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

Author(s):

Bing Zhao, MD[1]; Yuan Sun, MD[1]; Yong Qing Zhang, MD[1]; Chuan zhu Yan, MD, PhD[1,2]

Corresponding Author:

Chuan zhu Yan
czyan@sdu.edu.cn

Affiliation Information for All Authors: 1. Department of Neurology, Qilu Hospital of Shandong University(Qingdao), Cheeloo College of Medicine, Shandong University, 758 Hefei Road, Qingdao, Shandong, 266035, China; 2.Research Institute of Neuromuscular and Neurodegenerative Diseases and Department of Neurology, Qilu Hospital, Cheeloo College of Medicine, Shandong University, Jinan, China

Contributions:

Bing Zhao: Drafting/revision of the manuscript for content, including medical writing for content

Yuan Sun: Analysis or interpretation of data

Yong Qing Zhang: Major role in the acquisition of data

Chuan zhu Yan: Study concept or design

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A 15-year-old male 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 one month later (Figure). Analysis of blood mt-DNA revealed a homoplasmic m.14495T>C mutation, confirming the diagnosis of Leber's hereditary optic neuropathy (LHON). Idebenone 900mg/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].

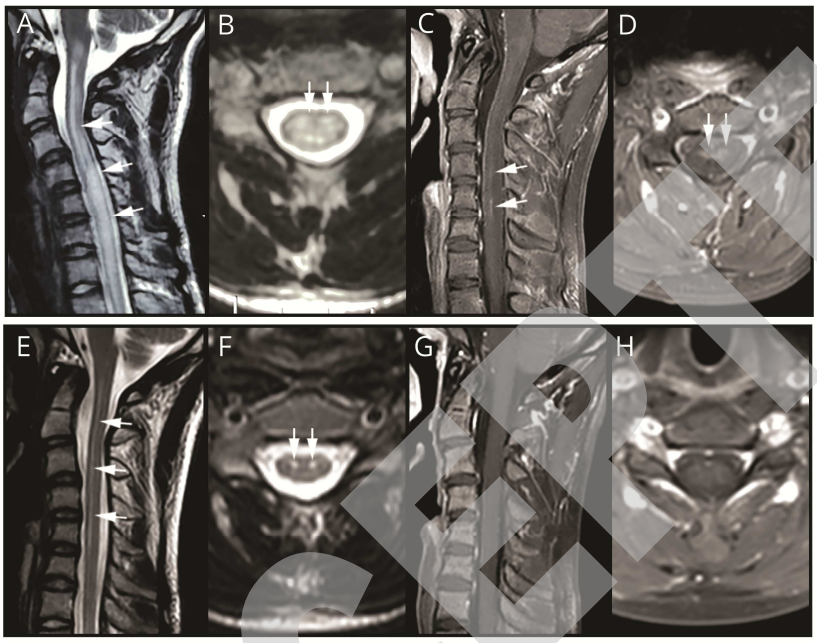
Appendix 1. Authors

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

Figure Legend

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).



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