Teaching *NeuroImages*: Anterior horn cell hyperintensity in Hirayama disease

Jamsheed A. Desai,
MD, BSc
Michel Melanson, MD,
FRCPC

Address correspondence and
reprint requests to Dr. Jamsheed
A. Desai, Queen's University,
Department of Medicine,
Division of Neurology, Kingston
General Hospital, Connell 7-76
Stuart Street, Kingston, ON,
Canada K7L2V7
3jd14@queensu.ca

Figure MRI findings in a 21-year-old man with Hirayama disease

(A) Abnormal kyphotic (straight) curvature of the cervical spine evidenced by dorsal aspects of the C3 through C6 vertebral bodies meeting a straight line (white) drawn from the dorsocaudal aspect of the C2 vertebral body to the dorsocaudal aspect of the vertebral body of C7. In addition, note the linear T2 intramedullary hyperintensity, representing atrophy and gliosis of the anterior horn cells (large arrow). (B) Transverse T2-weighted image demonstrating the owl eyes sign (hyperintensities in the anterior horns of gray matter), representing atrophy and gliosis of the anterior horn cells (white arrows).

A 21-year-old man presented with gradually progressive asymmetric bilateral weakness and atrophy of the intrinsic hand muscles that stabilized after a 3-year progression. EMG demonstrated both active denervation and chronic reinnervation of the intrinsic hand muscles. Cervical spine MRI demonstrated kyphosis of the cervical spine and high signal intensity within the anterior horns of the lower cervical cord (figure). Imaging features of Hirayama disease include 1) asymmetric lower cervical cord flattening, atrophy and hyperintense parenchymal signal, 2) abnormal cervical curvature as in our patient, and 3) dilated and enhancing epidural venous plexi and anterior shifting of the posterior dural sac on flexion

MRI.¹ Hirayama disease occurs in young individuals, with a male preponderance, and presents with distal upper extremity weakness due to segmental damage to lower motor neurons usually at the C8–T1 levels.²

AUTHOR CONTRIBUTIONS

Dr. Desai: drafting/revising the manuscript and analysis or interpretation of data. Dr. Melanson: drafting/revising the manuscript, study concept or design, and analysis or interpretation of data.

REFERENCES

1. Sonwalkar HA, Shah RS, Khan FK, et al. Imaging features in Hirayama disease. *Neurol India* 2008;56:22–26.
2. Hirayama K. Juvenile muscular atrophy of distal upper extremity (Hirayama disease). *Intern Med* 2000;39:283–290.

Neurology[®]

Teaching *NeuroImages*: Anterior horn cell hyperintensity in Hirayama disease

Jamsheed A. Desai and Michel Melanson

Neurology 2011;77:e73

DOI 10.1212/WNL.0b013e31822f02d0

This information is current as of September 19, 2011

Updated Information & Services

including high resolution figures, can be found at:
<http://n.neurology.org/content/77/12/e73.full>

References

This article cites 2 articles, 0 of which you can access for free at:
<http://n.neurology.org/content/77/12/e73.full#ref-list-1>

Citations

This article has been cited by 1 HighWire-hosted articles:
<http://n.neurology.org/content/77/12/e73.full##otherarticles>

Subspecialty Collections

This article, along with others on similar topics, appears in the following collection(s):
All Neuromuscular Disease
http://n.neurology.org/cgi/collection/all_neuromuscular_disease
All Spinal Cord
http://n.neurology.org/cgi/collection/all_spinal_cord
MRI
<http://n.neurology.org/cgi/collection/mri>

Permissions & Licensing

Information about reproducing this article in parts (figures, tables) or in its entirety can be found online at:
http://www.neurology.org/about/about_the_journal#permissions

Reprints

Information about ordering reprints can be found online:
<http://n.neurology.org/subscribers/advertise>

Neurology® is the official journal of the American Academy of Neurology. Published continuously since 1951, it is now a weekly with 48 issues per year. Copyright Copyright © 2011 by AAN Enterprises, Inc.. All rights reserved. Print ISSN: 0028-3878. Online ISSN: 1526-632X.