http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v10i10.22

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Case Report

Extensive acute disseminated encephalomyelitis with spinal cord involvement in a young boy responded with intravenous methylprednisolone

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Abstract
Acute disseminated encephalomyelitis is an immune mediated demyelination of central nervous system. It is a monophasic clinical syndrome though recurrences have been reported in rare cases which can have widespread involvement in brain and spinal cord. This syndrome usually seen in children and young adults following infection or vaccination. ADEM is characterised pathologically by perivascular inflammation, demyelination and edema within the brain and spinal cord. Clinically manifesting in wide varieties from focal to multifocal neurological dysfunction. We describe a case of 16-years old boy who presented to us with altered sensorium following a seizure episode with history of fever with loose stool since 4 days. After two days during hospital stay he developed weakness in all four limbs with bladder involvement. MRI brain showed multifocal cortical and sub-cortical edema involving bilateral frontal, parietal, temporal regions with partial effacement of adjacent sulcal spaces. MRI Spine showed hyperintense signal on T_2/STIR images involving cervical as well as dorsal cord particularly marked in lower cervical region extending from C_4-C_7 level with mild post contrast enhancement of involved parenchyma. He was diagnosed as ADEM and started on intravenous methylprednisolone for 5 days followed by oral prednisolone. He showed drastic improvement in weakness and at one month follow up he was normal clinically with resolution of radiological lesions.

Keywords: *Acute disseminated encephalomyelitis–ADEM, Quadriparesis, Methylprednisolone.*

Introduction

Acute disseminated encephalomyelitis is an immune mediated demyelination of central nervous system. It is a monophasic clinical syndrome though recurrences have been reported in rare cases which can have widespread involvement in brain and spinal cord. This syndrome usually seen in children and young adults following infection or vaccination. Many viral pathogens are implicated in its causation including Measles, Influenza, Epstein barr virus, Cytomegalovirus, Human herpes virus, Hepatitis A & B, Rubella and HIV. Similarly many vaccinations like diphtheria, tetanus, pertussis, rabies, measles Japanese encephalitis, hepatitis B, polio and influenza can leads to ADEM.[1] ADEM

is characterised pathologically by perivascular inflammation, demyelination and edema within the brain and spinal cord[11].Molecular mimicry is an important mechanism of immune activation due to sharing of epitopes between self-antigen in CNS and foreign antigen presented by pathogens or vaccinations.[2] clinical manifestation of ADEM includes altered sensorium, motor manifestation like hemiparesis, paraparesis, quadriparesis, cranial nerve involvement, seizure and optic neuritis.[1]Treatment modality of choice in management of ADEM includes controlling immune response against neural tissue by using steroid therapy. Other treatment option in unresponsive or refractory cases are plasma therapy, intravenous immunoglobulins. Here, we are describing a case of ADEM who presented with seizure followed by altered sensorium with history of Fever and loose stool 4 days back. After two days during hospital stay patient developed quadriparesis with urinary retention. This patient was treated with intravenous methylprednisolone for 5 days followed by oral prednisolone with clinical and radiological improvement at one month follow up.

Case Presentation

A 16 years old boys was brought by his father in altered sensorium following an episode of abnormal body movements involving all four limbs lasting for about 30 seconds with history of Fever and loose stool since 4 days. Fever was high grade, intermittent, not associated with chills and rigors, no rash, no burning micturition. Loose stool was sudden in onset, 3-4 episodes per day, watery in consistency, and few episodes of vomiting present, not associated with blood or pain abdomen. No history of preceding headache, blurring of vision, diplopia, Cough with or without hemoptysis. No significant past medical or surgical history. No history of recent immunization. On admission patient was in altered sensorium with GCS- $E_2V_1M_5$, Pulse rate-110/min, Blood pressure-100/58mmHg, Random blood glucose-156mg/dl, SpO_2- 86% on Room air, 94% with oxygen. Neck rigidity was absent, no other sign of meningismus. After few hours patient gained sensorium but was drowsy and maintaining SpO_2 on room air. Patient started on intravenous antibiotics and epileptics. No further episodes of fever, loose stool and seizure documented. On day 2 of admission patient started complaining of weakness in bilateral upper and lower limb which was sudden in onset, was more marked in lower limb with simultaneous onset in all four limbs with urinary retention for which patient was catheterised. On examination patient was conscious, oriented. Cranial nerves and fundoscopic examinations were normal. Motor system examination revealed mild hypertonia in all four limbs with power of 4/5 in bilateral upper limbs and 3/5 in bilateral lower limbs by Medical research council (MRC) grading. Deep tendon reflexes was exaggerated in bilateral upper and lower limbs with planter response was extensor bilaterally. Sensory system examination revealed reduced touch, pain, temperature sensation in all four limbs, no distinct upper level of sensory loss, with intact dorsal column sensation.

Investigation

1. Complete hemogram

Parameters	**Day1**	**Day2**	**Day3**	**Day4**	**Day 5**	**Day 6**
Hb	11.0	11.3	12.2	12.9	11.6	11.3
TLC	29100	13500	18600	13900	11000	8400
DLC	85/09/01/05	84/15/0/01	86/06/07/01	80/15/03/02	71/190/9/01	63/25/09/03
Plt Count	1.88 Lac	1.53 Lac	2.02 Lac	1.94 Lac	1.80 Lac	1.95 Lac

2. Biochemistry

	Day1	Day2	Day3	Day4	Day5	Day6
Blood Urea	20	29	42		33	22
Serum Cr	0.7	0.8	0.7		0.7	0.6
Ca^{++}	8.0	9.7	10.6		9.7	8.4
Na^{+}	139	137	138		138	
K^{+}	5.1	4.3	4.3		3.7	
AST	25	26	22		29	
ALT	45	58	41		37	
T. Bil	0.3	0.3	0.5		0.5	

In view of fever with seizure and altered sensorium CSF study was done which revealed colourless fluid with sugar level- 86mg/dl, protein level- 30mg/dl, acellular. CSF fluid gram stain, AFB stain and culture was negative. MRI brain showed multifocal cortical and sub-cortical T_2W hyper intensity involving bilateral frontal, parietal, temporal regions with partial effacement of adjacent sulcal spaces (**Figure 2 A-C**). MRI spine showed myelitis involving the cervical as well as dorsal cord with predominant involvement of cervical cord showing relatively swollen cord contours and mild post contrast enhancement of the involved parenchyma. Involved cord appears hypointense on T1W images and showed hyperintense signal on T2/STIR images (**Figure 1 A, B**). Covid19 RT-PCR was negative. CSF ELISA for Herpes simplex virus, Cytomegalovirus, HIV, Japanese encephalitis and Dengue was negative. Patient diagnosed as acute disseminated encephalomyelitis –post infectious with spinal cord involvement. Patient started on IV methylprednisolone 750mg once daily for 5 days followed by oral prednisolone 30mg once daily, which was continued for 4 weeks followed by slow tapering. Patient showed clinical improvement dramatically on Day 4 of steroid with steady gain of power in all four limbs, since than patient had gradual improvement in power. At one month follow up patient was clinically normal with resumption of all activities. Repeat MRI Brain and spinal cord done to document radiological improvement. Repeat MRI brain showed multiple subcortical areas of white matter hyperintensity in bilateral fronto-parietal and temporal lobes and with comparison to previous MRI there was significant resolution of lesions (**Figure 4 A-C**). MRI spine was normal and indicating complete resolution of lesions (**Figure 3**).

Discussion

ADEM forms 75% of cases of primary inflammatory demyelinating disorders of CNS with estimated annual incidence of 0.4-0.8 per 1,00,000.[3,4,5] Symptoms of ADEM develops 4-21days after the inciting event [1] but if inciting event is infection with subclinical onset and short latent period between infection and clinical symptoms than it may become difficult to differentiate the ADEM from acute viral encephalitis particularly in India where many viral encephalitis are endemic like Japanese encephalitis. It is very important to differentiate viral encephalitis from ADEM as steroid is the main therapy of ADEM which may leads to worsening of viral encephalitis.

ADEM involves both white and grey matter of CNS and like MS it can demonstrate periventricular brain lesion but in ADEM it is more likely ill defined, larger and more rounded lesions with predilection for deep gray matter and brain stem.[6,7] Spinal cord and brainstem involvement seen in approximately one third of patients with ADEM and its spinal characteristics are helpful in differentiating this condition from MS.8 these lesions are hyperintense on T_2 & FLAIR images and are usually of same age. Blood brain barrier disruption is seen in ADEM which is patchy and mainly due to perivenular inflammation.

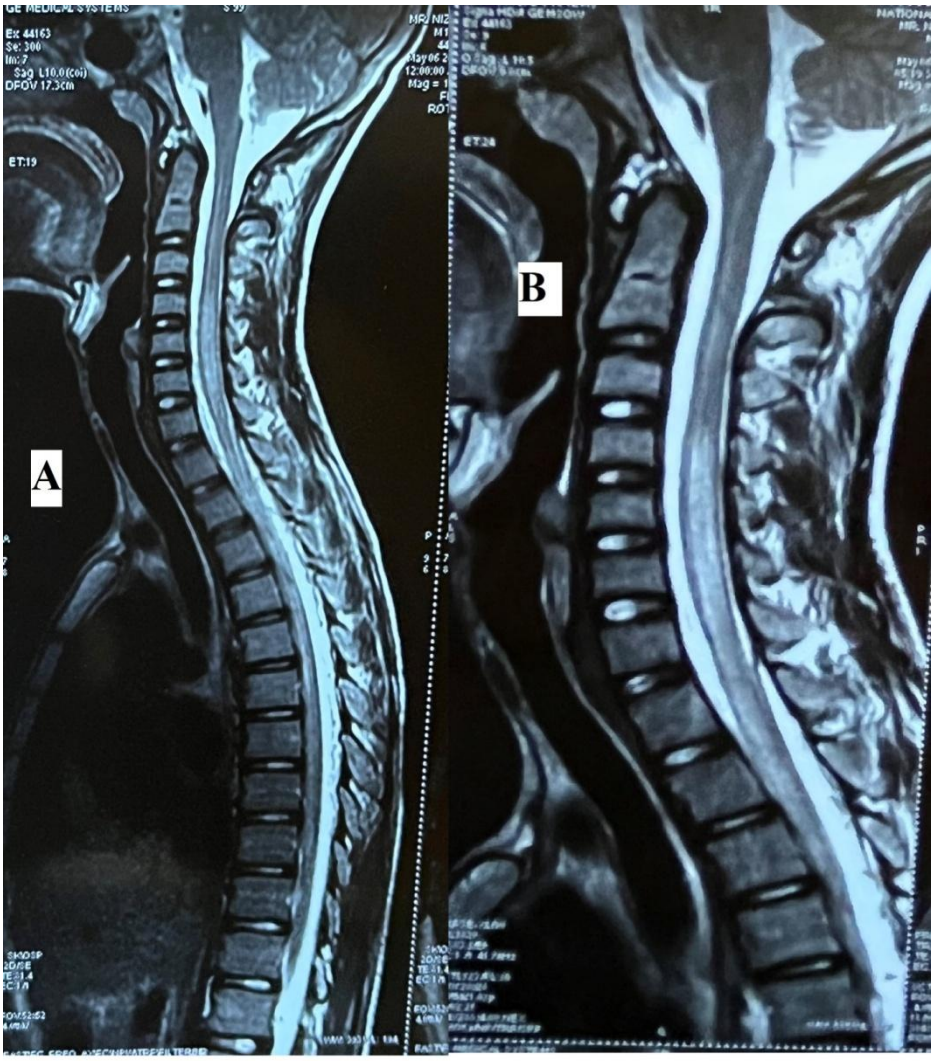

Figure 1 : A and B are T2-weighted sagittal section of spinal cord
showing hyperintense signal involving cervical as well as dorsal cord with relatively swollen cord contours particularly marked in lower cervical region extending from C4 down to C7.

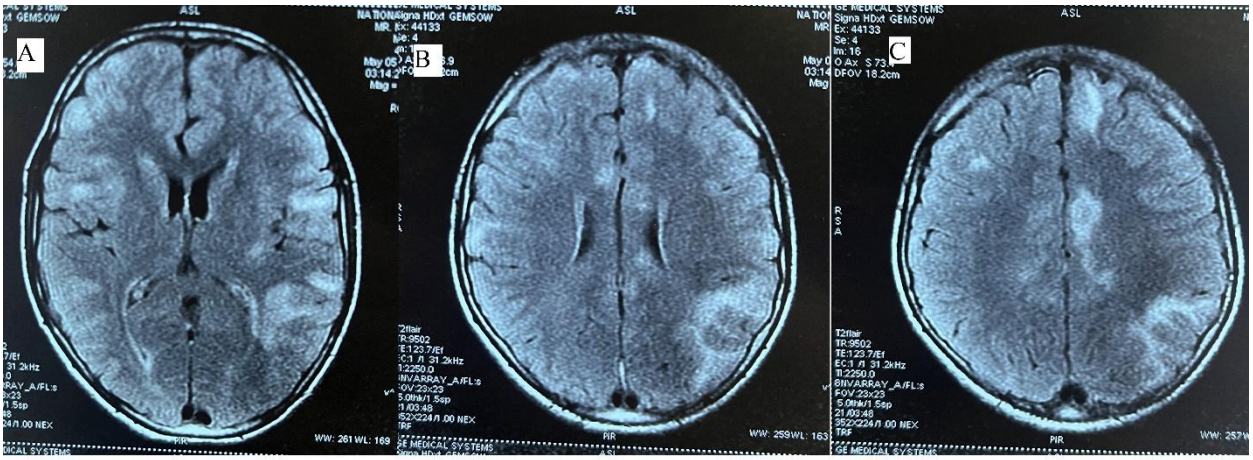

Figure 2: A,B,C are Axial section T2 FLAIR Images of brain showing hyperintense signal involving cortical and subcortical regions of bilateral frontal, parietal and temporal lobes

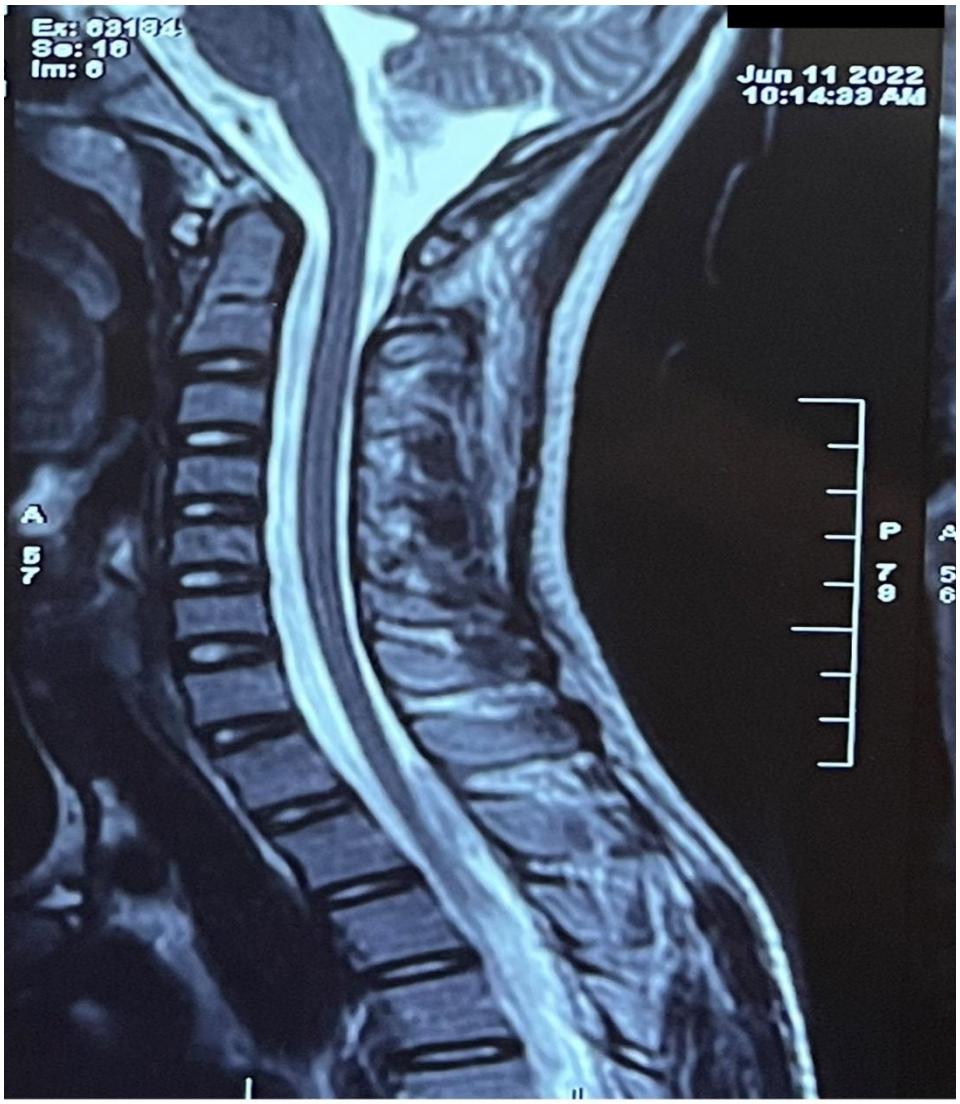

Figure 3: T2-weighted sagittal image of the spinal cord after 1 month showing complete resolution of lesions.

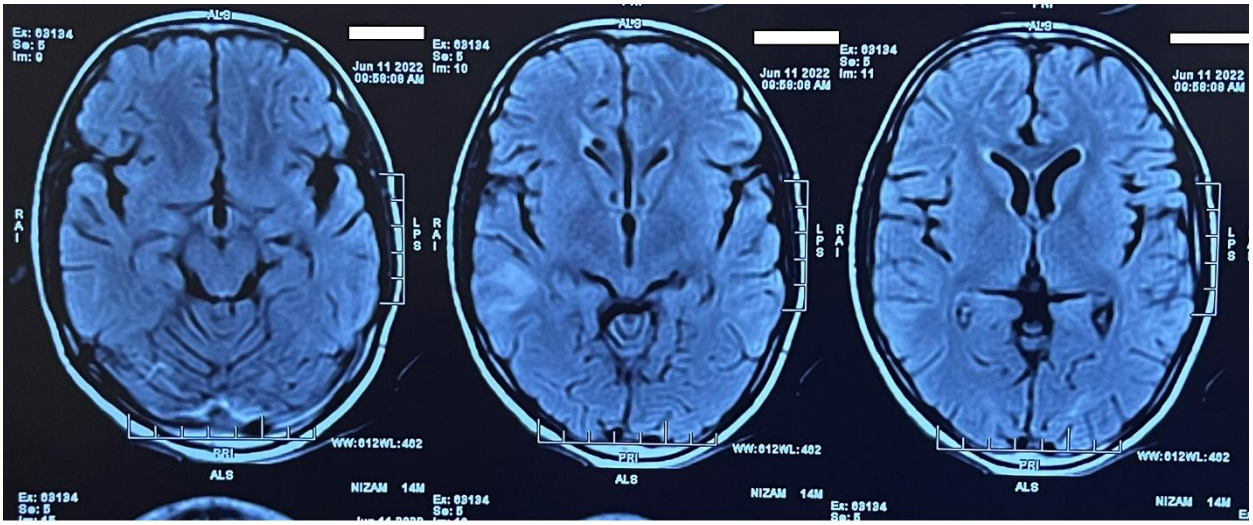

Figure 4: (A–C) T2 FLAIR MRI of the brain after 1 month showing significant resolution of lesions.

Hence the contrast enhancement pattern is variable as nodular, diffuse, gyral complete and incomplete ring. In our case MRI brain features are not characteristics for ADEM but simultaneous involvement of spinal cord favouring the diagnosis of ADEM. Absence of CSF pleocytosis and negative CSF ELISA for common viruses ruling out the viral encephalitis which is further supported by simultaneous extensive involvement of spinal cord which is unusual in acute viral encephalitis. We made the diagnosis of ADEM and started the patient on intravenous methylprednisolone 30mg/kg for 5 days along with supportive care and physiotherapy. Patient started on oral prednisolone after 5 days at dose of 1mg/kg/day for 4 weeks. Treatment modality of choice in management of ADEM includes controlling immune response against neural tissue by using steroid therapy. Steroid therapy as methylprednisolone/ dexamethasone for 3-5 days is usually used as first line therapy for ADEM .[4, 9, 10] This therapy needs to be tapered for 4-6 weeks as shorter tapering period has been linked with high tendency of relapse.[9,11] It is also recommended a combination therapy of steroid with immunoglobulin for patients who do not respond adequately to steroid alone.[4] Plasma exchange therapy is to be used in unresponsive or refractory cases of ADEM. There is no advantage of its early use.[4, 12] ADEM has good prognosis with full recovery but few reported cases displayed minor residual neurological deficit.[4, 12] In our case gradual clinical improvement was observed over 1 week with steadily improvement in power in all four limbs. Patient regained complete activities in one month. A repeat MRI brain show significant resolution of lesion with complete resolution of lesions in MRI spine.

Conclusion

This case exemplify the importance of early diagnosis and timely initiation of steroid therapy in ADEM as delay in initiation of therapy can lead to permanent neurological sequelae. Our patient received early steroid therapy leading to complete clinical resolution.

Patient's Consent: A full and detailed consent from the guardian has been taken. The patient's identity has been adequately anonymized.

Conflict of Interest: None

Disclosure: The authors hereby certify that the work shown here is genuine, original and not submitted anywhere, either in part or full. They transfer the full rights of the video to Neurology India. All the necessary permissions from the patient, hospital and institution has been taken for submitting this video to Neurology India

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