



## Dynamic myelopathy in Hirayama disease

An 18-year-old Portuguese Caucasian man presented with a gradual onset of painless weakness and atrophy of the right hand and forearm, accompanied by a numb sensation in the hand that had been worsening over the past 12

months despite physiotherapy. There was no history of cold paresis. Neurologic examination revealed atrophy in all muscles of the forearm and intrinsic hand muscles, no fasciculations, right upper limb (UL) hypotonia, distal and extensor predominant motor deficit, right UL pathologic hyperreflexia, and right C6–C8 dermatomes hypalgesia. Electrophysiologic study demonstrated severe chronic denervation of right C7–T1 myotomes, mild chronic

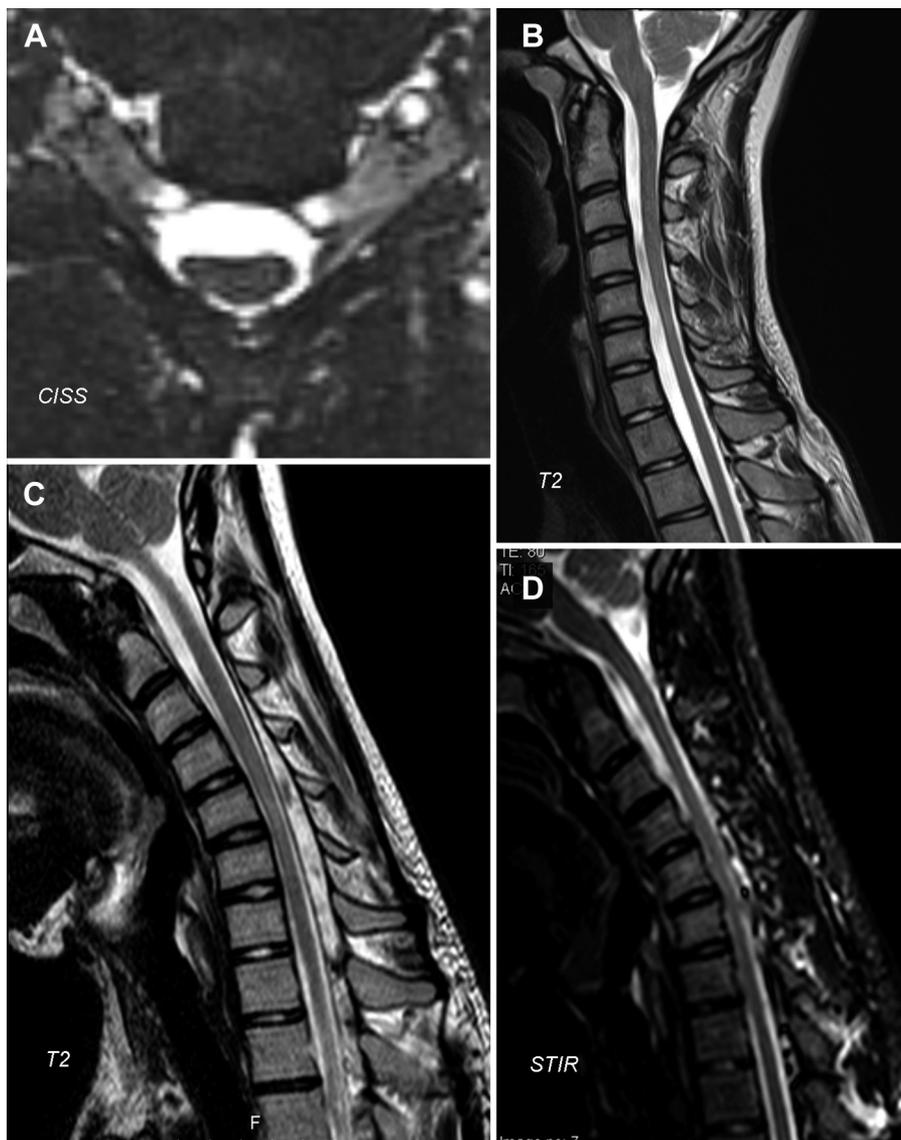


Figure. Cervical magnetic resonance imaging (MRI), dynamic study. (A and B) Cervical MRI in neutral position, showing posterior spinal cord displacement and asymmetrical flattening of lower cervical cord. (C and D) Cervical MRI in flexion position, showing anterior dural shift, parenchymatous T2-hyperintensity, and epidural venous plexus engorgement.

denervation of left C7–T1 myotomes, and marked reduction of right cubital CMAP amplitude. Cervical magnetic resonance imaging in neutral position showed an asymmetrical lower cervical cord flattening (Figure, A) and a posterior displacement of the spinal cord with no extra-axial lesion (Figure, B). Cervical flexion induced cervical cord compression and T2-hyperintensity, anterior dural shift (Figure, C), and epidural venous plexus engorgement (Figure, D). Patient was recommended to use a neck collar, to improve neck and back posture, to sleep without a pillow, and to avoid sports with risk of head and neck trauma. The patient has been stabilized for the last 9 months.

Hirayama disease, also known as juvenile muscular atrophy of distal extremity, occurs more frequently in young men and is characterized by insidious asymmetrical UL weakness and muscular atrophy [1]. The pathologic lesion is in the anterior horn cells of the spinal cord, most commonly at C7 and T1 [2]. Familial cases, bilateral UL involvement and sensory changes have been reported. The disease usually stops progressing months to years after onset. Electrophysiologic tests are important to exclude other conditions. Magnetic resonance imaging scan in the neutral position can show subtle cord atrophy [2]. Anterior displacement of the cord during flexion, as seen in this patient, has been proposed as the main pathophysiologic mechanism [2]. Several characteristics of this disease remain to be explained, including the male predominance. A genetic contribution has been suggested [3].

## References

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