

Teaching NeuroImage: Occipital Cortical Enhancement During Encephalopathy-like Episode in Neuronal Intranuclear Inclusion Disease

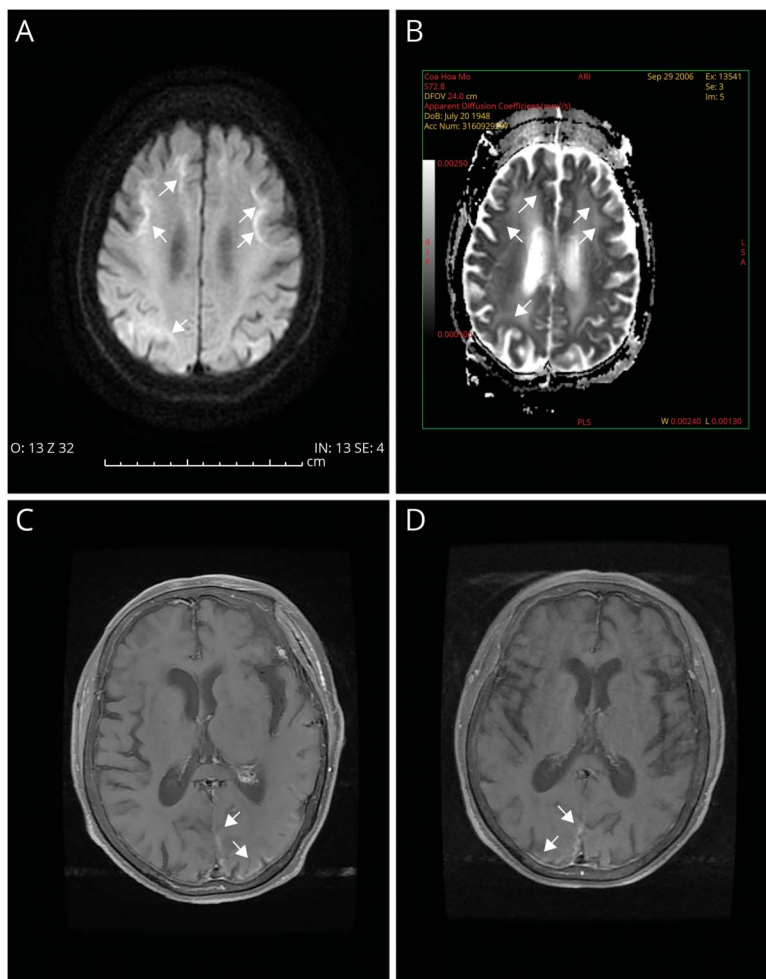
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Figure 1 Brain MRI Findings of an NIID Patient During Encephalopathy-like Episode



(A, arrow) Diffusion-weighted images showing high signals along the corticomedullary junction; the corresponding ADC images also show similar findings (B, arrow). (C, arrow) Postcontrast MRI showing enhanced lesions distributed in the left occipital lobe cortex and no enhancement in the right occipital lobe cortex. (D, arrow) Postcontrast MRI showing enhanced lesions distributed in the right occipital lobe cortex and no enhancement in the left occipital lobe cortex.

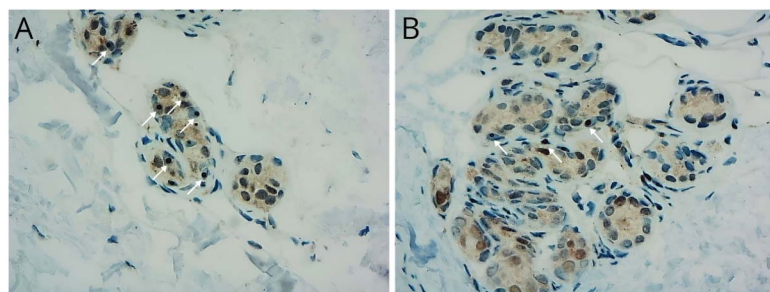
A 66-year-old woman developed behavioral abnormalities and altered sensorium and showed high signals along the corticomedullary junction on diffusion-weighted image (Figure 1A) and the corresponding ADC images (Figure 1B). Postcontrast magnetic resonance imaging showed enhancement of the left occipital lobe cortex (Figure 1C). One year later, she had recurrence of the same

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J.B Wu and Q.L Li contributed equally to this work.

Figure 2 Skin Biopsy Findings



(A, Arrow) Immunostaining showed p62-positive intranuclear inclusions in sweat gland cells ($\times 400$). (B, Arrow) Immunostaining showed ubiquitin-positive intranuclear inclusions in sweat gland cells ($\times 400$).

symptoms and her MRI showed enhancement of the right occipital lobe cortex (Figure 1D). These imaging findings correlated with her reversible encephalopathy episodes, manifested by behavioral abnormalities and altered sensorium. Finally, a diagnosis of neuronal intranuclear inclusion disease (NIID) was established based on skin biopsy (Figure 2A, B) and genetic testing.

Occipital cortical enhancement may be a diagnostic clue for NIID in patients with reversible encephalopathy episodes.¹

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Disclosure

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Jiping Yi, MD, PhD	Department of Neurology, The First People's Hospital of Chenzhou, The First Affiliated Hospital of Xiangnan University	Drafting/revision of the manuscript for content, including medical writing for content; study concept or design; analysis or interpretation of data

Reference

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