

Acute Encephalopathy in a Healthy 6-Year-Old

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Acute Necrotizing Encephalopathy of Childhood (ANEC) is a rare complication of post-infectious encephalopathy.

We report a six-year-old girl who presented to the emergency department with history of headache, seizure and rapid deterioration in the consciousness level, preceded by a history of upper respiratory viral infection. The neuroradiological images revealed extensive thalamic lesions bilaterally, suggestive of ANEC. The patient was treated with IV pulse steroid therapy, and the outcome was full recovery of the clinical condition.

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Acute Necrotizing Encephalopathy of Childhood is a rare disease that affects children and is usually preceded by viral infection^{1,2}. It typically presents with rapid progressive encephalopathy associated with convulsions after febrile illness².

The mainstay of diagnosing ANEC is the clinical presentation and the radiological finding of bilateral symmetrical lesions involving mainly the thalami³. ANEC is associated with poor prognosis and has a high rate of morbidity and mortality if not recognized and treated early⁴.

The aim of this presentation is to report a case of Acute Necrotizing Encephalopathy of Childhood and its relevant management.

THE CASE

A six-year-old girl who was previously healthy presented to the emergency room with two-days history of reduced level of consciousness. The family reported one episode of generalized tonic-clonic convulsion associated with eye up-rolling, frothy secretions from the mouth and urinary incontinence lasting few minutes; the attack was terminated after 10 mg Diazepam injection. The patient had upper respiratory tract infection (URTI) viral illness one week before her clinical presentation.

Physical examination showed normal vital signs with no fever. The patient was encephalopathic with Glasgow Coma Scale (GCS) of 10/15, in addition to brisk reflexes and Babinski's sign, characterized by an upgoing big toe and fanning outward of the other toes bilaterally. Urgent brain MRI was performed, which revealed bilateral mirror image enhancement of the thalami, see figures 1 and 2.

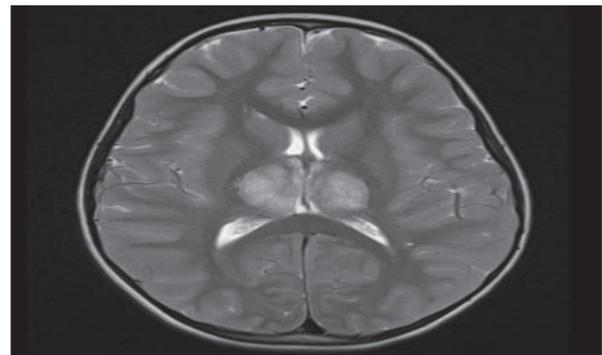


Figure 1: MRI of Brain Axial View Showing Bilateral Mirror Image Enhancement of the Thalami

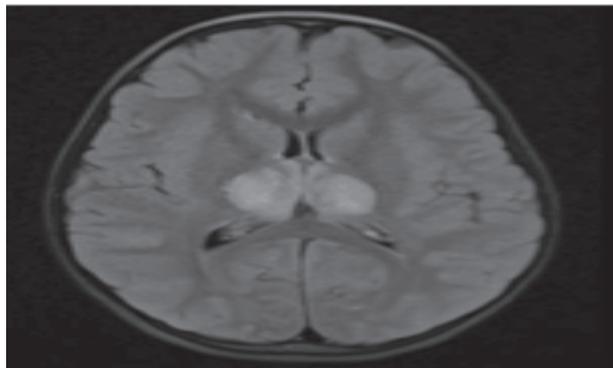


Figure 2: FLAIR sequence MRI of the Brain Showing Bilateral Mirror Image Enhancement of the Thalami

The patient showed rapid deterioration of consciousness and was transferred to ICU. IV Pulse steroid (Methylprednisolone) 600 mg was initiated, which was followed by dramatic improvement of clinical condition. She remained vitally stable

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with no further seizure attacks. Repeat brain MRI after steroid therapy showed a significant decrease in the size of thalamic lesions bilaterally, see figure 3. The patient was discharged after full recovery and no residual neurological deficits.

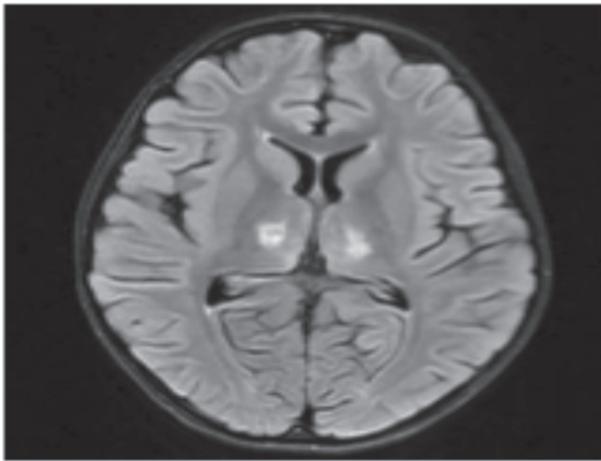


Figure 3: FLAIR sequence MRI of the Brain Axial View Showing Significant Decrease in the Size of the Thalamic Lesions after Steroid Therapy

DISCUSSION

Acute Necrotizing Encephalopathy is rare in pediatrics. Wang et al reported that ANEC is rare symmetrical involvement of bilateral thalami and other brain regions in previously healthy children¹. Our patient was the first case to be reported from our hospital, which is similar to what was reported by Wang et al¹.

The typical clinical presentations of ANEC include acute encephalopathy with seizures and decreased level of consciousness after viral illness². Our patient presented with rapid deterioration in the level of consciousness associated with convulsion preceded by viral infection.

Neurological images include CT brain and MRI, which are the diagnostic tools for ANEC. Our patient had brain MRI, which showed symmetrical bilateral involvement of both thalami and multiple lesions of the cerebral white matter which was highly suggestive of ANEC^{3,5}.

The prognosis of ANEC is usually poor and associated with high mortality rate. Manara et al reported that the outcome of ANEC is usually poor and approximately 70% of the patients die within few days from the onset of fever⁴.

Early identification and treatment with steroid were associated with good prognosis, especially in the absence of brainstem lesions. Okumara et al reported that early steroid treatment within 24 hours after the onset of symptoms is associated with better outcome⁶.

Our patient was treated within few hours after the onset of symptoms with IV Pulse steroid (Methylprednisolone) 600 mg, which resulted in full recovery and no residual neurological deficits; this was similar to the outcome of early and intensive use of IV corticosteroids reported by Araujo et al⁷.

CONCLUSION

Acute Necrotizing Encephalopathy of Childhood is a rare form of encephalopathy that occurs in previously healthy children with characteristic neurological image findings.

Early identification and treatment with steroid were associated with better prognosis and reduction in morbidity and mortality rate.

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REFERENCES

1. Wang HS, Huang SC. Acute Necrotizing Encephalopathy of Childhood. *Chang Gung Med J* 2001; 24(1):1-10.
2. Campistol J, Gassió R, Pineda M, et al. Acute Necrotizing Encephalopathy of Childhood (Infantile Bilateral Thalamic Necrosis): Two Non-Japanese Cases. *Dev Med Child Neurol* 1998; 40(11):771-4.
3. Hassanzadeh Rad A, Aminzadeh V. Acute Necrotizing Encephalopathy of Childhood (ANEC): A Case Report. *Iran J Child Neurol* 2017; 11(1):75-77.
4. Manara R, Franzoi M, Cogo P, et al. Acute Necrotizing Encephalopathy: Combined Therapy and Favorable Outcome in a New Case. *Childs Nerv Syst* 2006; 22(10):1231-6.
5. Albayram S, Bilgi Z, Selcuk H, et al. Diffusion-Weighted MR Imaging Findings of Acute Necrotizing Encephalopathy. *AJNR Am J Neuroradiol* 2004; 25(5):792-7.
6. Okumura A, Mizuguchi M, Kidokoro H, et al. Outcome of Acute Necrotizing Encephalopathy in Relation to Treatment with Corticosteroids and Gammaglobulin. *Brain Dev* 2009; 31(3):221-7.
7. Araújo R, Gouveia P, Fineza I, et al. Bilateral Thalamic Lesions in Acute Necrotizing Encephalopathy Due to H1N1 Infection. *Pediatr Neurol* 2016; 65:96-97.