ISSN : 1001-1781
Journal of Clinical Otorhinolaryngology, Head, and Neck Surgery

ROLE OF MRI IN ACUTE DISSEMINATEDENCEPHALOMYELITIS

Rushabh Raka[1]*, Ashutosh Patel[2], C. Vatsal Akhani[3]

[1]3rd year resident, [2]Assistant Professor, [3]3rd year resident

Department of Radiodiagnosis, SBKS Medical Institute and Research Centre, Sumandeep VidyapeethUniversity, Vadodara, Gujarat, India

*Corresponding author email: rushabhraka4@gmail.com

Abstract

Background: Acute disseminated encephalomyelitis (ADEM) is indeed an acute demyelinating disorder of the central nervous system (CNS). It is characterized by multifocal white matter involvement, leading to diffuse neurological signs. The disease is characterized by the presence of multifocal lesions in both the brain and spinal cord. It is important to note that ADEM does not involve the invasion of the central nervous system by external pathogens or infectious agents.

Instead, it is believed to be immune-mediated, involving inflammation and demyelination triggered by an immune response.

Aim: To evaluate role of MRI in acute disseminated encephalomyelitis.

Materials and methods: A prospective study of 10 cases which was conducted in the pediatric department in Dhiraj hospital. MRI brain was done on 1.5 Tesla MRI machine.

Results: Neuropathologically, ADEM is characterized by perivenular inflammation, which refers to inflammation around the small blood vessels in the affected areas of the CNS, and demyelination, which is the loss of the protective myelin sheath surrounding nerve fibers. These pathological

features are distinct from those seen in acute viral encephalitis, where the primary pathology involves the invasion and replication of viruses within the brain tissue.

Conclusion:

1. Gender Predominance: ADEM has no female predilection..

2. Association with Previous Infection: ADEM is more frequently associated with previous

infections rather than previous vaccinations. This suggests that certain infections may trigger an immune response leading to the development of ADEM.

3. Neurological Deficits: ADEM typically presents with neurological deficits, which can vary

widely depending on the location and extent of demyelination in the central nervous system.

4. Cerebrospinal Fluid (CSF) Findings: Analysis of CSF often reveals raised levels of protein.

5. MRI Findings: Magnetic resonance imaging (MRI) studies commonly show hyperintense lesions on T2-weighted (T2W) and fluid attenuated inversion recovery (FLAIR) sequences. These lesions are indicative of areas of demyelination within the brain and spinal cord.

6. Periventricular Association: ADEM is frequently associated with lesions in the periventricular area of the brain.

Key words: Acute disseminated encephalomyelitis, ADEM, MRI, CNS disorder.

Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune-mediated demyelinating disorder of the central nervous system (CNS). Clinically, it is characterized by the sudden onset of polyfocal

neurological symptoms. Importantly, the diagnosis of ADEM is often supported by neuroimaging

evidence, which typically reveals multifocal areas of demyelination within the CNS. This combination of clinical presentation and neuroimaging findings helps differentiate ADEM from other neurological conditions and guides appropriate management strategies.

Acute disseminated encephalomyelitis (ADEM) is a monophasic illness, meaning that it typically occurs as a single episode. It often manifests with the highest incidence in early childhood. However, ADEM can also occur in adults, albeit less frequently.

One of the concerning aspects of ADEM is its potential for severe outcomes. It carries a notable mortality rate, with studies reporting rates of up to 30%. Additionally, even among those who

survive, neurological sequelae or complications are common. These sequelae may include various degrees of neurological impairment, cognitive deficits, motor dysfunction, and other neurological impairments. Therefore, prompt recognition, diagnosis, and appropriate management of ADEM are crucial to mitigate these adverse outcomes and improve patient prognosis.

Signs and symptoms [4, 5]

Symptoms are more systemic rather than focal and include-

- Fever

- Headache
- Decreased level of consciousness varying from lethargy to coma
- Seizures
- Multifocal symptoms such as hemiparesis, cranial nerve palsies, movement disorders
- Behavioral changes like depression, delusion and psychosis may dominate the symptoms.

Triad of ADEM

- Prodromal illness or preceding vaccination,
- MRI signs of demyelination
- Acute presentation of neurologic symptoms

Materials and methods

A prospective study of 10 cases was conducted in the pediatric department in Dhiraj hospital. These cases were admitted with a chief complaint of fever, seizures and behavioral problems after the initial episode of infection or after vaccination. MRI brain was done on 1.5 Tesla MRI machine.

Demographic and epidemologic data

- 10 clinical cases were taken. Out of them 6were female and 4 were male.
- 8 Cases had the history of previous infections and 2 had history of previous vaccination.

Results

Clinical features were as per Table – 1. Physicalexamination was as per Table – 2.

Laboratory investigations

CSF analysis was done

- Raised protein level was found in 9 patients
- One patient had normal CSF protein.
- CSF lymphocytes were raised in all cases.

Table – 1: Clinical features.

Sign and Symptoms (10 Patients)	
Fever	9
Nausea and vomiting	5
Headache	7

Neck stiffness	1

Table – 2: Physical examination.

Neurological deficits	4
Ataxia	2
Hemiparesis	2
Paraparesis	1
Convulsions	3
Visual disturbances	2
Oculomotor nerve palsy	1
Facial nerve palsy	1
Impairment of consciousness	3

Cases

A two years-old previously healthy girl who presented with fever and altered sensorium in OPD, having history of infection.

She had hemiparesis on right side.

She developed postinfectious focal encephalitis.

MRI was done and showed fluid attenuated inversion recovery (FLAIR) sequences show extensive area of increased signal in left hemisphere and right basal ganglia (Photo -1).

T2 weighted MRI showing extensive area of increased signal in left hemisphere and right basal ganglia (Photo – 2).

Diffusion-weighted MRI (DWI) show a restriction of diffusion (Photo – 3).

A 14 year old male admitted for acute illness with lowered consciousness, right-sidedhemiparesis and fever.

Few days later, we note a progressively aggravate clinical symptoms evolving to coma. MRI was done - Axial Flair brain images:Bilateral and asymmetric involvement of the periventricular and subcortical white matter (Photo – 4).

13year old patient presented with visual disturbances, hemiplegia, single episode of seizures. FLAIR sequence shows hyperintense signal involving periventricular, subcortical white matter and grey matter on left side (Photo – 5).

ADEM can be distinguished from acute viral encephalitis because the disease is not the result of primary tissue invasion by an infectious organism. It is thought to be immune-mediated and is characterized neuropathologically by perivenular inflammation and demyelination.

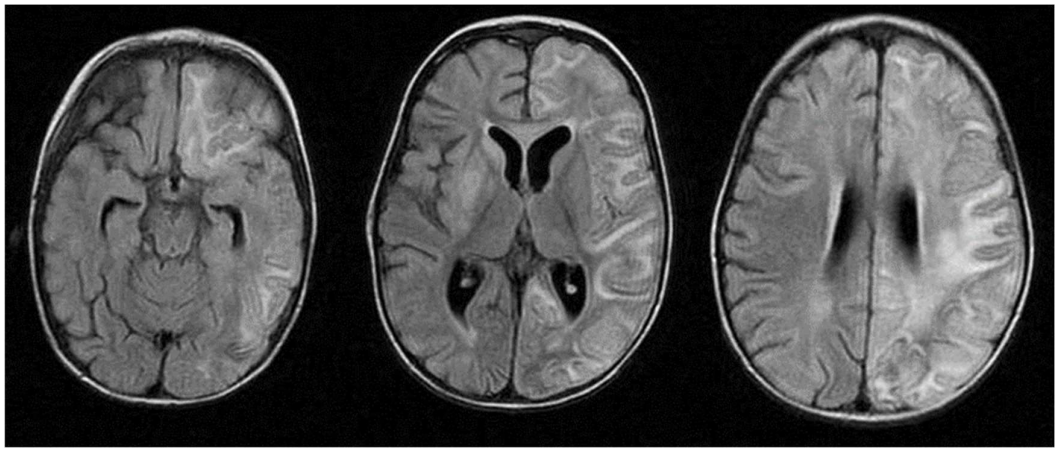
Table – 3: Hyper intense lesions on T2W and flair sequences.	
Bilateral and asymetrical in white matter	8
Diffuse involvement in white matter	2
Unilateral	1
Profound involvement of grey matter	1

Table – 4: Areas of brain affected.	
Periventricular area	7
Subcortical white matter	3
Basal ganglia	5
Thalamus	4
Cortical grey matter	1
Brainstem	2
Spinal cord	1
Cerebellum	0

Discussion

Demyelinating lesions of ADEM are better visualised by MRI. These demyelinating lesions of ADEM usually exhibit no mass effect and can be seen scattered throughout the white matter.

Photo – 1: Fluid attenuated inversion recovery (FLAIR) sequences show extensive area of increasedsignal in left hemisphere and right basal ganglia.



Characteristic lesions seen on MRI appear as patchy areas of increased signal intensity on conventional T2-weighted images and on fluid attenuated inversion recovery sequence (FLAIR).

Few MRI lesions may enhance after gadolinium administration. Extensive perifocal oedema may be seen.

Though white matter involvement predominates grey matter can also be affected, particularly basal ganglion, thalami, and brainstem

Thalamic involvement may be seen in 40% patients of ADEM, making this finding a potentially useful discriminator.

Photo – 2: T2 weighted MRI showing extensive area of increased signal in left hemisphere and right basal ganglia.

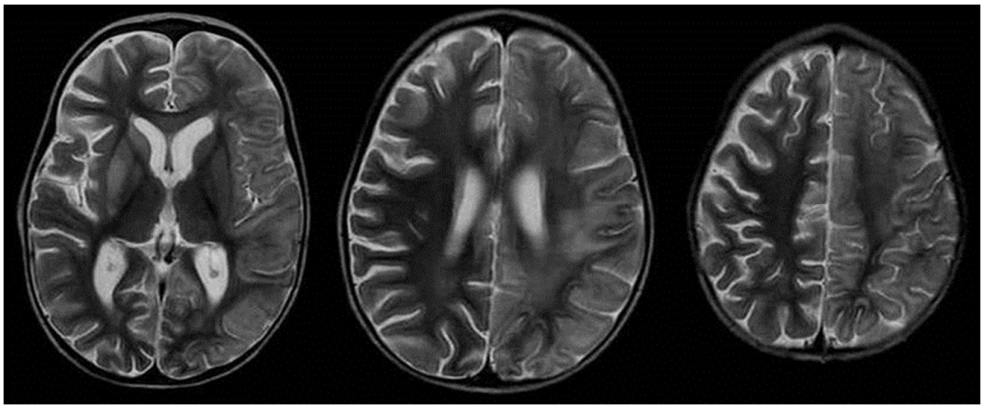


Photo – 3: Diffusion-weighted MRI (DWI) show a restriction of diffusion.

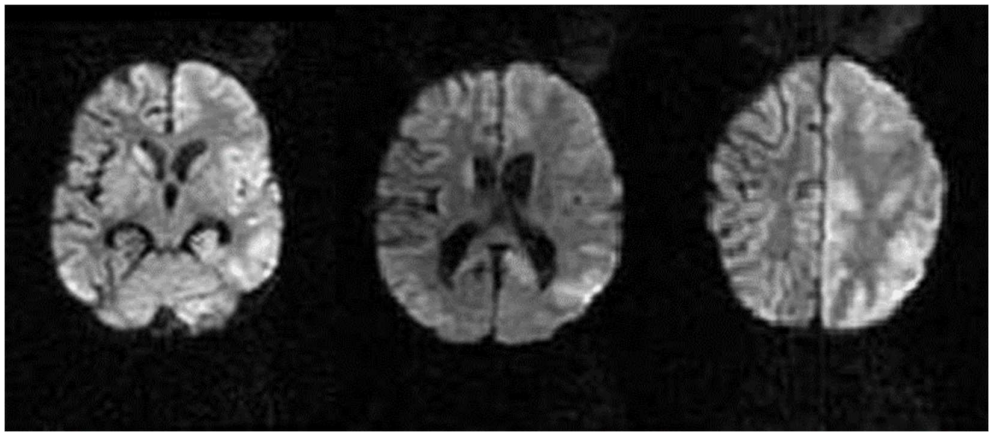
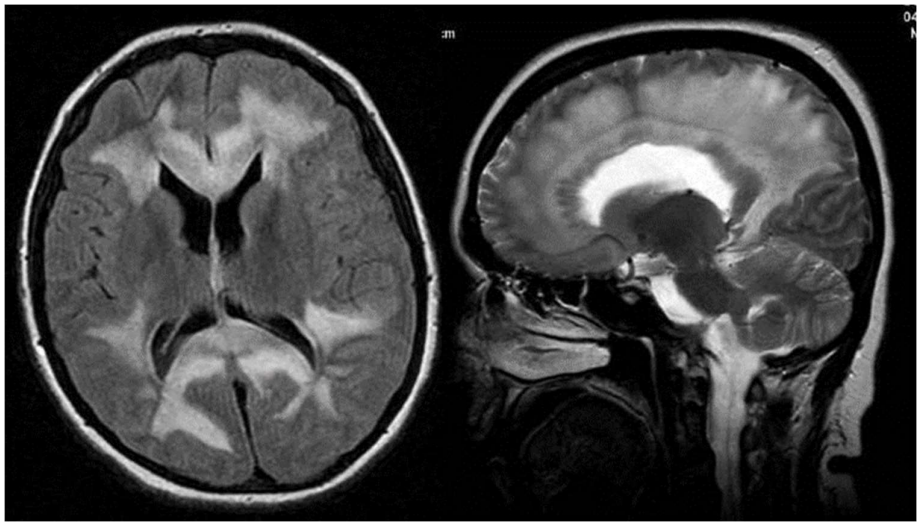
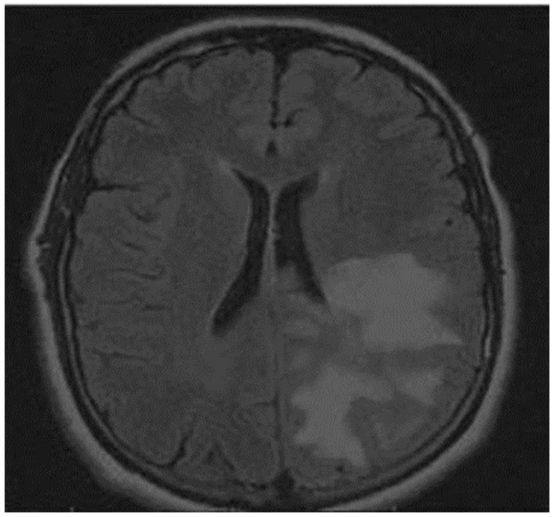


Photo – 4: MRI was done - Axial Flair brain images: Bilateral and asymmetricinvolvement of the periventricular and subcortical white matter.



Although ADEM is typically a disseminated process in the central nervous system, often with impaired sensorium, a few cases are dominated by spinal pathology.

Photo – 5: FLAIR sequence shows hyperintense signal involving periventricular, subcorticalwhite matter and grey matter on left side.



Conclusion

Acute disseminated encephalomyelitis (ADEM) is recognized as a potentially severe demyelinating disorder of the central nervous system (CNS). With advancements in medical imaging technology, particularly magnetic resonance imaging (MRI), the diagnosis of ADEM has become increasingly reliant on neuroimaging studies.

MRI is considered the investigation of choice for establishing the diagnosis of ADEM. It provides valuable information regarding the presence, location, and extent of demyelinating lesions within the CNS. The characteristic findings on MRI, such as hyperintense lesions on T2-weighted and fluid attenuated inversion recovery (FLAIR) sequences, aid in the differentiation of ADEM from other neurological conditions.

While other investigations, such as cerebrospinal fluid analysis, may provide supportive information, they are seldom as diagnostically valuable as MRI in the context of ADEM. Therefore, early recognition and prompt utilization of MRI are essential for timely diagnosis and initiation of treatment.

Early diagnosis and prompt treatment of ADEM are crucial for reducing morbidity associated with the condition. Timely intervention can help mitigate neurological sequelae, improve patient outcomes, and enhance overall quality of life. Therefore, clinicians should maintain a high index of suspicion for ADEM in patients presenting with acute encephalopathy and promptly initiate diagnostic evaluation, including MRI, to facilitate early diagnosis and management

References

1. Tenembaum S, Chitnis T, Ness J, Hahn JS. Acute disseminated encephalomyelitis. Neurology, 2007; 68(suppl 2): S23-S36.

2. Dale RC. Acute disseminated encephalomyelitis. Semin Pediatr InfectDis., 2003; 14(2): 90-5.

3. Rust RS. Multiple sclerosis, acute disseminated encephalomyelitis, and related conditions. Semin Pediatr Neurol., 2000; 7(2): 66-90.

4. Kleiman M, Brunquell P. Acute disseminated encephalomyelitis:response to intravenousimmunoglobulin. J Child Neurol., 1995; 10(6): 481-3.

5. Krupp LB, Tardieu M, Amato MP,Banwell B, Chitnis T, Dale RC, et al. International Pediatric Multiple Sclerosis Study Group criteria for pediatric multiple sclerosis and immune-mediated central nervous system demyelinating disorders: revisions to the 2007definitions. Mult Scler., 2013; 19(10): 1261-7.