

# Teaching NeuroImages: A case of Hirayama disease presenting with polymyoclonus



Jonathan J.Y. Ong,  
MRCP  
Furene S.J. Wang,  
MRCPCH  
Yee Cheun Chan, MRCP

Correspondence to  
Dr. Ong:  
jonathan\_ong@nuhs.edu.sg

**Figure 1** Symmetrical bilateral forearm wasting with sparing of the brachioradialis muscles



(A) Bilateral distal forearm wasting, sparing the brachioradialis muscles (arrows). (B) First dorsal interossei and (C) hypothenar and thenar eminence atrophy.

A 17-year-old boy developed bilateral hand tremulousness, weakness, and decreased muscle bulk over 6 months. Bilateral extremity wasting with sparing of brachioradialis muscles (figure 1) was evident. With arms outstretched, distal mini-polymyoclonus (video on the *Neurology*® Web site at Neurology.org) occurred. Neurophysiology suggests a pure motor neuropathy (chronic denervative changes in C7-T1 roots). MRI spine (dynamic flexion sequences)<sup>1</sup> (figure 2) revealed cervical cord atrophy, dural sac anterior shifting, and contrast-enhancing epidural compartment. The constellation of features suggests Hirayama disease,<sup>2</sup> postulated to be secondary to chronic segmental cord ischemia from repeated neck flexion over a tight dural sac. The polymyoclonus should not be mistaken for tremors.

#### AUTHOR CONTRIBUTIONS

Jonathan J.Y. Ong: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for

conduct of research and final approval. Furene S.J. Wang: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval. Yee Cheun Chan: drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, acquisition of data, study supervision.

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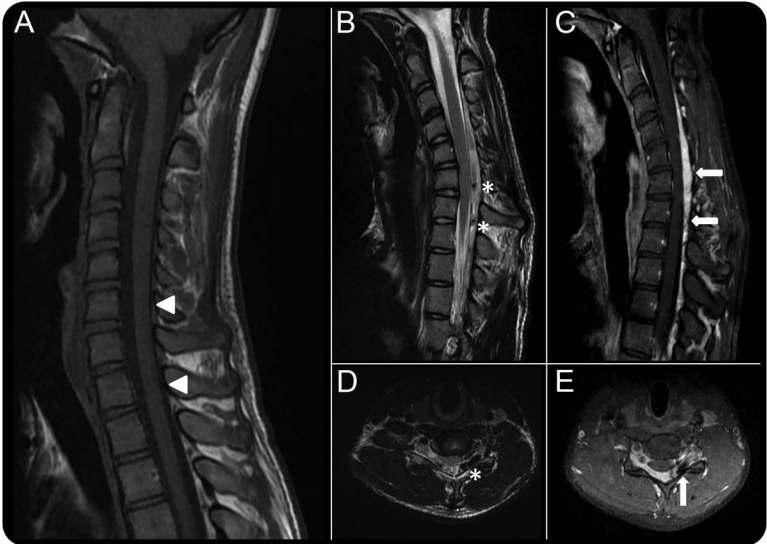
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From the Departments of Neurology (J.J.Y.O., Y.C.C.) and Pediatrics (F.S.J.W.), National University Hospital, Singapore.



(A) Nonflexion T1-weighted MRI shows C7-T1 cord atrophy (arrowheads). (B, D) Flexion T2-weighted MRI shows anterior shifting of posterior dural space with flattening of cervical cord. Engorgement of epidural venous plexus (asterisks) enhanced with contrast (C, E) (T1-weighted) (arrows).

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