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ACUTE NECROTIZING ENCEPHALOPATHY OF CHILDHOOD: A CASE REPORT FROM KATIHAR MEDICAL COLLEGE, BIHAR, INDIA.

Paediatrics

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ABSTRACT

Acute necrotizing encephalopathy of childhood (ANEC) previously thought as rare disease entity nearly exclusively seen in East Asian children including Japan and Taiwan. However, its sporadic cases have been reported from all over the world. The disease affects previously healthy young children and is characterized by respiratory or gastrointestinal infection, high grade fever accompanied by rapid alteration of consciousness and seizure. The hallmark of the disease is diffuse and symmetrical CNS lesions of thalami, brainstem, cerebellum, and white matter. The pathogenesis is still not known, most consider it secondarily after viral infection, however its genetic predisposition is also proposed. We were reported a case of 12 year old girl with ANEC hospitalized in Katihar Medical College and Hospital, Katihar and concluded that Acute Necrotizing Encephalopathy had poor outcome. And awareness for the detection of under diagnosed cases is needed to identify possible predisposing factors and improve the outcome of this dangerous acute illness of childhood.

KEYWORDS

Acute Necrotizing Encephalopathy, Seizure, Childhood, Bilateral Thalamic Involvement

INTRODUCTION:

Acute necrotizing encephalopathy of childhood is an atypical type of acute encephalopathy originally described in East Asia including Japan and Taiwan [1]. However; its sporadic cases have been reported from all around the world [2,3]. The exact etiology and pathogenesis of ANEC remain unclear; however, mycoplasma, influenza virus, herpes simplex virus, and human herpes virus-6 are among the most common infections that intensify the disease [4-6]. Although there is strong causal relationship between viral infection and ANEC, the exact pathogenesis remained unclear.It is believed that a "cytokine storm" resulting in brain injury and loss of immunomodulatory response is suspected to be the main pathogenesis of ANEC. Cytokines such as TNF-α and interleukins 1 and 6 can speed up the disease [6].

Imaging findings are the hallmark and are similar in cases of good or poor outcome. Thalamic involvement is a prerequisite, but lesions are also common in internal capsule, putamen, upper brainstem tegmentum, cerebral periventricular white matter, and cerebellar medulla, sparing other part central nervous system regions, such as anterior part of the putamen, subcortical white matter, optic nerve, pontine base, and spinal cord[7]. Despite the homogeneity of imaging and neuropathologic findings, the clinical expression of the disease ranges from a mild form of encephalopathy without sequelae to a fatal outcome [1,8,9]. However, the intensity of involvement and MRI lesions are clearly related with outcome[10].

There is no specific treatment or preventive measure for this disease and a poor prognosis with less than 10% of complete recovery is generally expected [11]. Most of them experience rapid neurological decline and death.

CASE REPORT:

A previously healthy 12 year old girl, product of non-consanguineous marriage, admitted in pediatric department with complain of sudden onset of high grade fever for 3 days not associated with chills or rigor, loose stool 2 days back 4- 5 episodes (semi-solid to watery) and two episodes of non-bilious vomiting, sensorium started deteriorating after 2 days of onset of symptoms, multiple episode of GTC seizure from morning. Her GCS at the time of admission was 3/15, pupil B/L constricted non-reactive to light, eye ball B/L medially deviated, downturned. Tone decreased, DTR—areflexic. Respiratory rate was 50/min with intercostal retraction, fast and shallow breathing pattern. B/L crepitation heard all over the lung field.Features suggestive of DIC.P/A soft with no hepatospleenomegaly. Temperature at the time of admission was 105.4 F, B.P 80/50 mm Hg in right arm. Her BMI was 12.33Kg/m^2. After admission her first routine examination report are as following table.1.:-

Table.1. Showing laboratory findings

Haematological examination	WBC-21000(N-88%, L-9%, M-3%, E-1%, B-0%) Platelet- 39000/dl Hb- 14.1gm% C-reactive protein- 12 mg/ml S.Urea-72mg/dl S. Creatinine- 1.5mg/dl
Electrolyte	Na+-138meq/l K+-5 meq/l Cl--106meq/l Ca++ -9.3 mg/dl
LFT	SGOT-1820 IU/dl SGPT- 1310 IU/dl ALP- 225 IU/dl PT- 14.7 sec (Control- 12Sec)
ROUTINE URINE	U. Albumin- 1+ Leucocyte/HPF- 1-2 Epithelial cell- A few cell Erythrocyte/HPF- PLENTY
CSF	4 cell/dl, lymphocytic predominant Protein- 42mg/dl Sugar- 66mg/dl ELISA for JE- Negative
Serology	Dengue – Negative Anti Pv/Pf (Parahit Total kit)- Negative

Amino acid chromatography and virus isolation was not done.
Patient was treated with injectable ceftriaxone, meropenem, linezolid, and acyclovir, anticonvulsant (phenytoin, valproate, leveteracetam and midazolam).

During the course of treatment GCS gradually improved over 6 days, she followed simple command but developed aphasia, tone in lower limb gradually increased, bladder control poor.

MRI brain showed symmetrical oval areas of altered signal intensity involving B/L thalami and brain stem tegmental tracts with subacute haemorrhage in thalamic lesion. Few small focal round to oval lesions in B/L corona radiata with evidence of restriction on DWI/ADC.

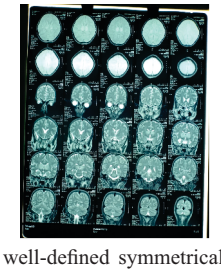


Figure.1. Showing well-defined symmetrical oval areas of altered

signal intensity (appearing hyperintense on T1W1, isointense to slightly hyperintense on T2W1 & not suppressed on FLAIR) noted involving B/L thalami & brain stem tegmental tracts (posterior part of midbrain & pons).

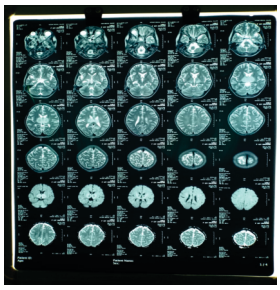
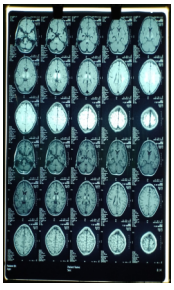


Figure.2. Showing well-defined symmetrical oval areas of altered signal intensity (appearing hyperintense on T1W1, isointense to slightly hyperintense on T2W1 & not suppressed on FLAIR) noted involving B/L thalami & brain stem tegmental tracts (posterior part of midbrain & pons).



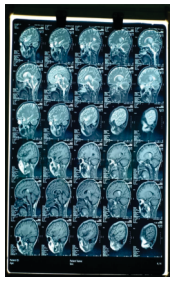


Figure. 3. MRI findings **Figure.4. MRI findings**

Figure 3 and 4: Showing few small focal round to oval lesions (appearing hypointense on T1W1, hyperintense on T2W1 & FLAIR) seen in B/L corona radiata with evidence of restriction on DWI/ADC

Overall MRI findings (figure 1 to 4) were suggested of acute necrotizing encephalopathy of childhood (ANEC).

DISCUSSIONS:

Acute necrotizing encephalopathy of childhood is found mostly in previously healthy young child. It was earlier thought to be found in almost exclusively in previously healthy young children or infants of East Asian including Japan and Taiwan [1]. However,its sporadic cases have been reported from all around the world [2,3]. Our case was a previously healthy child with high fever, loose stool, vomiting and convulsion with fast neurological deterioration. It initiated with sudden onset of fever accompanied by loose stool and vomiting which was followed by sudden deterioration of sensorium and intractable seizure which was controlled by phenytoin, valproate, leveteracetam, and midazolam infusion. Later features of DIC developed for which FFP and fresh whole blood cell transfusion was done. The exact etiology of ANEC has yet to be to determined, but ANEC has been linked to certain respiratory infection such as influenza and genetics have also been described to be playing a role [12]. Qalab et al has reported case of ANEC after Dengue in Pakistan in 2017 [13]. In Skelton and associates' study, a case of ANEC was reported after fever and diarrhea [5]. Salehi Omran MR, also reported case of ANEC after fever and diarrhea in Iran [11]. In our case however virological study except JE and Dengue, was not done. JE and Dengue serological study came to be negative. Before illness the child had normal development as per age with normal intelligence. The child was product of non-consanguineous marriage, no history of seizure disorder in any family member for three generation or history of sudden child death. Urine amino-acid chromatography was not done.

During the course of treatment, consciousness improved gradually in 6 days, starts following simple command in 20 days, recognise relatives but was unable to speak and sit, unable to swallow solid or liquid food. Patient was discharged on nasogastric tube feeding and physiotherapy was advised. During follow-up after 7 days of discharge child was able to follow simple command, bladder bowel become normal, however unable to swallow liquid and solid food, tone of lower limb was increased, knee jerk increased with psychological and dysarthria persisting. Further long term follow-up is needed.

CONCLUSIONS:

Acute Necrotizing Encephalopathy is an under reported case because of its significant intercase variability with unclear etiology.The recognition of ANEC is not easy despite the distinctive clinical and imaging features. Further characterization is difficult in both the mild and severe form of the disease because the appropriate investigations (eg. Mitochondrial enzymes, cytokines, brain and other organ biopsies) are not easily available at the acute stage. It had got poor outcome. Awareness for the detection of underdiagnosed cases is needed to identify possible predisposing factors and improve the outcome of this dangerous acute illness of childhood.

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