



Acute necrotizing encephalopathy of childhood: a Case Series from a tertiary care center of Pakistan

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Background

Acute Necrotizing Encephalopathy of Childhood (ANEC) is a severe form of encephalopathy, predominantly affecting Asian children aged 6 to 18 months (1). ANEC often follows viral infections such as influenza, HHV-6, or enterovirus, including potential associations with SARS-CoV-2 during pandemics (2). Clinical manifestations include fever, altered consciousness, seizures, and characteristic brain imaging findings, notably bilateral thalamic lesions (3).

Despite its severity, ANEC is frequently misdiagnosed or unrecognized, leading to delayed treatment and poor outcomes. While a unified treatment protocol is lacking, steroids and IVIG are commonly used for immunomodulation therapy (4). This study illuminates the diverse presentations and challenges of ANEC, particularly in the context of SARS-CoV-2, underscoring the importance of early diagnosis and tailored management strategies.

Case Report

Table 1	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age	2 years	3 years	6 years	2.5 years	9 months	2 years 9 months
Presenting Symptoms	Fever, Vomiting, Irritability	Fever, Vomiting, Cough, Irritability	Fever, Vomiting, Irritability	Stiff body, Stiffness, Irritability	Fever, Irritability	Fever, Irritability
GCS Score	12/15		7/15	9/15	8/15	6/15
Preceding Infections	SARS-CoV-2, E. coli, Urinary tract infection	RSV, SARS-CoV-2, and Dengue virus	Aspergillus flavus, influenza B	Not isolated	Dengue virus	SARS-CoV-2
Neurological Symptoms	Seizures, Altered consciousness	Seizures, Altered consciousness	Seizures, Drowsy, Decorticate posturing	Stiffness, Altered consciousness	Seizures, Altered consciousness	Seizures, Altered consciousness, Hypertonia
Radiological Findings	B. thalami, centrum semi vale and periventricular white matter	B. thalami, Subcortical white matter	B. thalami centrum semi vale, B temporal lobes, B. cerebellar hemispheres	B. thalamic lesions	B. thalami, cerebral, cerebellar white matter (CT brain)	B. thalami, B. cortical and subcortical white matter in frontal, parietal and occipital lobes.
Treatment	Oseltamivir, Methylpred, IVIG, Antibiotics	IVIG, Methylpred, Antiepileptics, IV antibiotics	Methylpred IVIG, Antiepileptics, IV antibiotics	Methylpred, IVIG, Antiepileptics, Antibiotics	ostelmavir, IVIG, Methylpred Antiepileptics, Antibiotics	IVIG, Methylpred Antiepileptics, Antibiotics
Outcome	Discharged home, Follow/up advised	Discharged home, Follow/up advised	Discharged home, Follow/up advised	Discharged home, Follow/up advised	Expired	Discharged home, Follow/up advised

Abbreviations: B. Bilateral; IVIG Intravenous immunoglobulin

Method

A retrospective chart review of six pediatric patients diagnosed with ANEC at the Aga Khan University Hospital in Karachi were enrolled. The study analyzed clinical, radiological findings, underlying etiologies, treatment approaches, and its outcomes.

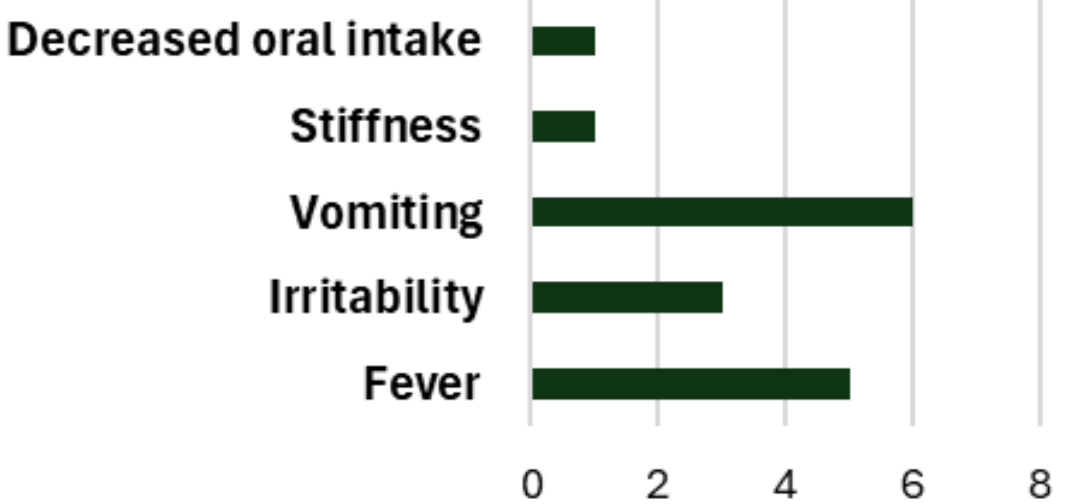
Results

The cases in this series exhibited varying clinical presentations, including fever, seizures, and altered consciousness. Neuroimaging consistently revealed bilateral thalamic lesions, a hallmark of ANEC, which were seen in 83% of the patients and 33% of the patients showed white matter involvement. All patients received both intravenous immunoglobulin (IVIG) and Steroids along with different combinations of therapies such as antibiotics, antiviral medications, and supportive care. As for the outcomes, 4 patients (67%) successfully recovered and discharged home while two patient died with mortality rate (33%) was determined.

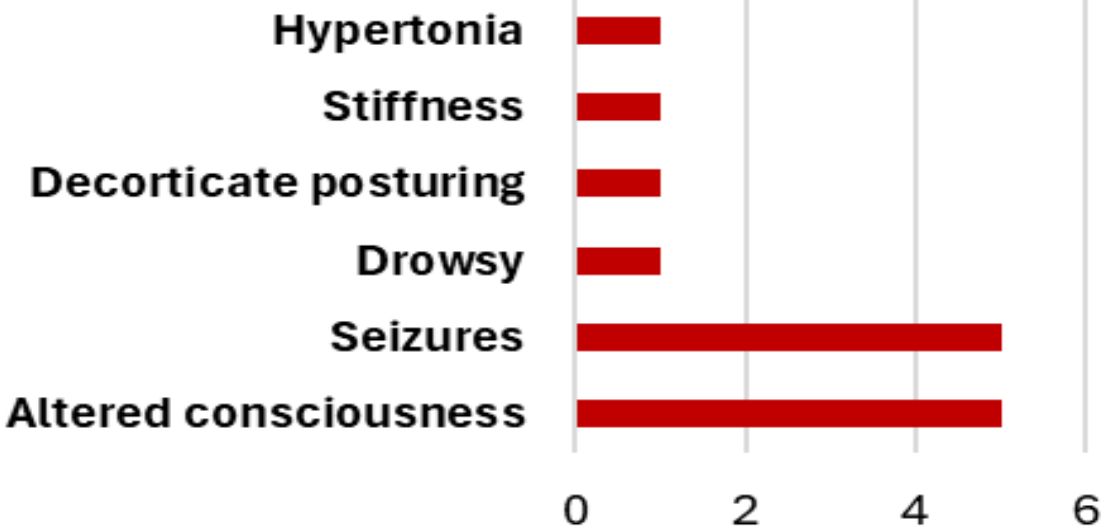
Conclusions

In conclusion, this case series sheds light on ANEC's clinical spectrum, diagnostic challenges, treatment strategies, and outcomes. It stresses the importance of early recognition, accurate diagnosis, and tailored treatment to improve outcomes. Cases associated with SARS-CoV-2 highlight the need for ongoing research into this rare disorder and its viral connections, making this study essential for healthcare providers managing ANEC cases.

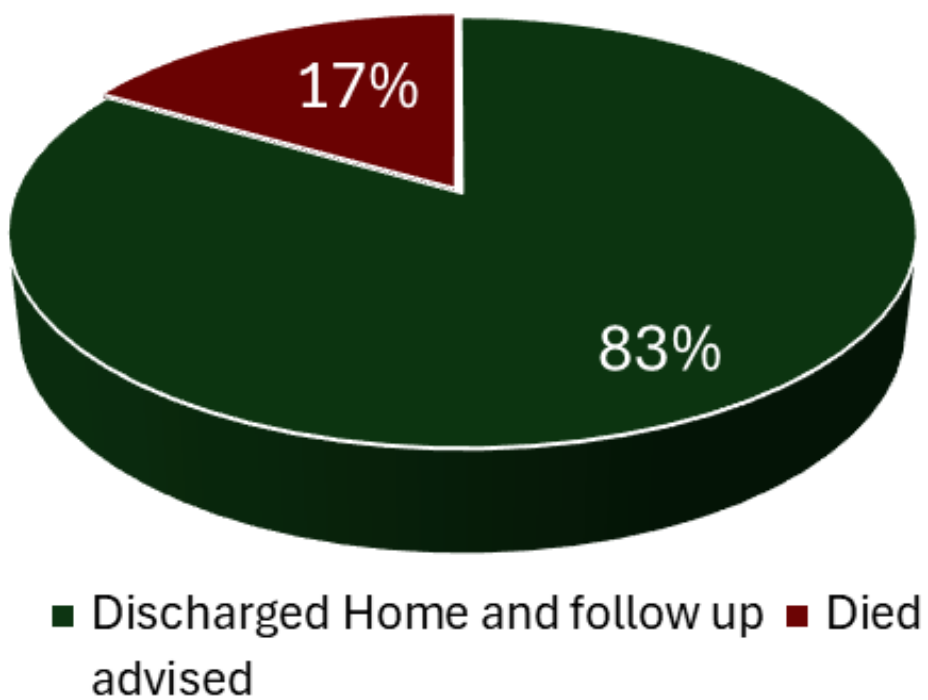
Systemic symptoms



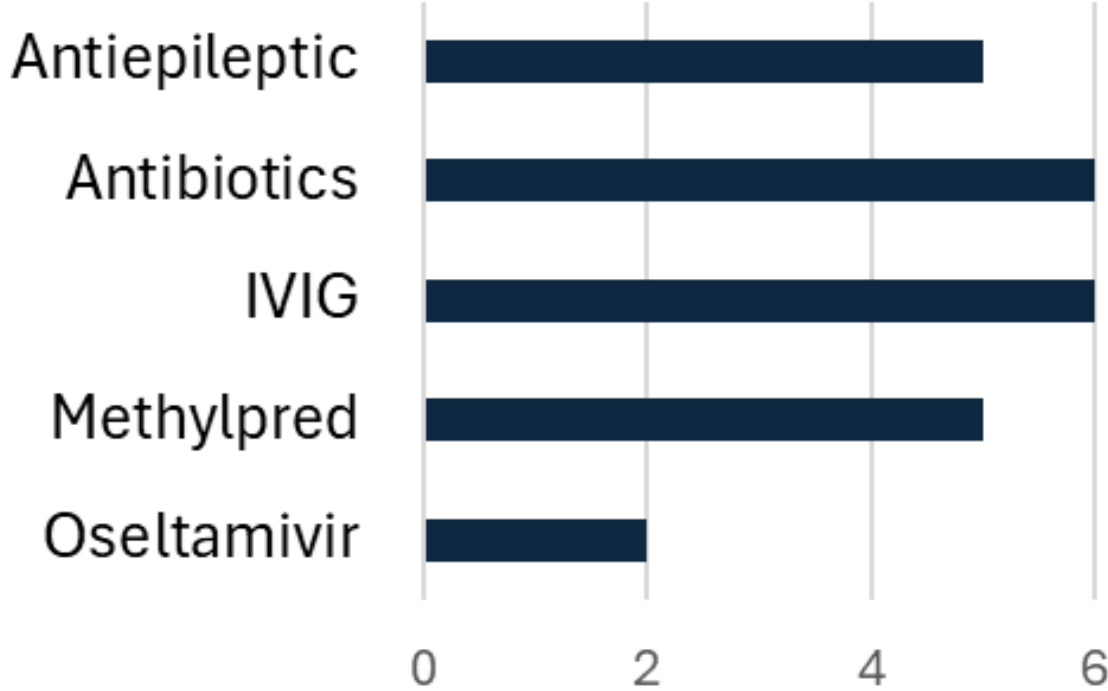
Neurological symptoms



Treatment Outcome



Frequency of Medication use



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