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Case Report

A rare case of acute necrotising encephalitis: a case report

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ABSTRACT

Acute necrotising encephalopathy of childhood (ANEC) is a rare brain disorder that occurs most commonly after viral infection, predominantly seen in countries like Japan and Taiwan. The goal of this article is to discuss a case of ANEC and highlight the need for early intervention and treatment, as the prognosis is usually poor. Diagnosis is based on clinical signs and radiological imaging- which shows the involvement of multiple parts of brain matter. There are no standardised guidelines for management, however, most literature supports emphasis on infection control, steroids for neuroprotection, and immunotherapy. Prompt intervention can prevent rapid neurological decline- which is seen in many cases.

Keywords: ANEC, Radiological imaging, Steroids, Immunotherapy

INTRODUCTION

Acute necrotising encephalitis is a rare brain disorder that occurs most commonly after viral infection, predominantly seen in countries like Japan and Taiwan.[1] The exact incidence rate is unknown and very few cases have been reported in literature. Definite diagnosis is usually via imaging (CT or MRI) which shows lesions affecting the thalamus, brainstem, periventricular white matter, and cerebellum.[2] Here we discuss a case and highlight the management of a severe case of acute necrotising encephalitis, emphasising the need for early intervention to prevent morbidity and mortality in these patients.

CASE REPORT

A 1 year 2-month-old developmentally normal male child, immunised up to date, was brought with complaints of fever with seizures and loose stools since 1 day. The child had 2 episodes of focal tonic seizures of upper limbs lasting for 10 seconds at presentation in triage. On examination, the child had tachycardia along with CFT prolongation and hypotension. The child had mild respiratory distress as well. He was drowsy with a GCS of 6/15 and depressed anterior fontanelle. CNS examination revealed miotic pupils which were 2 mm in size. The tone was increased in all 4 limbs with brisk deep tendon reflexes and bilateral plantar response was extensor in nature.

The child's clinical status improved with 2 Ringer lactate boluses. His seizures were controlled with levetiracetam (maintenance dose of 20 mg/kg/day). His investigations also showed hypokalemia with hypoglycemia. The child was started on severe dehydration correction, with KCL, and intravenous antibiotics (inj. cefera-T and inj. amikacin).

Potassium levels normalised on day 2 of admission. No further episodes of hypoglycemia were noted. The child had 2 further of episodes of seizures on day 2 of admission- general tonic-clonic type lasting for about 2 mins. These seizures were aborted with inj. midazolam and inj. levera (maintenance dose of 30 mg/kg/day). However, his sensorium progressively worsened.

An EEG was performed which was suggestive of generalized electrical disturbance- an indicator of severe encephalopathy. CSF analysis showed elevated protein with 50 RBCs after an atraumatic tap. A diagnosis of viral encephalitis was considered, so he was started on 2/3rd maintenance fluids and inj. acyclovir. Subsequently, however, viral and autoimmune encephalitis CSF PCR panels were negative.

The child was started on 1/3rd feeds on day 3 of admission and Ryle's tube feeds were slowly upgraded. MRI with contrast showed features of ANE.

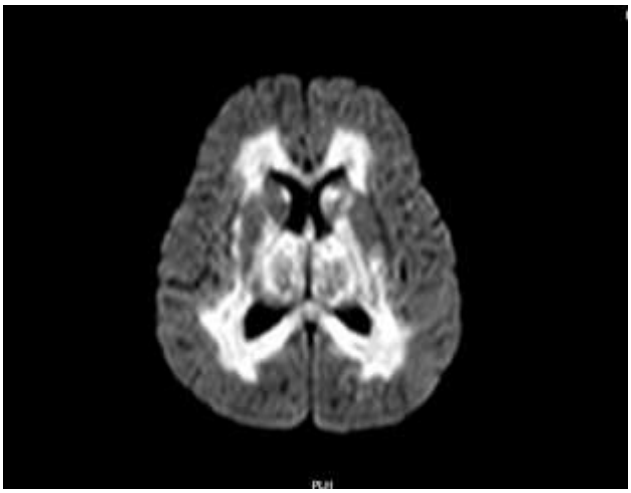

Figure 1: MRI showing bilateral thalamus and periventricular necrosis- highly suggestive of ANEC.

The child was then given 1 gm/kg IV IG, inj. dexamethasone (later converted to oral) and IV antibiotics were continued inj. The child clinically improved with the above measures and was discharged on day 15 with a GCS of 11 and stable vitals.

DISCUSSION

ANE is an extremely rare disease and the exact incidence rate is unknown.

Encephalopathy can be suspected when a developmentally normal child presents acutely with psychomotor changes and regression.[1] Most commonly, symptoms at initial presentation are seizures along with altered mental status.[3] This was consistent with how our patient presented. It is believed that TNF alpha and IL 1 and 6 are involved in the rapid decline of neurological status.[4]

Clinically, ANE can resemble other conditions such as Reye syndrome, Leigh encephalopathy, and Wernicke encephalopathy.[2] CSF findings are increased protein with or without pleocytosis.[1] Our patient's CSF analysis showed elevated protein and RBCs- which is similar to that of HSV encephalitis.

Diagnosis of ANEC is radiological (CT/MRI) showing lesions affecting the thalamus, brainstem, periventricular white matter, and cerebellum. There is involvement of gray and white matter, along with evidence of the breakdown of the blood-brain barrier.[2]

Management is often focused on infection control, steroids for neuroprotection, and immunotherapy, along with symptomatic management.[5,6] Our child showed an initial rapid neurological decline and then gradual improvement with 1 gm/kg IV IG, inj. dexamethasone, and continuation of IV antibiotics.

Acyclovir is the drug of choice to combat viral etiology.[7] Early initiation of immunotherapy leads to improved outcomes in pediatric patients. Studies show good outcomes with IL-6 blockers as well.[5] There is no current data showing the superiority of methylprednisolone over dexamethasone is available.

Very few patients recover completely, and the prognosis is usually poor which may be determined by the amount of brain involvement on imaging. Many patients have long-lasting neurological sequelae and subsequent developmental abnormalities.[8] The role of multi-disciplinary therapy cannot be emphasised on further as regular physiotherapy, feeds, and various medical teams are required to manage such patients to ensure their journey towards recovery.

CONCLUSION

ANEC is a rapidly progressing, severely debilitating disorder, with a high mortality rate. A high index of suspicion and early initiation of treatment may prevent morbidity and mortality. As this disease is a diagnostic and therapeutic challenge, hence, it is important to discuss several such cases to pinpoint the presentation, causes in different regions of the world, and management algorithms.

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