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Acute Disseminated Encephalo Myletis [ADEM]

Subhasri Balaji[1], Kedarnath Reddy[2]

Abstract: ***ADEM is a childhood acquired demyelinating disorder occurring following a viral infection or vaccination. It presents with acute onset of polyfocal neurological deficits accompanied by encephalopathy and changes compatible with demyelination on brain MRI. It is treated with pulse therapy and IV steroids which results in clinical improvement***

Keywords: ADEM, Demyelinating, post infection, post vaccination, disseminated, focal deficits, encephalopathy, MRI Brain, IV Pulse steroids

1. Introduction

- Acute disseminated encephalomyelitis (ADEM) is an inflammatory demyelinating event of CNS presenting with acute onset of polyfocal neurologic deficits accompanied by encephalopathy and changes compatible with demyelination on brain MRI.
- It occurs following an episode of fever due to infection or post vaccination
- It affects children in the age group of 5 to 8 years with slight male preponderance.
- Encephalopathy is the hallmark of ADEM. Initial symptoms include fever, lethargy, headache, vomiting, seizures
- Focal neurological deficits are seen in ADEM and include visual lossataxia, motor and sensory deficits, bladder bowel dysfunction.

2. Case Report

A 6year old female child presented to our hospital with diminution of vision. History of fever since 5 days prior to the onset of illness. Fever was moderate grade, intermittent, not associated with rigors.

History of non - productive cough and running nose for 3 days

On day 6 of fever, child developed blurring of vision in both eyes followed by difficulty in reaching out to objects followed by complete loss of vision. No complaints of convulsions, headache, vomitings.

No complaints of falls or unsteadiness in gait

Child had normal perinatal history, normal developmental milestones

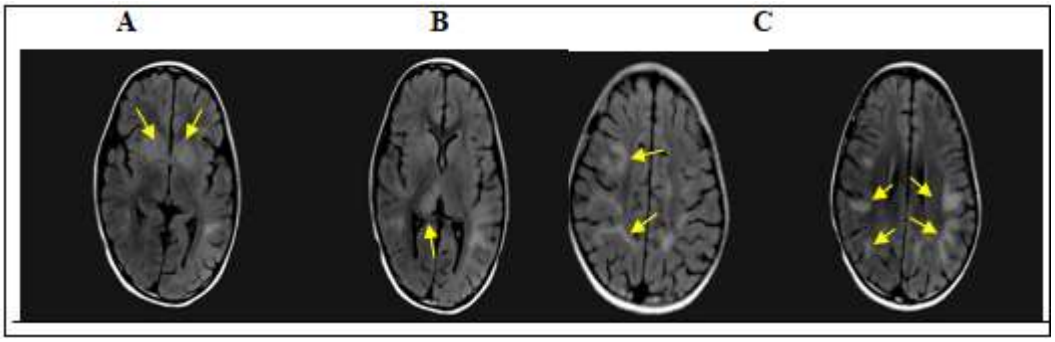
O/E
Child was conscious, coherent

Pupil: Bilateral measured 4mm, sluggishly reacted to light

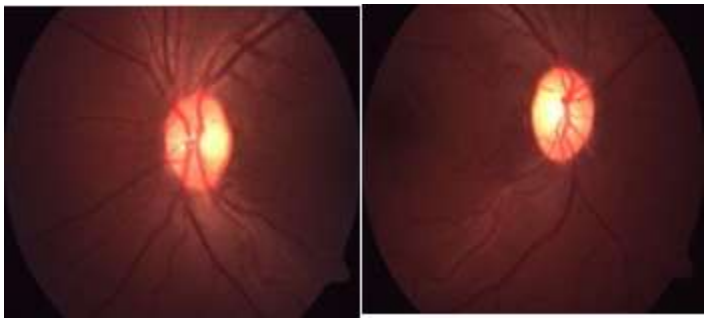
No facial asymmetry & Extra Occular movement intact

Motor System Examination:

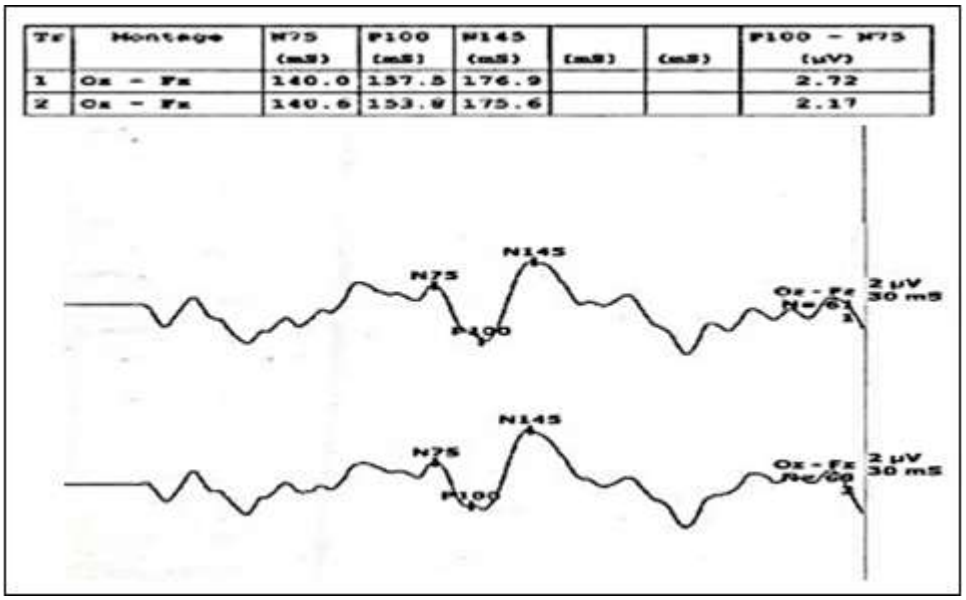
	R	L
T ONE	N	N
POWER	Paucity of movements	
DTR	1+	1+
PLANTAR	↓	↓



Hyperintensities in **A**. B/L Basal ganglion **B**., Right Thalamus, **C**. Subcorticalwhite matter



B/L Hyperaemic Optic Discs



Prolonged P100 latency - in both eyes, Amplitude within Normal limits s/o B/L Optic Neuritis

Clinical Course

3. Investigations

Child was evaluated with the following investigations:

1) MRI BRAIN: Multiple patchy FLAIR hyperintensities in bilateral caudate, lentiform and right thalamus and subcortical white matter in bilateral cerebral parenchyma with diffusion restriction without ADC drop.
2) CSFANALYSIS: Elevated CSF Proteins
3) FUNDUS EXAMINATION: Disc hyperemia and tortuosity of blood vessels in both eyes suggestive of bilateral Optic neuritis,
4) VISUAL EVOKED POTENTIAL: P100 Latency in both eyes is prolonged, amplitude is within normal limits suggestive of bilateral optic neuritis.

4. Treatment

In our case, child was treated with IV. methylprednisone pulse therapy 30mg/kg/dose, 450mg diluted in 50ml NS for 5days.

There was marked improvement in vision from day 3 of the treatment and child was discharged on oral prednisolone 30mg daily with a tapering plan. Child was advised to attend for follow up after 6w wherein there were no new symptoms suggesting relapse. MRI brain was repeated which showed reduction in size of the T2/FLAIR hyper intense lesions. Fundus examination in both eyes showed normal optic disc with sharp margins and vessels having normal course and caliber. VEP showed an improvement in response in both eyes

5. Discussion

1) ADEM which is a childhood acquired demyelinating inflammatory disorder of CNS heralded by an antecedent viral infections like influenza, measles, varicella, mumps, HSV - 1, EBV or a preceding vaccination with H1N1 MMR, JE, DPT which induce a molecular mimicry to produce CNS auto antigens
2) The clinical course consists of a non specific prodrome of fever, cough, myalgias for 3 - 4days followed by neurological symptoms. Encephalopathy, the hallmark of ADEM ranges from mild irritability to severe form of somnolence, obtundation, coma.
3) Other findings include seizures, vomitings, headache, cranial nerve palsies, diminution of vision, cerebellar signs like ataxia, nystagmus
4) Glucocorticoids are the mainstay of treatment
   a) IV methylprednisolone: 20– 30 mg/kg/day x 5 days (maximum –upto1000mg/day) followed byOralprednisolone: 1 - 2mg/kg/day (maximum upto 40 - 0mg/day) over 4–6weeks
   b) IVIG: 2g/kg given over 2– 5 days
   c) Plasmapheresis (5–7 Exchanges A/D in refractory cases)

6. Conclusion

ADEM which is a childhood acquired demyelinating disorder of CNS occurring following a viral infection or post vaccination which when treated with pulse therapy of IV steroids results in brisk improvement in most children with resolution of clinical and radiological features. IVIG is initiated if there is no clinical improvement within 7 days of completing pulse steroids

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