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Teaching NeuroImages: Hirayama Disease with Symmetric Atrophy of Bilateral Distal Upper Extremities

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A 15-year-old boy presented with progressive left to bilateral hand weakness and cold paresis over one year. Examination revealed atrophy of upper limb musculature, especially bilateral dorsal interossei muscle. Neutral MRI showed cord thinning and intramedullary hyperintensity at C5-C7 levels. Neck-flexion MRI demonstrated bilaterally symmetric spinal cord compression due to dural sac anterior shifting (Figure), suggestive of Hirayama disease (HD). HD mimics ALS-like symptoms, and features an expansion of the dural sac on neck-flexion MRI[1]. The majority of HD is unilateral or asymmetric while bilaterally symmetric involvement is reported in 10% of the patients, and indicates more severe affliction[2].

Appendix 1: Authors

Name	Location	Contributions
Ye Liu, M.D. Ph.D.	Fudan University affiliated Huashan Hospital, Shanghai, 200040, China	Drafting/revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data
Yue Zhang, M.D.	Fudan University affiliated Huashan Hospital, Shanghai, 200040, China	Drafting/revision of the manuscript for content, including medical writing for content; Study concept or design
Qiang Dong, M.D.	Fudan University affiliated Huashan Hospital, Shanghai, 200040, China	Major role in the acquisition of data; Study concept or design

Teaching Slides-http://links.lww.com/WNL/B302

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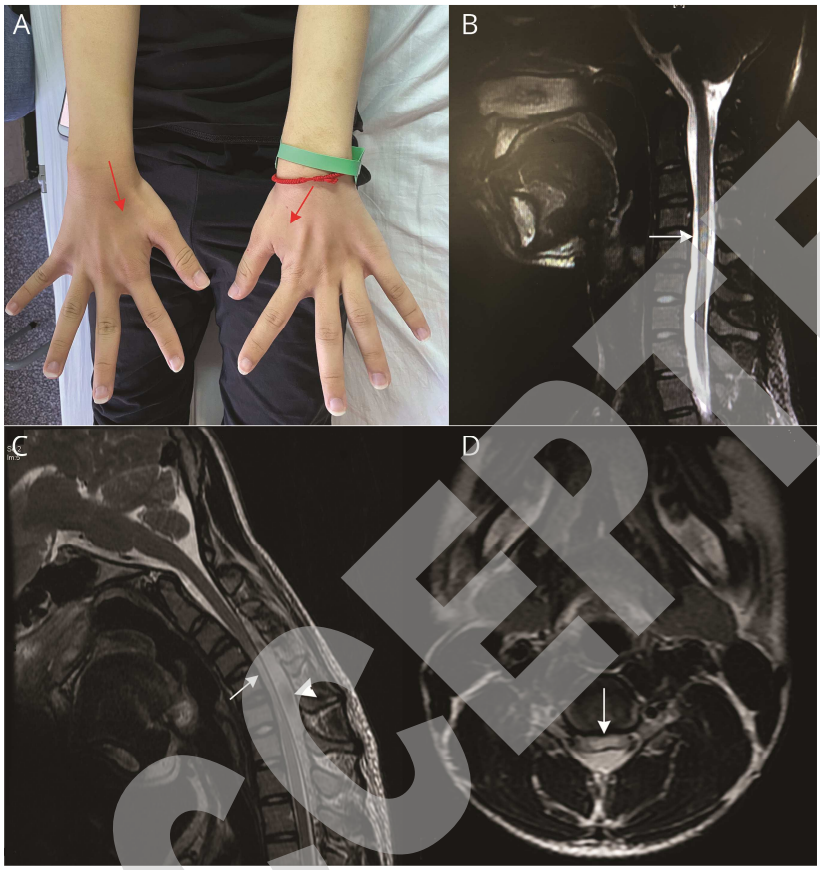
2. Pradhan S. Bilaterally symmetric form of Hirayama disease. Neurology 2009;72:2083-2089.

Figure. Photography of the patients' hands and neck MRI imaging
Wasting of bilateral dorsal interossei muscle (A). Neutral sagittal T2-weighted MRI shows cord thinning and hyperintense signal at C5 to C7 levels (B). Neck-flexion sagittal (C) and axial (D) T2-weighted MRI showed crescent-shaped enlarged posterior epidural space below C3 with flow void causing bilaterally symmetric flattening of the lower cervical cord.



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