Teaching NeuroImages: The zigzag edging sign of adult-onset neuronal intranuclear inclusion disease

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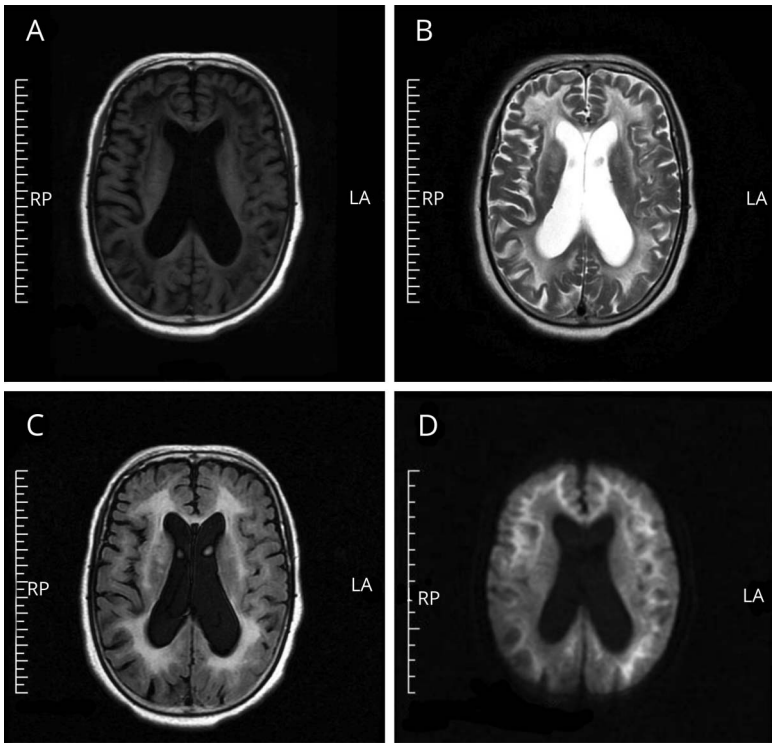
Figure 1 Brain MRI



Brain MRI shows bilateral cerebral white matter lesions, hypointensity on T1 (A), hyperintensity on the T2 (B) and fluid-attenuated inversion recovery (C), and high signal intensity along the corticomedullary junction on diffusion-weighted imaging (D).

A 66-year-old woman presented with dementia, urinary incontinence, and episodic coma for 3 years without remarkable family history. Brain MRI showed leukoencephalopathy and a high signal intensity along the corticomedullary junction on diffusion-weighted imaging (DWI) (zigzag edging sign) (figure 1). Skin biopsy revealed intranuclear inclusion bodies in sweat glands and perivascular cells (figure 2). FMRI CGG permutation was not present. Finally, the diagnosis of neuronal intranuclear inclusion disease (NIID) was confirmed.

Adult-onset NIID is a clinically heterogeneous neurodegenerative disorder. The zigzag edging sign on DWI is a strong diagnostic clue.[1] A definite diagnosis requires skin biopsy revealing intranuclear inclusions and genetic evaluation ruling out fragile X syndrome.[1,2]

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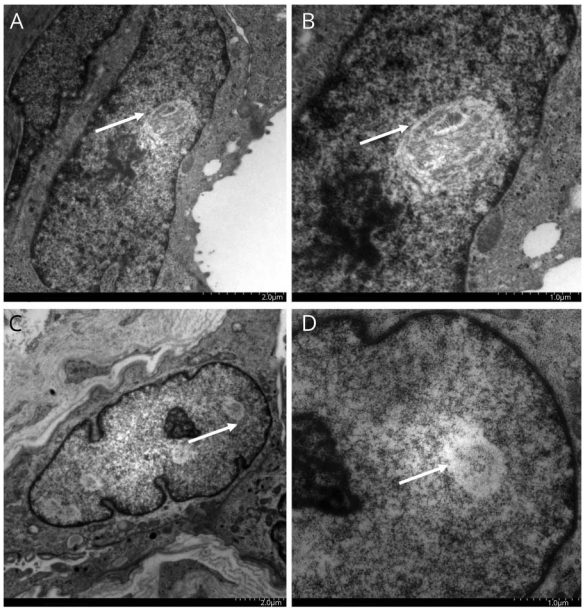
Go to Neurology.org/N for full disclosures. Funding information and disclosures deemed relevant by the authors, if any, are provided at the end of the article.

Figure 2 Skin biopsy



Electron microscope shows intranuclear inclusion bodies in sweat gland (A, B) and perivascular cells (C, D) (arrows).

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Disclosure

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Appendix Authors

Name	Location	Role	Contribution
Lizhang Chen	West China Hospital, Sichuan University	Author	Acquisition of data and wrote the manuscript
Anshan Chen	Dazhu County People's Hospital	Author	Major role in the acquisition of imaging data
Song Lei	West China Hospital, Sichuan University	Author	Electron microscope analysis of the skin specimen
Li He	West China Hospital, Sichuan University	Author	Study supervision
Muke Zhou	West China Hospital, Sichuan University	Author	Critical revision of the manuscript

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1. Sone J, Mori K, Inagaki T, et al. Clinicopathological features of adult-onset neuronal intranuclear inclusion disease. Brain 2016;139:3170–3186.
2. Sone J, Tanaka F, Koike H, et al. Skin biopsy is useful for the antemortem diagnosis of neuronal intranuclear inclusion disease. Neurology 2011;76:1372–1376.

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Teaching NeuroImages: The zigzag edging sign of adult-onset neuronal intranuclear inclusion disease

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In the article "Teaching NeuroImages: The zigzag edging sign of adult-onset neuronal intranuclear inclusion disease" by Chen et al.,[1] the second-to-last sentence in the first paragraph should read "FMR1 CGG permutation was not present." The publisher regrets the error.

Reference

1. Chen L, Chen A, Lei S, et al. Teaching NeuroImages: The zigzag edging sign of adult-onset neuronal intranuclear inclusion disease. Neurology 2019;92:e2295–e2296.

Opinion and Special Articles: Self-management in epilepsy

Web-based seizure tracking applications

Neurology® 2019;93:415. doi:10.1212/WNL.0000000000007480

In the article "Opinion and Special Articles: Self-management in epilepsy: Web-based seizure tracking applications" by Casassa et al.,[1] first published online November 19, 2018, NIH Grant T32NS048005 should have been listed as a funding source. The authors regret the error.

Reference

1. Casassa C, Rathbun Levit E, Goldenholz DM. Opinion and Special Articles: Self-management in epilepsy: web-based seizure tracking applications. Neurology 2018;91:e2027–e2030.

Cost of illness in Charcot-Marie-Tooth neuropathy

Results from Germany

Neurology® 2019;93:415. doi:10.1212/WNL.0000000000007916

In the article "Cost of illness in Charcot-Marie-Tooth neuropathy: Results from Germany" by Schorling et al.,[1] first published online March 27, 2019, the published-online-ahead-of-print version should have presented figures in USD rather than euros. They are presented correctly in the April 23 issue. The editorial office regrets the error.

Reference

1. Schorling E, Thiele S, Gumbert L, et al. Cost of illness in Charcot-Marie-Tooth neuropathy: results from Germany. Neurology 2019;92: e2027—e2037.

Comprehensive systematic review summary: Treatment of tics in people with Tourette syndrome and chronic tic disorders

Neurology® 2019;93:415. doi:10.1212/WNL.0000000000007918

In the article "Comprehensive systematic review summary: Treatment of tics in people with Tourette syndrome and chronic tic disorders" by Pringsheim et al.,[1] first published online May 6, 2019, the data supplement link in the first paragraph should have been: links.lww.com/WNL/A882. The authors regret the error.

Reference

1. Pringsheim T, Holler-Managan Y, Okun MS, et al. Comprehensive systematic review summary: Treatment of tics in people with Tourette syndrome and chronic tic disorders. Neurology 2019; 92:907–915.