Case Report

A Case of Deep Cerebral Venous Thrombosis Presenting like Acute Necrotizing Encephalopathy

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Abstract

Acute necrotizing encephalopathy (ANEC) is a rapidly progressive neurologic disorder that occurs in children after common viral infections such as influenza A and herpes simplex virus. It is observed more commonly in Asian countries. Magnetic resonance imaging findings in ANEC include symmetrical brain lesions, preferentially affecting the thalamus bilaterally. However, similar neuroimaging findings are also observed in deep cerebral vein thrombosis, which can lead to misdiagnosis. We report a case of 2½-year-old child who presented like acute necrotizing encephalopathy but on further investigations was found to be having deep cerebral vein thrombosis.

Keywords: Acute necrotizing encephalopathy, deep cerebral vein thrombosis, magnetic resonance imaging, thalamus

Introduction

Acute necrotizing encephalopathy (ANEC) is a rare, severe encephalopathy observed more commonly in Asian countries.[1] It is thought to be triggered by a viral infection (influenza and human herpes virus-6 [HHV-6]) in a genetically susceptible host.[2] Magnetic resonance imaging (MRI) findings are characterized by symmetrical brain lesions, preferentially affecting the thalamus bilaterally, which help to make a prompt diagnosis of acute necrotizing encephalopathy.[3] However, neuroimaging findings in such cases should be interpreted very cautiously because venous sinus thrombosis of deep cerebral veins can produce similar neuroimaging findings characterized by infarction and vasogenic edema of bilateral thalami.

Case Presentation

We report a case of 2½-year-old girl who presented with complaints of irritability, which lasted for 2 days followed by altered sensorium for 1 day. Two weeks before this presentation, the patient was admitted for bronchopneumonia and was discharged after 7 days of intravenous (IV) antibiotics. Seven days after being discharged, she developed excessive irritability with decreased intake of oral feeds that lasted for 2 days, followed by altered sensorium. She was developmentally normal with no history of any other major illness in past. On admission, the patient’s Glasgow coma scale was E2V1M5 with blood pressure 116/68 mm Hg, heart rate 120/min, and respiratory rate 26/min. Her cranial nerves were intact. She had no weakness, ataxia, sensory disturbance, or meningeal signs. Motor examination revealed increased tone of both upper limb and lower limb, power <3/5 with brisk deep tendon reflexes, and extensor plantar response. She developed seizures on admission, and was started on IV phenytoin, valproate, and levetiracetam.

MRI of brain revealed symmetrical large areas of restricted diffusion appearing hyperintense on T2-weighted (T2W)/fluid attenuation inversion recovery image and hypointense on T1-weighted (T1W) images involving bilateral thalami, suggestive of acute necrotizing encephalitis [Figure 1].

On the basis of clinical presentation and neuroimaging findings, a tentative diagnosis of acute necrotizing encephalopathy was made. The patient was started on anticoagulation, and repeat MRI showed improvement in the neuroimaging findings, indicating resolution of deep cerebral vein thrombosis.

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encephalitis was made. The child was started on IV methylprednisolone pulse therapy (30 mg/kg OD). However, she did not respond to the treatment, magnetic resonance venography was planned, which revealed deep cerebral venous thrombosis involving right transverse and sigmoid sinus, straight sinus, bilateral internal cerebral veins, and vein of Galen [Figure 2].

As the diagnosis of deep cerebral venous sinus thrombosis was made, methylprednisolone was stopped and low-molecular-weight heparin was started. The patient’s prothrombotic workup was sent. Her condition improved gradually, and oral feed was started. She was discharged after 1 month with a plan of repeat magnetic resonance venogram after 3 months on follow-up.

**DISCUSSION**

The real etiology and the pathogenesis of ANEC remain unclear. Usually, it develops secondary to viral infections, including influenza A, herpes simplex virus, HHV-6, and enterovirus. Pathologically, there is edema, hemorrhage, and necrosis in thalamic region.

In most of the patients of ANEC, bilateral symmetrical thalamic involvement is observed. MRI of brain shows multiple areas of restricted diffusion involving bilateral thalami.

*Figure 1:* MRI of brain showing large areas of restricted diffusion appearing in T1W hypointense (A) and T2W hyperintense (B) images involving bilateral thalami, neuroimaging findings were consistent with acute necrotizing encephalitis. Patchy areas of blooming (C) on susceptibility weighted images are seen in the region of bilateral thalami
thalam. Lesions are hypointense on T1W and hyperintense on T2W images. These findings can be extensive. Multiple areas of hemorrhagic spots, cavitations, and post-contrast enhancement are also observed. Deep cerebral venous thrombosis also causes vasogenic edema, hemorrhage, and necrosis in the bilateral thalam, which results in similar neuroimaging findings.

CONCLUSION
This case report highlights that in the presence of bilateral thalami lesions, deep cerebral venous thrombosis must be considered in addition to ANEC. Delay in the diagnosis of cerebral venous thrombosis and commencement of anticoagulant therapy can lead to unfavorable outcomes.

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Conflicts of interest
There are no conflicts of interest.

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