Monomelic Amyotrophy (Hirayama’s Disease)

Last month’s case presented weakness and atrophy of the right hand muscles in a 47-year-old man.

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The lesion is likely located at the level of:
1. Motor neurons
2. Motor nerves
3. Brachial plexus
4. Axons
5. Myelin

MONOMELIC AMYOTROPHY (HIRAYAMA’S DISEASE)

• The presence of diffuse weakness and denervation that spans multiple nerve roots with preservation of sensory responses and reduced reflexes suggest a lower motor neuron lesion.
• Multifocal motor neuropathy with conduction block may present similarly, but the presence of conduction blocks and the lack of diffuse denervation would distinguish the case. This patient was treated with IVIG before referred to our center with no improvement.
• Cervical polyradiculopathy due to an infiltrative lesion of the motor nerve root, such as lymphoma, is possible, but the long duration and normal cervical MRI ruled out this possibility.
• ALS may start with focal lower motor neuron signs, but it usually spreads to other muscles and evolves into a typical picture within a year or two. Distal spinal muscular atrophy is usually bilateral and distal.
• Chronic focal lower motor neuron disease is well known.
• It is sporadic and affects males 10 times more often than females. Age of onset is 13-15 years.
• Starts with distal single limb weakness, right is twice as likely affected as left.
• Subclinical involvement of the other arm is common, and in 10 percent of cases proximal weakness is evident.
• 40 percent of times it spreads to the other side. It progresses over five years and then plateaus for decades.
• Most denervation is in C8-T1 muscles, but proximal and even lower extremity muscles may be affected. Cervical MRI: cord atrophy is seen in 30-50 percent of cases.
• It is speculated that posterior epidural venous plexus engorgement during flexion contributes to the pathology, but that does not explain the presence of denervation in the legs.