ORIGINAL RESEARCH PAPER

General Medicine

A CASE REPORT ON ACUTE NECROTIZING ENCEPHALOPATHY OF CHILDHOOD

KEY WORDS: ANEC, hyperntense lesion, thalamus, brainstem.

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ABSTRACT

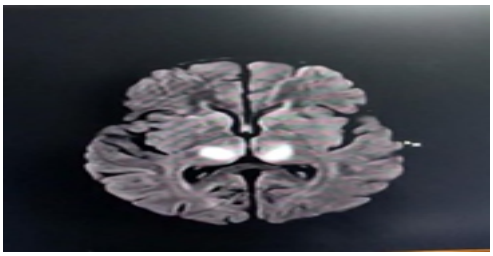
Acute necrotizing encephalopathy of childhood is an atypical encephalopathy seen almost exclusively in previously healthy young children or infants. After viral infection it can be diagnosed by bilateral symmetrical lesions predominantly observed in thalami & brainstem of infants and children. Here we report a case of 13 year old girl child presented with fever, seizure, altered sensorium to our emergency department. MRI brain with contrast showing non enhancing T2W hyperintense lesions in bilateral thalamus, posterior limb of internal capsule with focal areas of hemorrhage suggestive of –Acute Necrotizing Encephalopathy of Childhood.

INTRODUCTION :

Acute necrotizing encephalopathy of childhood is a specific type of encephalopathy reported in young children and infants after viral infection. neuro pathologically viral infections cause foca l breakdown in blood brain barrier characterized by bilateral symmetrical hyperintense lesions predominantly observed in thalamus & brainstem. The real etiology & pathogens of ANEC remains unclear; however mycoplasma, influenza virus, herpes simplex virus, human herpes virus -6 are among the most common infections that intensify the disease. Acute encephalopathy following viral infection with seizure and altered consciousness, and absence of CSF pleocytosis with occasional increased level of protein are characteristic of ANEC.

Case Presentation:

A 13 year old girl child presented with h/o fever of 7 days which is high grade , intermittent ,associated with headache , with h/o seizure one episode ,h/o vomitings -3 episodes , and altered sensorium for 4 days .patient initially got treated in local private hospital ,there diagnosed to have dengue fever (serology shows IgM antibodies against dengue), then patient being referred to SVRRGGH for further evaluation , on initial evaluation patient GCS was –E3V2M5 & hemodynamically stable . on neurological examination patient had hypertonia of all limbs ,biceps & knee reflexes on both sides were 3+, triceps ,supinator, ankle reflexes were 2+. Both pupils were 2mm reacted to light, With b/l withdrawal plantar response & there were no signs o f meningeal irritation . routine blood investigations like CBC, LFT, RFT, SERUM ELECTROLYTES ,urine microscopy were normal, FUNDUS –was normal , in CSF examination TC of 3 cells, protein -40 ,glucose-80 mg/dl, CBNAAT-negative, cultures were normal . she underwent MRI, which showed non enhancing T2W hyperintense lesion in bilateral thalami ,posterior limb of internal capsule with focal areas of hemorrhage. She started on injection acyclovir 400 mg iv tid, inj cefotaxim 1gm iv tid, injection methylprednisolone 20 mg/kg /day for 3 days & injection intravenous immunoglobulin 0.4 mg/kg/day for 5 days . along with anticonvulsant. Patient condition improved dramatically, and her cognition was improved and discharged ambulatory.



MRI BRAIN shows –hyperintense lesion in b/l thalami

DISCUSSION :

Acute necrotizing encephalopathy of childhood is a disease entity seen in previousy healthy children . our case was a previously healthy child presented with fever, headache, seizures, altered sensorium. Differential diagnosis for this case were encephalitis & meningitis but here in this case the CSF analysis was normal. Japanese encephalitis can also involve thalam i bilaterally but the disease is endemic in nature and is not symmetrical as seen in ANEC. Reye syndrome, leigh syndromes are also the differential diagnosis for ANEC , but reye syndrome is associated with hypoglycemia, hyperammonemia, and lactic acidosis. and the leigh syndrome is associated with hypoglycemia, hyperammonemia, & lactic acidosis. clinically ANEC may be differentiated from ADEM by an early onset of encephalitic features just after the prodromal illness while in ADEM , it may take 1 to 2 weeks to develop encephalitis features and there is no symmetrical lesion but patchy demyelination in white matter, gray matter & junction is seen In ADEM .

CONCLUSION :

ANEC is a rare fulminant disease in children ,approximately 1/3 rd of individuals with ANEC do not survive their illness, of those who survive about half will have permanent brain damage. Other individuals who survive their illness appear to recover completely .early intervention with use of intravenous immunoglobulin and iv methylprednisolone may change the outcome.

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