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

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Case summary

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). Post-contrast magnetic resonance imaging showed enhancement of the left occipital lobe cortex (Figure 1C). One year later she had recurrence of same 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].

References

Figure legend

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) Post-contrast MRI showing enhanced lesions distributed in the left occipital lobe cortex and no enhancement in the right occipital lobe cortex.

(D, arrow) Post-contrast MRI showing enhanced lesions distributed in the right occipital lobe cortex and no enhancement in the left occipital lobe cortex.
Figure 2: Skin biopsy findings

(Figure 2A, arrow): Immunostaining showed p62-positive intranuclear inclusions in sweat gland cells (×400).

(Figure 2B, arrow): Immunostaining showed ubiquitin-positive intranuclear inclusions in sweat gland cells (×400).
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