IJNRD.ORG ISSN : 2456-4184
INTERNATIONAL JOURNAL OF NOVEL RESEARCH AND DEVELOPMENT (IJNRD) | IJNRD.ORG
An International Open Access, Peer-reviewed, Refereed Journal

Multiphasic Acute Disseminated Encephalomyelitis (ADEM) - A Rare Case Report

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Abstract

Background: Acute Disseminated Encephalomyelitis (ADEM) is an immune-mediated demyelinating disorder of the central nervous system (CNS), often triggered by infections or vaccinations. While traditionally considered monophasic, recurrent and multiphasic forms (MDEM) have been increasingly recognized.

Objective: This study aims to provide an overview of MDEM, including its clinical presentation, imaging findings, differential diagnoses, and management strategies.

Methods: A case study of a 12-year-old female with progressive cerebellar ataxia is presented, alongside a review of relevant literature. Clinical history, neurological examination, imaging, and laboratory investigations were analyzed to establish a definitive diagnosis.

Results: The patient exhibited chronic progressive cerebellar ataxia, speech abnormalities, and tremors, with MRI findings suggestive of multiphasic ADEM. Investigations ruled out autoimmune, infectious, metabolic, and degenerative causes. Treatment was initiated with immunotherapy, leading to clinical stabilization.

Conclusion: MDEM remains a diagnostic challenge due to its overlap with other demyelinating disorders. Early recognition and aggressive immunotherapy can improve outcomes. Further research is needed to establish optimal treatment protocols and long-term prognosis**.**

Keywords: Acute Disseminated Encephalomyelitis (ADEM), Multiphasic ADEM (MDEM), Demyelinating Disorders, CNS Autoimmune Disorders, Pediatric Neurology, Neuroinflammation, MRI Brain, Immunotherapy, Ataxia, Myelin Oligodendrocyte Glycoprotein (MOG).

Introduction :

Acute Disseminated Encephalomyelitis (ADEM): is a postinfection, postimmunization disorder, also called parainfectious encephalomyelitis. Once considered a purely monophasic illness, recurrent and multiphasic forms (MDEM) of ADEM are now recognized. ADEM is an immune-mediated CNS demyelinating disorder.

More common in childhood, typically between 5 and 8 years of age. The majority of children with ADEM have a nonspecific febrile illness preceding onset. Monophasic ADEM is the most common type. Recurrent ADEM is

characterized by a second episode occurring within 2 years after the initial illness, involving the same anatomic area as the original illness.

Multiphasic ADEM (MDEM): Characterized by one or more subsequent events involving a different anatomic area, demonstrated by a new lesion on MRI or a new focal neurologic deficit. MDEM is more common in children and frequently associated with myelin oligodendrocyte glycoprotein (MOG) seropositivity. More than half of all patients recover completely within 1-2 months after onset, whereas approximately 20% experience some residual functional impairment. Overall mortality in recent series is low.

ADEM: IMAGING

Brain:
• Multifocal T2/FLAIR hyperintensities → Bilateral but asymmetric white matter lesions, small round/ovoid lesions, hazy flocculent "cotton balls" (>2 cm, common in children), ± Basal ganglia, posterior fossa, cranial nerves.
• Enhancement varies → Multifocal punctate, linear, partial ring; can be perivenular; large "tumefactive" lesions less common.

Spinal Cord:

• Patchy/longitudinally extensive T2 hyperintensity.

• Strong but patchy enhancement

Case:

Patient Details: 12-year-old female, normal vaginal home delivery, achieved milestones appropriately, dropped out in 5th standard.

Chief Complaints: Progressive difficulty in walking (4-5 yrs), unsteady gait, swaying, worsened in 1.5 months (now dependent). Tremulous hands (3-4 yrs), worsened by holding objects. Difficulty eating (food spillage). Speech abnormality (1 yr) - slurred, loud, explosive, comprehension intact. Recurrent LRTI (4-5 yrs). Abdominal pain, vomiting (2 months ago).

No H/O: Alcohol/drug/toxin intake, eye-head lag, abnormal involuntary movements, chest pain, palpitations, DOE, joint pain, oral ulcers, hair loss, photosensitivity. **Family H/O:** Negative.

General Examination: Conjunctival telangiectasia, mild malar rash. **CVS:** S1/S2 heard, no murmurs. **RS:** Clear. **P/A:** Soft, non-tender, no organomegaly.

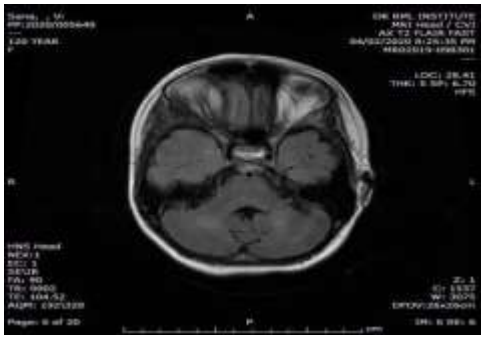
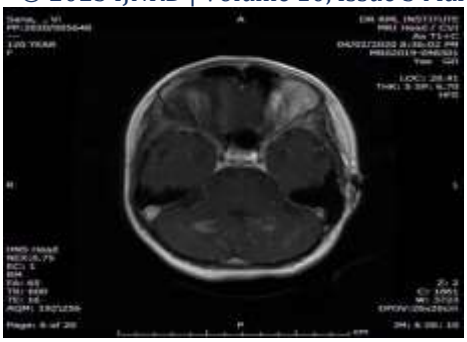
Neurological Examination: MMSE: 30/30. **Fundus:** Normal. **Speech:** Scanning type. **Tone:** Decreased (all limbs). **Power:** 5/5 (all limbs). **Sensation:** Normal. **Cerebellar Signs:** Dysdiadochokinesia, gaze-evoked nystagmus, impaired finger-nose, knee-heel test, truncal/stance ataxia, impaired tandem walking, broad-based gait with sway.

Clinical Diagnosis: Chronic symmetrical progressive cerebellar ataxia.

Differential Diagnoses: • **Degenerative:** Ataxia-Telangiectasia, ARCA1, Ataxia w/ Oculomotor Apraxia (Type 1/2), Friedreich's Ataxia, SCA (1,2,3,7,13) • **Autoimmune:** Ataxia w/ gluten sensitivity • **Inflammatory:** Multiphasic ADEM, NMOSD, SLE • **Infective:** HIV, Whipple's, EBV, VZV, HSV1, Lyme • **Toxic/Metabolic:** Hypothyroid, Vit B1 def, Phenytoin

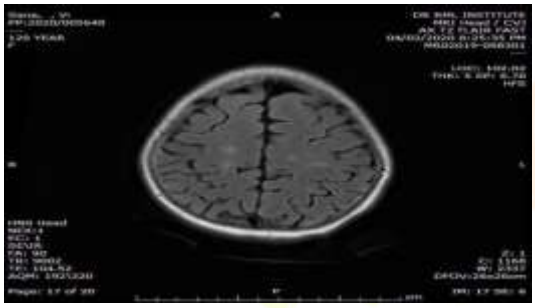
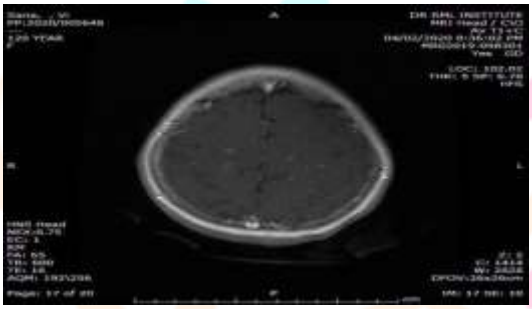
Investigations: • **MRI Brain:** **Axial T2 FLAIR** S/o Bilateral asymmetric hyperintensity of cerebellar hemisphere with involvement of dentate nuclues .

Axial T1 Contrast : Post contrast enhancement of bilateral hyperintensity of cerebellar hemisphere with involvement of dentate nuclues

• **MRI Brain:** **Axial T2 FLAIR** S/o Bilateral asymmetric hyperintensity of white matter in frontal region

Axial T1 Contrast : Post contrast enhancement of bilateral asymmetric hyperintensity in frontal region

• **CBC:** Hb: 11.8, WBC: 7700, PLT: 298K • **RFT:** Creat: 0.32, Urea: 11, Na: 138, K: 3.71, RBS: 102 • **LFT:** Bili: 0.41, SGOT/PT: 26/13 • **Coagulation:** PT-INR: 13.3/1.02 • **Lipid Profile:** Chol: 176, HDL: 33.3, LDL: 113, VLDL: 30, TG: 128 • **Immunoglobulins:** IgA, IgM, IgG - Normal • **Viral Serology:** HIV, HBsAg, HCV: Negative • **Tumor Markers:** AFP: 2.30, CEA: 0.01 • **Thyroid:** T3: 145, T4: 8.2, TSH: 1.69 (N) • **Autoimmune:** ANA: ++ (1:80, speckled), PCNA: +, dsDNA: - • **Lactate:** 44.9 mg/dL • **CSF:** Normal, JE/HSV: Negative, CALAS: Negative, GeneXpert: Negative • **Vitamin B12:** Normal • **2D Echo:** Normal • **USG Abdomen:** Normal • **NCS:** Normal • **VEP:** Normal

Final Diagnosis: Multiphasic Acute Disseminated Encephalomyelitis (ADEM)

Discussion :

Multiphasic Acute Disseminated Encephalomyelitis (MDEM) is a rare, immune-mediated, demyelinating disorder of the central nervous system (CNS), characterized by recurrent episodes affecting different anatomical locations. Unlike monophasic ADEM, MDEM presents with multiple relapses over time, often with new neurological deficits and MRI lesions.

In this case, a **12-year-old female** presented with **progressive cerebellar ataxia, speech abnormalities, tremors, and worsening gait**, with a history of **recurrent lower respiratory tract infections (LRTI)**. These findings raised suspicion for a **chronic, immune-mediated demyelinating disorder**. The differential diagnoses included **degenerative ataxias, autoimmune conditions (NMOSD, SLE), infectious causes (HIV, Whipple's, Lyme), and metabolic/toxic etiologies**. However, the **clinical course, MRI findings (multifocal T2/FLAIR hyperintensities with asymmetric white matter involvement), and autoimmune markers (ANA positivity)** strongly supported **MDEM**.

Pathophysiology

ADEM and MDEM are believed to result from **molecular mimicry**, where a prior infection or immunization triggers an autoimmune response against CNS myelin. Myelin oligodendrocyte glycoprotein (MOG) antibodies are frequently implicated, especially in pediatric cases.

Imaging & Investigations

MRI findings in this patient were **consistent with MDEM**, showing:

- Bilateral, asymmetric white matter lesions
- Multifocal hyperintensities on T2/FLAIR
- Patchy enhancement without tumefactive lesions

Laboratory tests ruled out infectious, metabolic, and degenerative causes. The presence of **ANA positivity with a speckled pattern** suggested a potential autoimmune process, though **dsDNA negativity** ruled out active systemic lupus erythematosus (SLE).

Management & Prognosis

Treatment primarily involves **high-dose corticosteroids (IV methylprednisolone), IV immunoglobulins (IVIG), or plasmapheresis** in severe cases. Immunomodulatory therapy, such as rituximab, may be considered in recurrent cases.

Prognosis in MDEM is generally **favorable**, with **most patients achieving significant recovery within 1–2 months**. However, **20% may have residual neurological deficits**. Long-term follow-up is essential to **monitor relapses and distinguish MDEM from multiple sclerosis (MS) or neuromyelitis optica spectrum disorder (NMOSD)**.

Conclusion

This case highlights the **diagnostic complexity** of **chronic progressive cerebellar ataxia in children**. The **clinical presentation, imaging, and autoimmune markers** strongly favored **Multiphasic ADEM**. Early recognition and immunotherapy are crucial for **optimal neurological recovery and preventing long-term disability**.

Ethical Compliance Statement

This study complied with all ethical standards and did not require approval from the Institutional Ethics Committee. Written informed patient consent was obtained. All authors have read and complied with the journal's ethical publication guidelines.

Financial Support and Sponsorship
Nil.

Conflicts of Interest
There are no conflicts of interest.

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