Acute disseminated encephalomyelitis: a rare case report of a 15 years old from Bangladesh

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ABSTRACT

Acute disseminated encephalomyelitis (ADEM) is rare autoimmune neurological multifocal disorder characterised by inflammatory demyelination of brain and spinal cord usually in response to infection or sequelae of immunisation, most commonly affecting the children.Though a rare one it is estimated that 1 in 125,000-250,000 population is affected by ADEM each year. And such a case of 15 years old girl presented with acute onset of fever and headache and single episode of seizure followed by unconsciousness with GCS 6(E2V2M2).signs of meningeal irritation like neck rigidity, Kernig's sign and Brudzinski's sign were positive. investigation report showed Hb-11.4 g/dl ,neutrophlic leukocytosis , normal range of serum electrolytes ,ICT for malaria was negative and MRI of brains showed multiple abnormal T2 hyperintense signal in both parieto-frontal white matter. so she was diagnosed a case of ADEM and treated with intravenous methylprednisolone for 5 days followed by oral corticosteroid in tapering dose for 6 weeks.

Introduction

ADEM also known as post -infectious encephalomyelitis is a rare autoimmune inflammatory neurological disorder of nervous system mainly central which is characterised by demylination of the brain and spinal cord as a result of infection or sequelae of immunisation. Children are the most affected one but adult case of ADEM occurs following vaccination against Mumps,Measles and rubella(1)(2) . Though rare one estimated of 1 in 125,000-250,000 individuals are affected by ADEM each year(1). More commonly male are the most affected one with a male and female ratio of 1.3 to 1.The risk factors for ADEM include genetics, exposure to infectious organisms,immunization exposure and lighter skin pigmentation. All the ethnic groups are affected globally. The majority of cases follow either viral or bacterial infection and in majority of cases causative pathogen is unidentified(1). The pathogenesis is not fully understood but antigentic exposure in genetically susceptible individuals may cause ADEM either through molecular mimicry and or due to T cell mediated immune mechanism. It has also been suggested that ADEM may occur due to CNS injury secondary to generalised inflammation of brain and spinal cord(4).ADEM patients usually have presentation like rapid onset of fever, headache ,nausea or vomiting confusion, fatigue and neurological features like numbness, difficulty swallowing, vision, seizure, difficult in coordinating voluntary movements such as walking and some degree of impairment of consciousness(1)(3). Though there is no diagnostic criteria for ADEM but presence of signs and symptoms and radiological findings can help in diagnosis of ADEM. In patients with ADEM T2-Weighted and FLAIR magnetic resonance imaging (MRI) of brain shows bilateral,hyperintense lesion that are diffuse and poorly demarcated(4).ADEM being a autoimmune disease treatment includes I/v Methyl prednisolone at dosage of 20 to 30 mg/kg (max 1g/day) for 3 to 5 days followed by oral corticosteroids in a tapering dose for 4 to 6 weeks.(5). Full recovery is seen in 50 to 70% of cases and ranging of 70 to 90%

with some minor residual disability. In some case severity ranges to 8 to 30% with ataxia and hemiparesis.(6)

Case report;

We hereby present a case of 15 years old girl brought to the Mymensingh Medical College by her parents with the complaints of fever and headache for 3 days. Fever was high grade ,acute in onset without any chills and rigor with no evening rise of temperature . Headache was severe and prolonged for 3 days following which she suddenly developed single episode of seizure followed by unconsciousness for which she was brought to the hospital.

On examination she had altered consciousness with GCS 6(E2V2M2),Temperature was raised, blood pressure was 100/80 mm of Hg and signs of meningeal irritation like neck rigidity, Kernig's sign and Brudzinski's sign were positive and bilateral planter extensor was found.Other systematic examination findings were normal. As per the clinical findings she was treated for meningoencephalitis with Inj.Ceftriaxone 1g I/V BD, inj. Acyclovir 250mg 1 vial I/v BD ,inj.dexamethasone 1 amp i/v TDS and investigation showed

Hb-11.4 g/dl with increased sedimentation, neutrophilic leukocytosis with WBC Count 25000/mmˆ3 and 87% of neutrophils, Serum electrolytes and blood glucose were within the normal limit.

ICT for malaria was negative .

MRI OF BRAIN revealed. -Multiple abnormal T2 hyperintense signal in both parieto-frontal white matter suggestive of acute disseminated encephalomyelitis(ADEM)

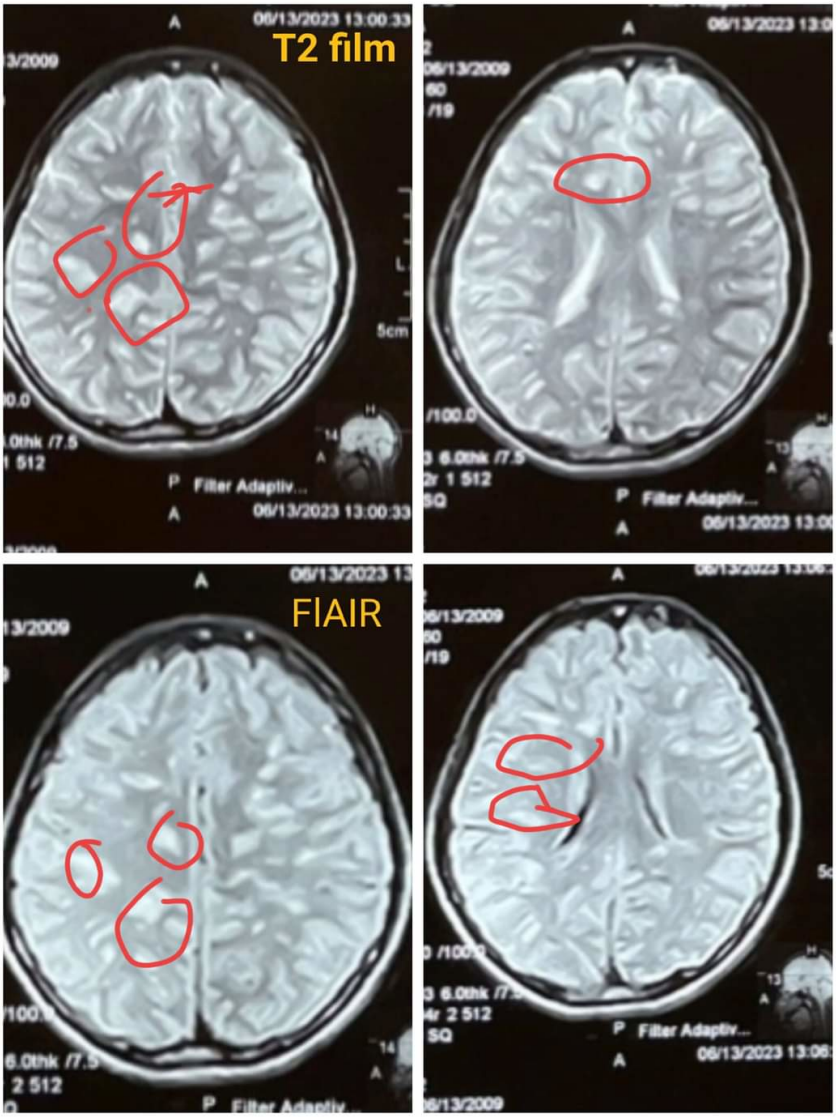

FIG 1:-MRI of brain showing T2 AND FLAIR view of temporo-parietal region

So she was diagnosed as case of ADEM and was treated accordingly with i/v methylprednisolone 20mg/kg body weight for 5 days followed by oral corticosteroids in tapering dose for 6 weeks.

Discussion

ADEM commonly referred to as post infectious encephalomyelitis affects the central nervous system and is characterised by demylination of the brain and spinal cord. (1) A rare autoimmune disorder known to be affecting 1 in 1,25,000-1,50,000 population most commonly affects the younger age group (2),presents with symptoms ranging initially from non specific such as fever, nausea, vomiting, fatigue, lethargy, irritability, malaise, nausea, weight loss, confusion, stupor and coma.(9)The specific neurological symptoms comprise of leg/arm weakness, tingling or numbness, Seizures,altered mental status, vision loss as it occasionally involves the optic nerve(1,9).The extent of presentations vary between the affecting individuals depending on the age of onset and the location of the lesions (9). In the later stages the patients can also present with ataxia,

slurring of speech and cranial nerve dysfunction.(1,2,9) Our patient presented with fever, headache and one episode of seizure with altered mental status, loss of consciousness, reduced GCS , signs of meningeal irritation bilateral plantar extensor mimicking meningoencephalitis.She was treated with empirical antibiotics and antiviral for meningoencephalitis. According to Suvasini sharma et al(10) headache and vomiting are reported in 38 to 52% of the patients, seizure in 13 to 47% , meningismus in 5 to 43%. Fever and seizure are more frequently associated with ADEM as compared to other demyelinating syndromes.(10) Seizures are common ranging from focal togeneralised(7).The exact pathophysiology isn't understood but it is most commonly considered to be associated with immune response following infections or vaccination .(1,2,9) There is no specific diagnostic criteria, a good history, clinical examination and relevant examination findings helps the establishment of the diagnosis. In two third of patients abnormal CSF findings are seen i.e. moderate pleocytosis with raised proteins and absence of oligoclonal bands.(7) MRI of the brain shows hyperintense lesion on T2 weighted, FLAIR, proton density and echo planar trace diffusion free sequences.(1)ADEM can also present as space occupying lesion(SOL) (7) involvement can be present either as a single lesion or in the form of multiple lesions .(1). Our patient's CSF findings were normal but the MRI of brain showed multiple abnormal T2 hyperintense signal in both parieto-frontal white matter.Treatment Options include high dose corticosteroids preferably methylprednisolone and dexamethasone.Intravenous immunoglobulin(IVIG) can be considered if the patient doesn't respond to steroids.Clinical trials also suggest plasmapheresis to be considered as one of the treatment Options. Rehabilitation therapy should be considered for those having cognitive or speech impairment, epilepsy , visual and motor problems (7). We started high dose IV methylprednisolone for our patient and she showed dramatic improvement following the treatment which was later followed by oral steroids. Clinical progress of ADEM is rapidly progressive with rapid deterioration within 2 to 5 days.Rarely owing to brain stem involvement, respiratory failure may occur (10) Children usually have a good prognosis but severe cases may have residual abnormalities, one fourth of the cases havea poor prognosis(8). On the first follow up our patient had mild generalised weakness otherwise she was normal.on the 2nd follow up after 2 weeks , she was completely fine with no any residual abnormalities. Conclusion

Though ADEM is a rare autoimmune neurological disorder with a incidence of 1 in 125,000 -250,000 it still can be treated by early diagnosis and treatment. The main goal of the authors here is to highlight a case of ADEM and where eary diagnosis and treatment was beneficial for the patient. Children are the ones who are affected by ADEM so it is very much crucial to suspect ADEM for early and diagnosis and treatment. After covid-19 and its vaccination cases of ADEM are increasing so we must keep in mind it can be ADEM if there will be presentation like that of this case.

Conflict of interest-None to disclose

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