Teaching NeuroImages: Acute necrotizing encephalopathy of childhood

Neuroimaging findings

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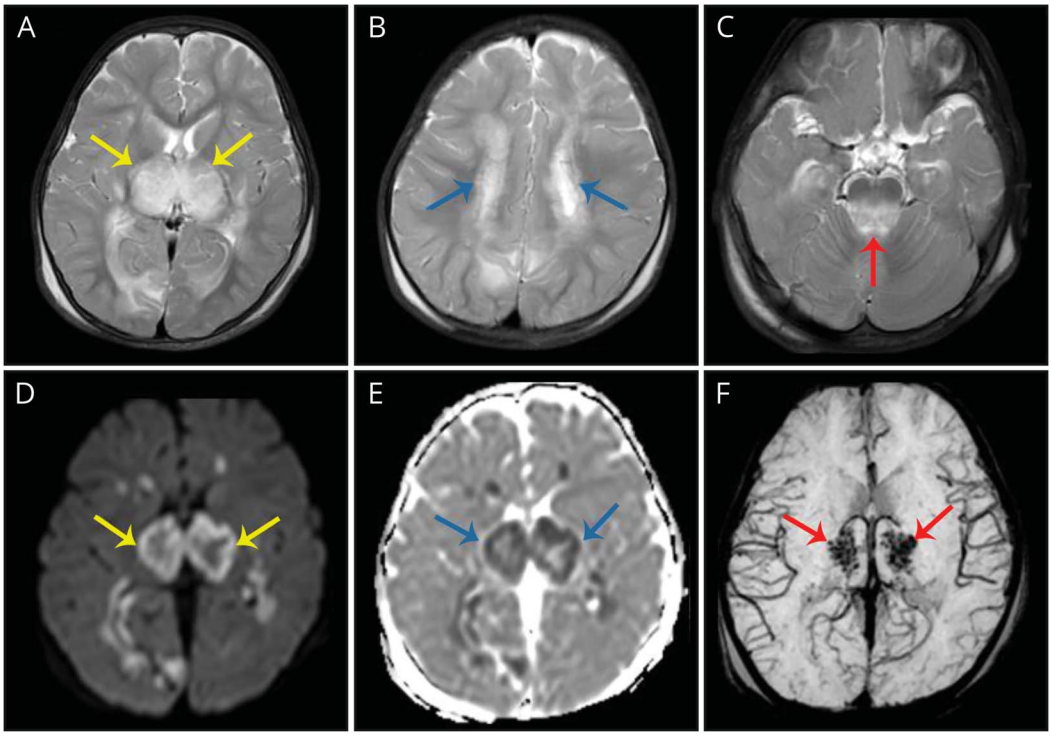
Figure 1 MRI shows symmetric thalamic swelling, brainstem and white matter involvement, and trilaminar appearance on apparent diffusion coefficient (ADC)



Axial T2-weighted images (A–C) show swelling and hyperintensity involving the thalami (A), cerebral white matter (B), and dorsal brainstem (C). The thalamic lesions show a trilaminar appearance on the diffusion-weighted imaging and ADC images (D, E). Punctate hemorrhagic foci are noted on the susceptibility-weighted imaging images (F).

A 10-month-old infant was brought to the hospital in status epilepticus, preceded by a 2-day history of fever and loose stools. Brain MRI revealed swelling and T2 hyperintensity involving the thalami, white matter, and dorsal brainstem (figure 1). The thalamic lesions showed a trilaminar appearance on the diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) images, with hemorrhagic foci on susceptibility-weighted imaging (figures 1 and 2).

Acute necrotizing encephalopathy of childhood is a fulminant encephalopathy affecting infants and children.[1] Viral infections (influenza, rotavirus, human herpesvirus–6), immune-mediated, and genetic (*RANBP2* mutation) etiologies have been implicated as causative factors.[2,3] The trilaminar appearance on DWI and ADC images and symmetric thalamic involvement is characteristic.

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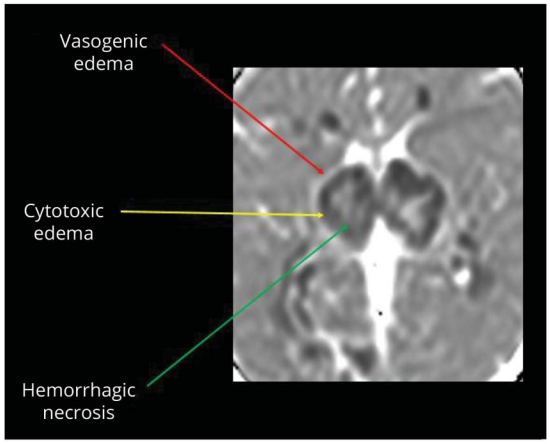
Figure 2 Axial apparent diffusion coefficient (ADC) image shows the characteristic trilaminar appearance



Axial ADC image shows the trilaminar appearance in better detail. The inner region of facilitated diffusion is due to hemorrhagic necrosis, middle layer of restricted diffusion is due to cytotoxic edema, and outer layer of facilitated diffusion is due to vasogenic edema.

Author contributions

Asthik Biswas: concept, image interpretation, and writeup of manuscript. Mugil Varman: image interpretation and preparation. Aditya Gunturi: image interpretation and preparation. Sangeetha Yoganathan: clinical input, critical revision for intellectual content. Sridhar Gibikote: critical revision for intellectual content.

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Disclosure

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References

1. Mizuguchi M, Abe J, Mikkaichi K, et al. Acute necrotising encephalopathy of childhood: a new syndrome presenting with multifocal, symmetric brain lesions. J Neurol Neurosurg Psychiatry 1995;58:555–561.
2. Mizuguchi M. Acute necrotizing encephalopathy of childhood: a novel form of acute encephalopathy prevalent in Japan and Taiwan. Brain Dev 1997;19:81–92.
3. Neilson DE, Adams MD, Orr CMD, et al. Infection-triggered familial or recurrent cases of acute necrotizing encephalopathy caused by mutations in a component of the nuclear pore, RANBP2. Am J Hum Genet 2009;84:44–51.