Acute Necrotizing Encephalopathy: The Importance of Imaging Features

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A previously healthy 2-year-old male toddler presented with high fever, cough, upper limb tremor and gait ataxia, rapidly evolving to neurologic deterioration, drowsiness, and bradycardia and, therefore, requiring intensive care. Complete blood cell count, C reactive protein, aminotransferases, ammonia, and lactate were normal and the toxicology screen was negative. Nasopharyngeal swab polymerase chain reaction (PCR) was positive for rhinovirus and negative for influenza and other respiratory viruses. Lumbar puncture revealed pleocytosis of 24 cells/µL with predominant mononuclear cells, normal glucose, and mildly elevated protein levels (61.7 mg/dL), with sterile cerebrospinal fluid cultures and negative PCR for herpes virus and enterovirus. Other infectious agents were also excluded. There was a mild blood-brain barrier disturbance, with no oligoclonal bands or intrathecal synthesis of immunoglobulins. The electroencephalogram showed an encephalopathic pattern with irregular and slow background activity (Fig. 1) and brain magnetic resonance imaging (MRI) showed multifocal bilateral thalamic lesions (Fig. 2).

Acute necrotizing encephalopathy was admitted, and methylprednisolone 30 mg/kg/day initiated. Due to persistent altered mental status, associated with aphasia and dysphagia, and as biotin-thiamine-responsive basal ganglia disease could not be excluded, these vitamins were also initiated. Clinical improvement was noticed after five days, with full recovery of swallowing and speech. Eight months after the diagnosis, he showed lower right-hand dexterity and left-hand dominance. The metabolic evaluation revealed normal amino acids and redox potential and genetic study of biotin is ongoing, even though there is no family history of acute necrotizing encephalopathy.

Acute necrotizing encephalopathy is a severe neurological disorder characterized by rapid neurologic

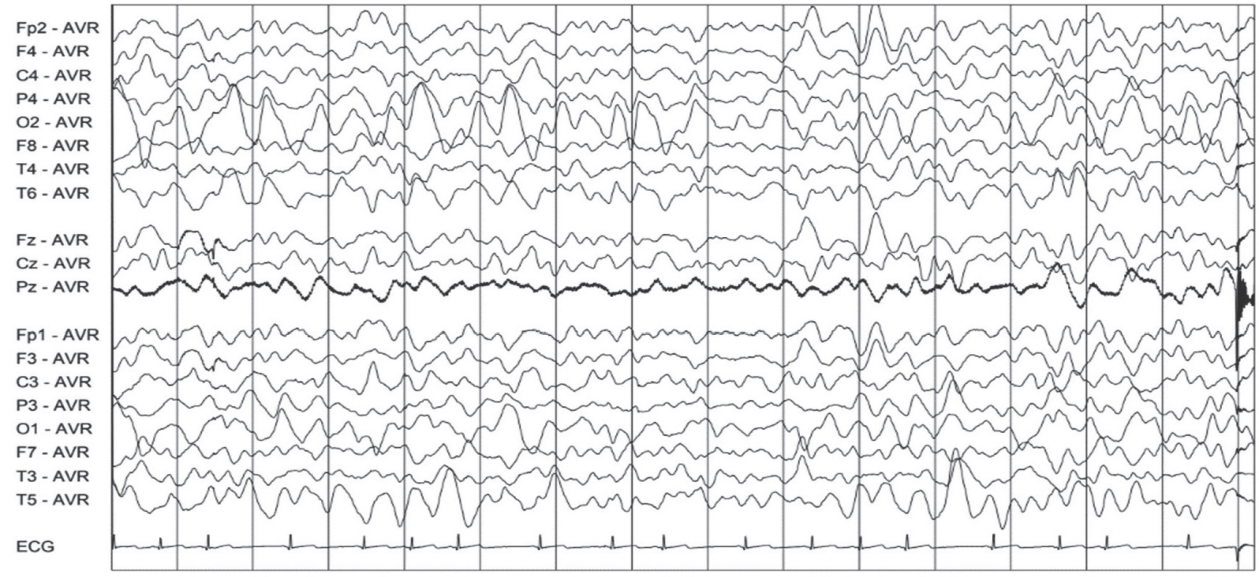

Figure 1. Diffuse and moderately slow and Diffuse and moderately slow and irregular encephalopathic pattern, moderately reactive to stimulation. No asymmetries. No focal anomalies or paroxysmal activityirregular encephalopathic pattern, moderately reactive to stimulation. No asymmetries. No focal anomalies or paroxysmal activity.

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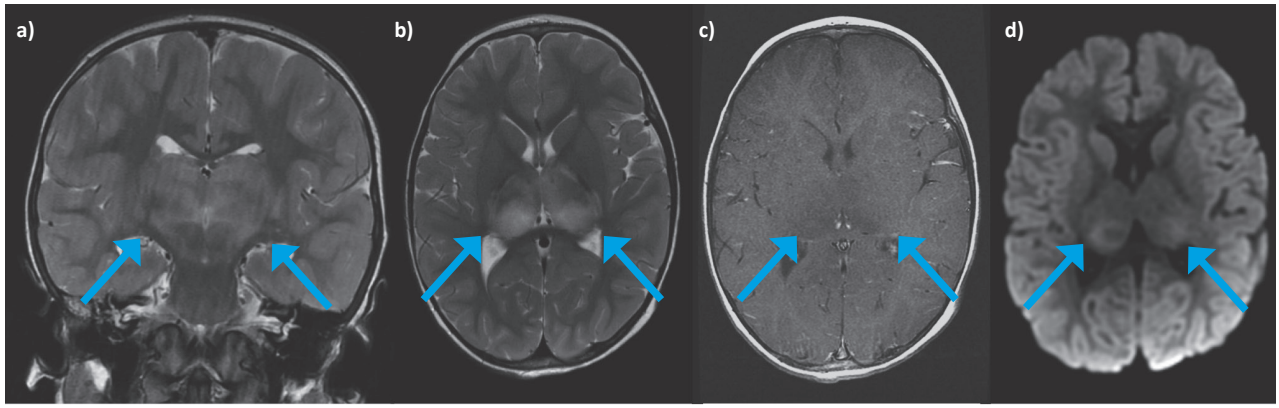

Figure 2. Coronal T2 (a), axial T2 (b), axial T1 with gadolinium (c) and axial DWI (d): Bithalamic lesions with extension to the midbrain and internal capsule, symmetrical, hyperintense in T2, hypointense in T1, without gadolinium enhancement and with peripheral restricted diffusion. No haemorrhage or cavitation.

deterioration secondary to a virus febrile illness. It is imagiological diagnosed by a distinctive feature of multifocal symmetric brain lesions, affecting the bilateral thalamus, brainstem, cerebral periventricular white matter, and cerebellum.[1] Common etiologic agents include influenza virus and other infectious diseases as well as toxic, metabolic, and inflammatory/vascular central nervous system disorders.[2] In this case, we question the pathogenicity of rhinovirus associated with acute necrotizing encephalopathy, as it has not been described before. The genetic result is also important, even without a positive familial history of acute necrotizing encephalopathy, as if positive for the Ran binding protein 2, it may predict new episodes and early management can improve the frequently poor prognosis.[3,4] In children without brainstem lesions, improvement in the outcome was described particularly when steroids were started within the first 24 hours from the onset.[5]

Keywords: Child, Preschool; Leukoencephalitis, Acute Hemorrhagic/diagnosis

WHAT THIS REPORT ADDS

- This case report aims to raise awareness for the importance of magnetic resonance distinctive features that should raise the suspicion of underlying acute necrotizing encephalopathy.
- When acute necrotizing encephalopathy is diagnosed the early start of steroids can improve the diagnosis.

Conflicts of Interest

The authors declare that there were no conflicts of interest in conducting this work.

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Consent for publication was obtained.

Confidentiality of data

The authors declare that they have followed the protocols of their work centre on the publication of patient data.

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