RESIDENT & FELLOW SECTION

Teaching NeuroImage: Leber Hereditary Optic Neuropathy With Longitudinal Spinal Cord Lesion Mimicking Spinal Cord Infarction

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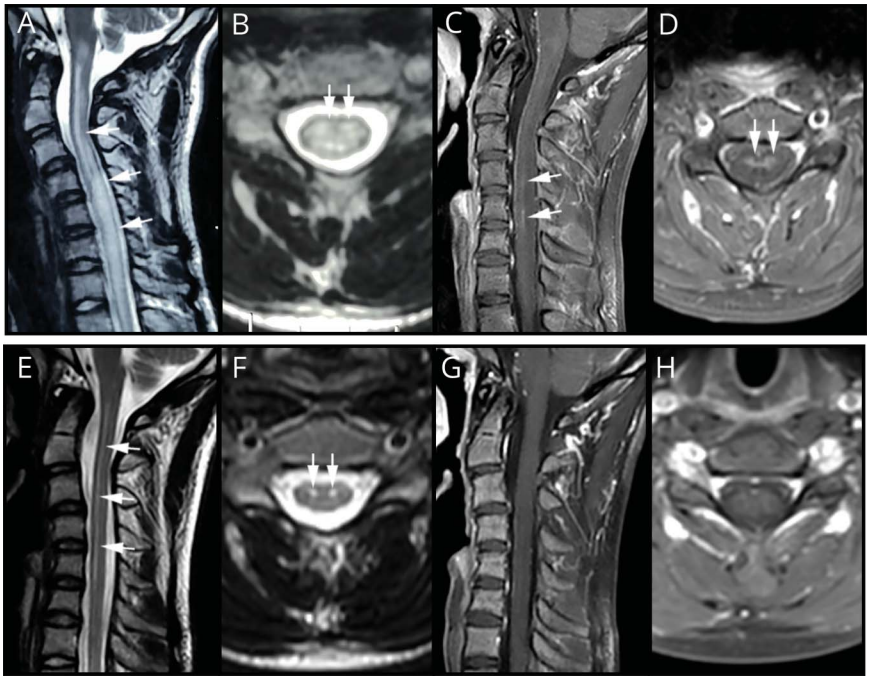
Figure Spinal Cord MRI



MRI in the acute phase demonstrated a longitudinally extensive T2 hyperintensity lesion extending from the lower medulla oblongata to the thoracic level (A, B, arrows) and a linear strip corresponding to the anterior gray matter after gadolinium enhancement (C, D, arrows). MRI in the recovery phase demonstrated the pencil-like T2 hyperintensity on sagittal views corresponding with the "snake-eye" appearance on axial planes (E, F, arrows), without any enhancement (G, H).

A 15-year-old boy with a maternal family history of "optic neuropathy" presented with acute flaccid paralysis in both upper limbs. Cervical MRI demonstrated longitudinally extensive T2 lesions, with significant enhancement of the anterior gray matter. Typical "snake-eye" appearance was observed in the follow-up MRI 1 month later (Figure). Analysis of blood mt-DNA revealed a homoplasmic m.14495T>C mutation, confirming the diagnosis of Leber hereditary optic neuropathy (LHON). Idebenone 900 mg/d was given to him, and he recovered completely 3 months later.

Spinal cord involvement was rare in LHON.[1] The enhancement pattern in this case mimicked spinal cord infarction rather than demyelinating diseases.[2]

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Name	Location	Contribution
Bing Zhao, MD	Department of Neurology, Qilu Hospital of Shandong University (Qingdao)	Drafting/revision of the manuscript for content, including medical writing for content
Yuan Sun, MD	Department of Neurology, Qilu Hospital of Shandong University (Qingdao)	Analysis and interpretation of data
Yong Qing Zhang, MD	Department of Neurology, Qilu Hospital of Shandong University (Qingdao)	Major role in the acquisition of data
Chuanzhu Yan, MD, PhD	Department of Neurology, Qilu Hospital of Shandong University (Qingdao)	Study concept or design

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