

Acute Necrotizing Encephalopathy: A Rare Case

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Abstract

11 year boy was presented with fever, vomiting and GTC with GCS of 6 and neurological signs with shock. He was given ventilator and circulatory support. MRI was suggestive of Acute disseminated encephalomyelitis [ADEM]. But acute presentation and rapid deterioration, absence of CSF pleocytosis and raised proteins, raised liver enzymes and thalamic involvement on MRI was suggestive of Acute Necrotizing Encephalopathy [ANE] than ADEM. He was treated with IV antibiotics, Acyclovir, Methyl Prednisolone and Intravenous immunoglobulin. In spite of all the vigorous measures, our patient died.

Key words: Acute disseminated encephalomyelitis (ADEM), Acute Necrotizing Encephalopathy (ANE), Children

Introduction

ANE is a rare entity characterized by brain damage that is preceded by an acute febrile disease, mostly viral infections like Influenza A, Herpes Simplex Virus and human herpes virus [1]. ANE is exclusively seen in healthy young children or infants of East Asian including Japan and Taiwan [2]. Most cases are sporadic, though few cases have recurrent and/or familial suggesting of inherited pattern, There is no specific treatment and has poor prognosis with less than 10% of complete recovery [3].

Case Report

A 11-year boy admitted in our hospital with a history of two days fever, moderate grade, not associated with chills and rigor and relieved on taking medication along with five episodes of vomiting on the day of admission after which he was taken to the local physician where he had episode of generalized tonic clonic convulsion for which he was referred to our hospital. During transport he had four episodes of tonic posturing which lasted for about 10-15 seconds. There was no history of cough, cold, diarrhea, joint pains, rash also no history of animal bite, recent travel or any vaccinations. He had received orally Paracetamol and Ondansetron by local physician prior to admission.

On admission, his heart rate was 112 bpm, respiratory rate was 48 cpm, blood pressure 90/60 mm Hg, SpO₂ of 98%, peripheral pulses were feeble and CRT 5 seconds. On neurological examination his Glasgow Coma scale was 6 [E1V2M3]. Pupils were pinpoint not reacting to light. Power was grade 1 in upper and lower extremities. Deep tendon reflexes were brisk and plantar were extensor. Signs of meningeal irritation were absent. Fundus examination revealed left retinal hemorrhages but no papilledema. The child had normal WBC count and platelet count. He had raised SGOT (157 U/L), SGPT (169 U/L), ALP (206 IU/L) levels. His Renal function tests were normal. Serology for Dengue and Japanese Encephalitis were negative. His CSF revealed high proteins (551 mg/dl), normal sugar and normal cell count and CBNAT for Tuberculosis was negative.

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Case Report

On CT scan (Fig1), diffuse white matter hypodensity seen involving extensive areas of bilateral occipito -parietal lobe, bilateral frontal lobes was seen. As child was on ventilator only diffusion weighted and FLAIR sequences MRI were done (Fig 2) which suggested of ill defined hyper intense lesions in bilateral fronto- parietal and occipital temporal predominantly subcortical white matter and cerebellar white matter was seen. Deep grey matter nuclei, basal ganglia-thalami as well as mid brain, pons were also involved suggestive ADEM. There were no findings suggestive of intracranial hemorrhage or space occupying lesion. Based on the clinical symptoms, a laboratory and MRI finding, diagnosis of acute necrotizing encephalopathy was made.

The child was intubated and ventilated on PCV mode with settings FiO₂ 100%, RR 20 PIP 10 and PEEP 5 as his GCS was 6 on admission. Child was empirically started with Injection Ceftriaxone [100mg/kg/day] and Acyclovir [20mg/kg/day]. He was also given 3% NaCl 5ml/kg/dose to reduce intracranial pressure. As the child was hemodynamically unstable, Nor-adrenaline and Dobutamine drip were started. Child was also received injection methyl prednisolone 20 mg/kg/day and IVIg 500mg/kg/day 8 hourly. But child did not respond to our vigorous management and deteriorated further and died after 72 hours of admission.

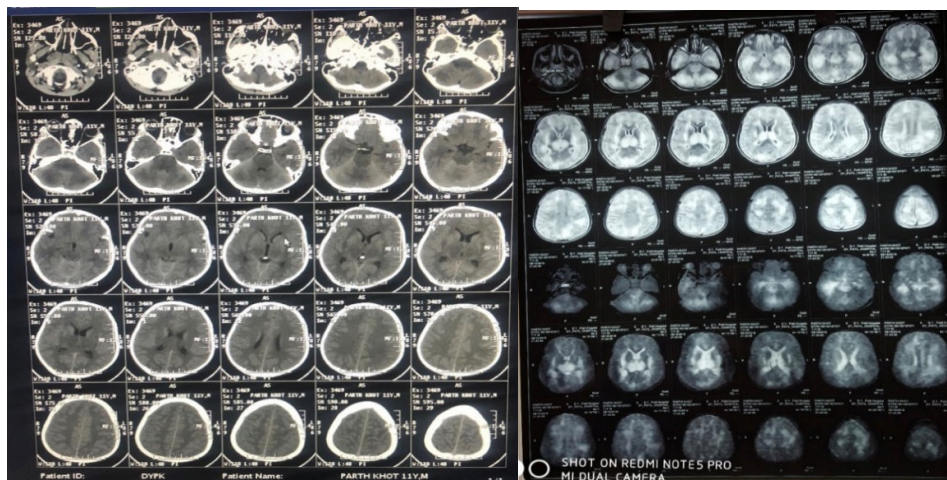


Fig.1

Fig.2

Discussion

ANE is a rare disease and has rapid deterioration. Encephalopathy is followed by respiratory or gastrointestinal infection and high fever [4]. The prodromal symptoms are followed by seizures, altered consciousness that rapidly progresses to coma, neurological deficit and liver problems [5]. ANE is followed mostly by viral infections. Though etiology is unclear it might be followed by Influenza A, Influenza B, Human Herpes virus infections. ANE may be familial or sporadic. Familial cases may be due to RANBP2 gene mutations, and are known as “infection-induced acute encephalopathy 3 (IIAE3) [6,7]. It is believed that virus or its variant increases intracranial cytokine production (TNF alpha, IL 1 and 6), which progresses disease rapidly [8].

The disease is characterized by absence of inflammatory cells in brain parenchyma which differentiates from other entities like ADEM and acute hemorrhagic encephalitis. ANE is diagnosed by multiple, symmetrical lesions showing T2 prolongation in the thalami, frequently with accompanying lesions in

the brain stem tegmentum, periventricular white matter, putamen, and cerebellum [9]. In our patient acute clinical presentation with rapid deterioration, absence of CSF pleocytosis and raised proteins normal sugar, raised liver enzymes and thalamic and brainstem involvement on MRI was suggestive ANE and differentiates from ADEM even if there was predominant white matter affected on MRI [10].

Okumura et al stated that administration of steroids within 24 hours after the onset was related to better outcome of children with ANE without brainstem lesions [11].

Conclusion

ANE is a rare disease which should be differentiated from ADEM with fatal prognosis in spite of vigorous management.

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