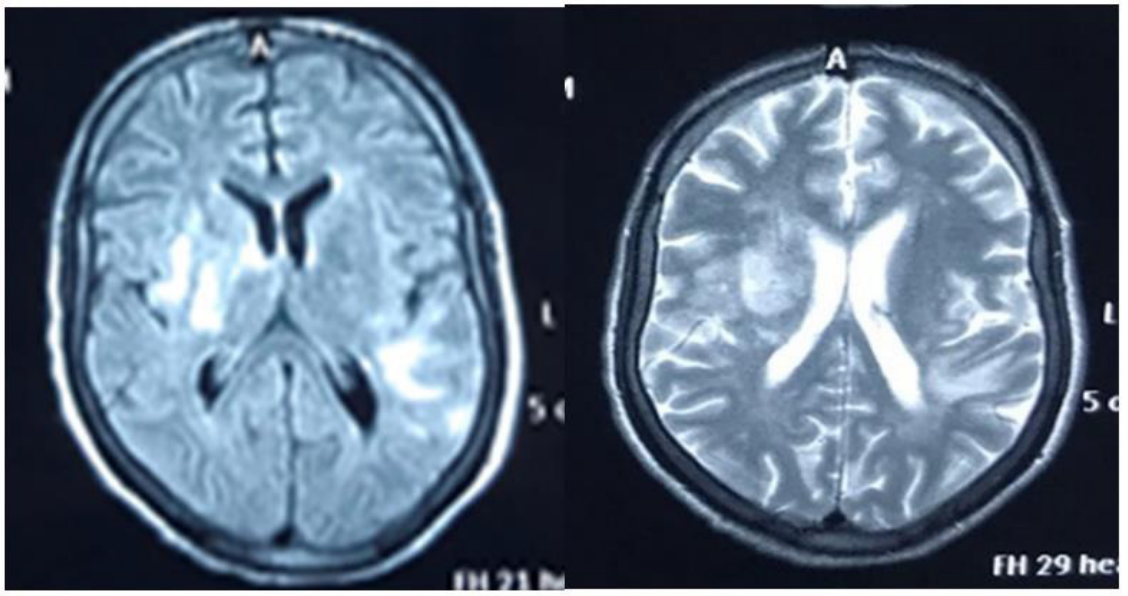

Figure 1: Magnetic resonance imaging brain showing focal hyperintense lesion (FLAIR& T2) in cerebral cortex and subcortical white matter indicating demyelination of brain in the first episode (July 2018).

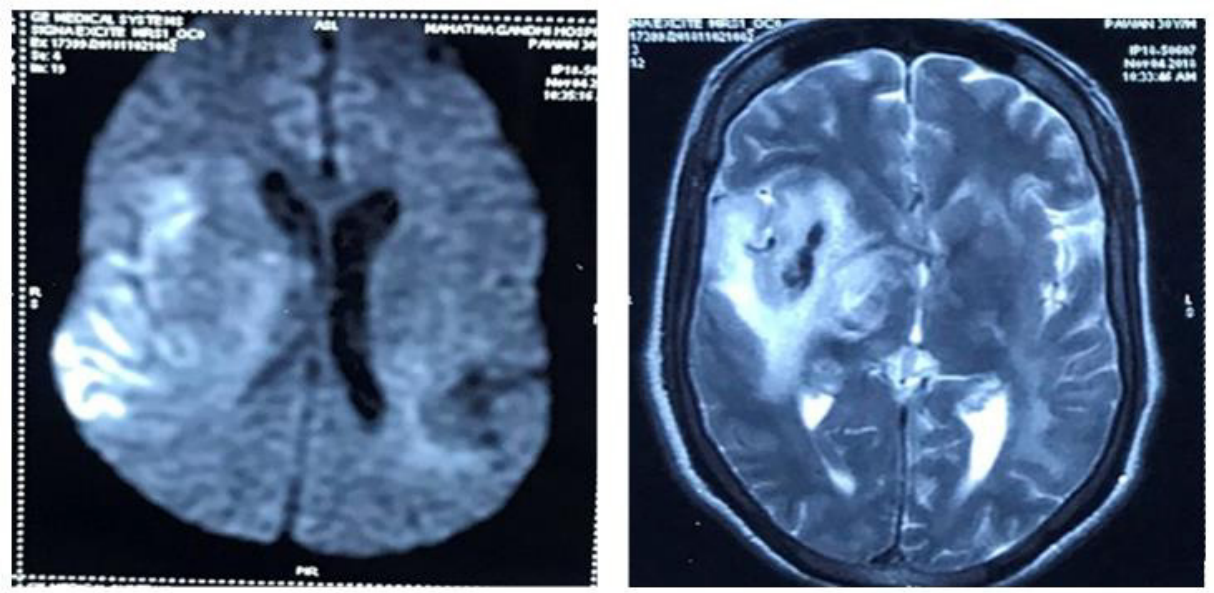

Figure 2: Magnetic resonance imaging brain showing focal hyperintense lesion (DWI& T2) in cerebral cortex and subcortical white matter indicating demyelination of brain in the second episode (Nov 2018).

Cerebrospinal fluid (CSF) study was done which shows Sugar 45mg/dl, protein 248mg/dl, cell count 25/cmm (99% lymphocytes), negative for Gram-stain or Ziehl–Neelsenstain, and no growth in culture, viral encephalitis panel, and TB PCR was negative. Keeping diagnosis of ADEM, he put on intravenous immunoglobulin (IVIG) for 5 days then oral prednisolone and antiepileptic, and was discharged in stable condition. After 3 months, in November 2018, he was again admitted with a similar severe headache, left hemiparesis grade 2/5, left UMN type facial palsy with left focal seizure and secondary generalization, and global aphasia. MRI brain showed focal hyperintense, demyelinating lesions [(Figure 2)-Nov 2018], in the same brain territory as in the previous episode [(Figure 1)-MRI July 2018]. Immunological markers: Antinuclear antibody, antineutrophil cytoplasmic antibody, anticardiolipin antibody, lupus anticoagulant were negative. Serum Lactic acid and PBF for Sickle cell were normal. MR Venography and CT angiography brain plus neck vessel were normal. His treatment was IVIG for 5 days, followed by oral Prednisolone in the tapering dose and anti-epileptics and the patient showed improvement. After follow-up on 1 month, clinically/MRI brain was normal (Figure 3).

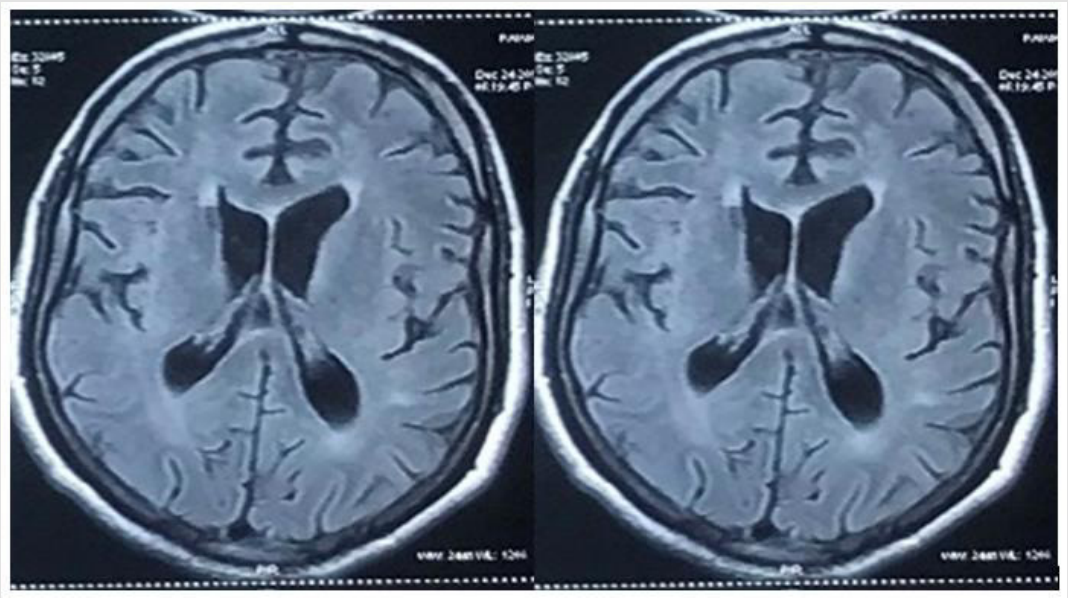

Figure 3: MRI brain after 1 month showed disappearance of lesions.

Discussion

The index case presented with recurrent episodes of headache, seizures, encephalopathy, a focal neurological deficit in form of hemiparesis and aphasia. The differential diagnosis of recurrent CNS lesions is considered [6]:

1) Recurrent Meningo Encephalitis

2) Reversible cerebral vasoconstriction syndrome(RCVS)

3) MELAS

4) Primary CNS vasculitis/Angiitis

5) Recurrent CNS inflammatory demyelinating disorder like NMO, MS, ADEM

Of the above, Systemic diseases like MELAS were excluded as, S. Lactic acid was normal, no family history, and no multiorgan manifestations like myopathy, hearing impairment, short stature, dementia, and Recurrent meningoencephalitis was excluded as there was no stiff neck or signs of meningismus and CSF HSV PCR was negative. Multiple sclerosis (MS) and ADEM, the two major CNS inflammatory demyelinating diseases, are difficult to differentiate in the initial episode. MS is a continuous demyelinating disease with a characteristically relapsing-remitting course [6]. Although recurrence is characteristic of MS, a second ADEM is described. Furthermore, there are no definite guidelines to differentiate MS and ADEM [6].

Recurrent ADEM (RADEM) is defined as the occurrence of a new episode with a recurrence of the first symptoms and signs, 3 or more months after the first ADEM event and after at least 1 month completing therapy, without a new central nervous system (CNS) lesion (clinical or neuroimaging). The incidence of the second episode has occurred in 10–18% of cases [7]. The category of recurrent ADEM was eliminated in the 2013 criteria, and replaced by the term multiphasic disseminated encephalomyelitis (MDEM), describing 2 episodes consistent with ADEM, separated by at least 3 months [8]. In our patient, 2 episodes of headache, seizures, encephalopathy, a focal neurological deficit in form of hemiparesis and aphasia separated by a period of 3 months, elevated CSF protein, an absent oligoclonal band in CSF, same territory MRI lesions, and complete clinic neuroradiological recovery clinch to the diagnosis RADEM. In previous studies, there are only a few adult RADEM have been reported [8-10]. However, more than two ADEM should be suspicious for MS. Neuropsychiatric features may be the main presentation of a relapse. Since recurrent ADEM is a corticosteroid-responsive condition, awareness and early diagnosis are mandatory [10].

Conclusion

RADEM can be diagnosed clinically, initiated treatment at the earliest because it is most important for the outcome.

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