Cervical spinal cord compression complicating the clinical course of Charcot-Marie-Tooth type 1

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DESCRIPTION

A 73-year-old woman with Charcot-Marie-Tooth type 1 (CMT1) had followed a typical slowly progressive course since onset. She had bilateral distal upper and lower limb weakness and walked with a single stick from 50 years of age, requiring two sticks following a left internal capsular stroke at 63 years of age. Neurological examination was consistent with a severe, length-dependent sensorimotor neuropathy: distal wasting/weakness in all limbs, with additional mild right lower limb pyramidal pattern weakness consistent with the previous stroke. There was decreased vibration sense and proprioception to the knees, with loss of pin sensation to knees and elbows. Reflexes were absent. Plantar responses were flexor. Neurophysiology revealed homogenous slowing of motor nerve conduction velocities (24 m/s in right median nerve), with absence of temporal dispersion, conduction block, and of accentuated proximal and distal slowing, consistent with an inherited demyelinating neuropathy.

Following years of slow progression, at 73 years of age she was admitted following three

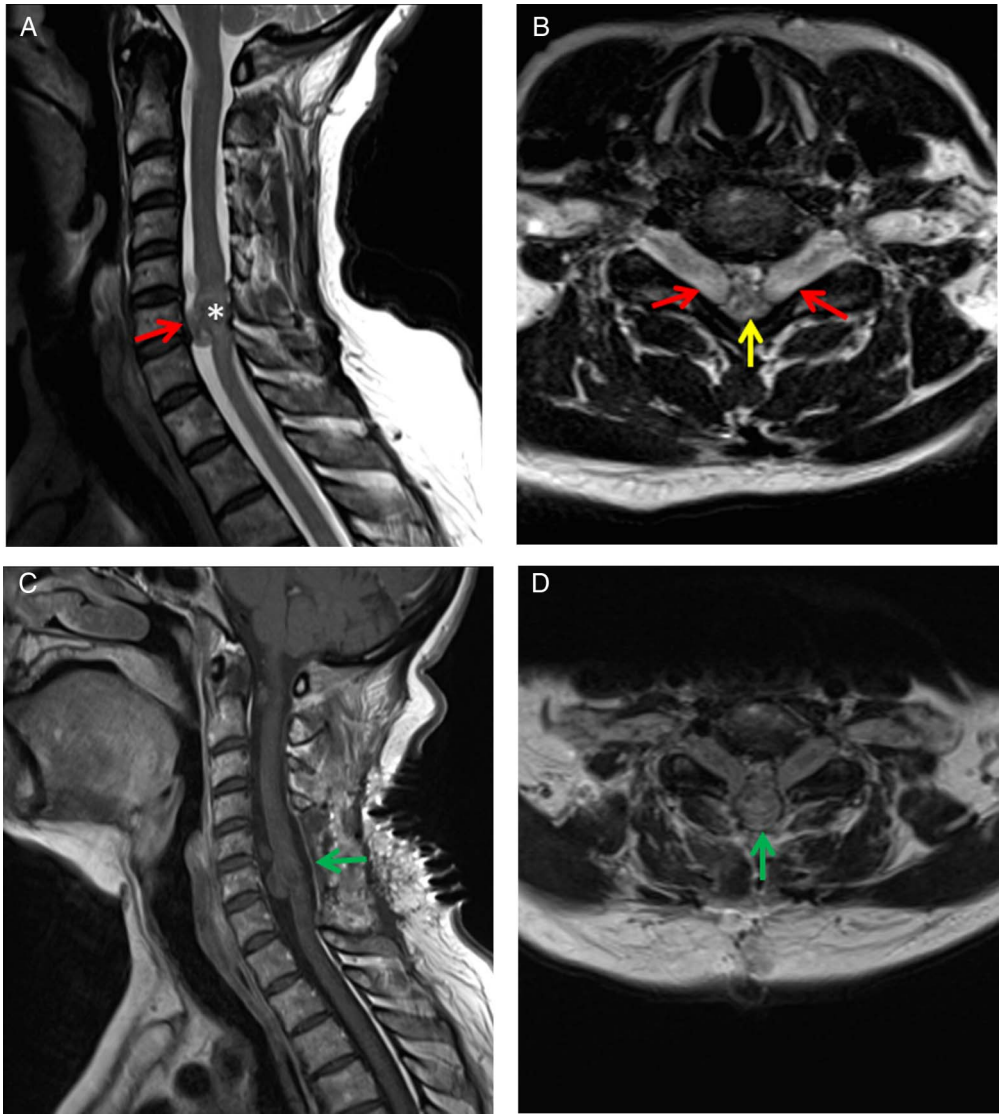

Figure 1 Preoperative T2-weighted sagittal (A) and axial (B) MRI of the neck showing thickened C7 nerve roots (red arrows) causing compression (asterisk) and intramedullary signal change (yellow arrow) in the spinal cord. Postoperative gadolinium-enhanced T1-weighted sagittal (C) and axial T2-weighted (D) images demonstrating adequate decompression (green arrows).

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falls in 1 month. Her right leg had become significantly weaker over the preceding 6 months and she could no longer mobilise. There was no sphincter involvement. There was now marked right lower limb pyramidal pattern weakness with an extensor plantar response; the remainder of the examination was unchanged from the previous visit. A central lesion was suspected.

Brain MRI was unremarkable, however, enlarged preganglionic cervical nerve roots, documented on an MRI study performed a decade prior, had enlarged further, resulting in compression and intramedullary signal change in the mid-lower cervical spinal cord (figure 1A, B). The patient underwent urgent C5–7 laminectomy, postsurgical imaging confirming adequate thecal decompression (figure 1C, D). Following 6 weeks of intensive neurorehabilitation, she returned to independent ambulation.

Learning points

- Spinal cord compression is a serious, albeit rare complication of enlarged nerve roots, secondary to both chronic inflammatory demyelinating polyradiculoneuropathy[1] and inherited demyelinating neuropathies.[2 3]
- This case accentuates the importance of meticulous re-examination and consideration of an alternative diagnosis in patients with Charcot-Marie-Tooth type 1 presenting with deterioration out of keeping with the expected natural history.

Contributors MRBE, ML and MMR wrote and edited the imaging case report. HC prepared and reported imaging.

Competing interests None declared.

Patient consent Obtained.

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