Clinical Reasoning: A 17-year-old baseball player with right hand weakness

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Section 1

A healthy 17-year-old baseball player presented with a 2-year history of right elbow pain and hand weakness. He first noticed pain on the lateral aspect of his right elbow exacerbated by movement and heavy lifting. Following a 4-month period of rest and physical therapy, he resumed playing baseball. Shortly thereafter, he developed the same right elbow pain, but also felt that his right grip was getting progressively weaker. With repetitive flexion at the elbow, he would feel a "snap" that caused tingling down his forearm. His right hand felt cooler than his left and his grip strength seemed weaker when in cold weather. He otherwise denied neck pain, bowel/bladder dysfunction, fasciculations, and cramps. He did not notice any weakness or sensory change in his other extremities. There was no antecedent trauma, infection, or vaccination. He did not take medications or use illegal substances. Family history was unremarkable. His neurologic examination revealed intact cranial nerves. His tone and bulk were normal except for mild atrophy of his right hypothenar eminence and the ulnar side of his distal forearm (figure 1), which is known as the reverse split hand sign.[1] Passive flexion of the right elbow caused tingling down his forearm. Finger adduction in addition to fifth digit flexion and abduction were 4/5. There were no fasciculations. His reflexes were 2+ throughout. There was no sensory disturbance in either hand. The remainder of his examination including gait and coordination were normal.

Questions for consideration:

1. Where would you localize the lesion?
2. What is your differential diagnosis at this stage?

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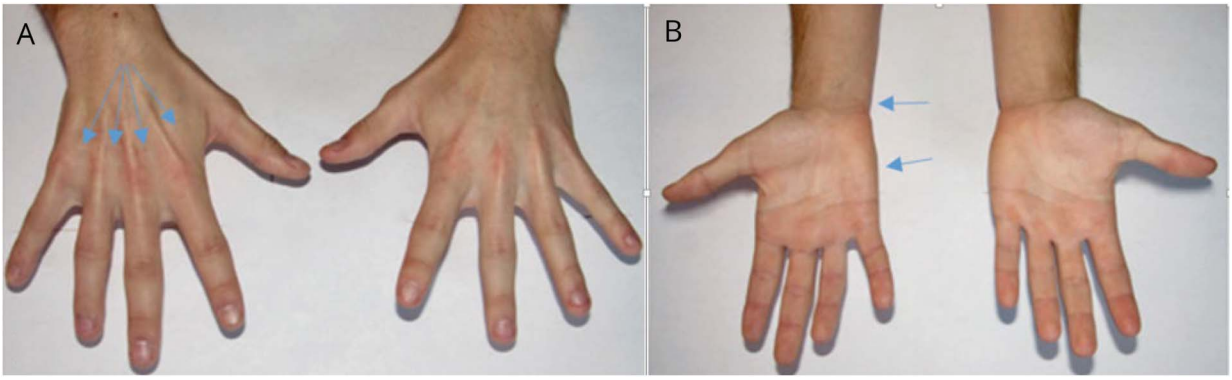
Figure 1 Examination findings



Reverse split hand sign: note the atrophy of the (A) interossei, (B) hypothenar eminence, and distal forearm in the right upper extremity (arrows).

Section 2

Given the findings of hand weakness with focal atrophy and no increase in tone or reflexes, the lesion localizes to a lower motor neuron process. The atrophied muscles get their motor innervation from anterior horn cells, which give rise to the C8 and T1 nerve roots, medial cord, and ulnar nerve. Given the history of sensory disturbance with elbow flexion, the differential diagnosis should include compressive ulnar neuropathy at the elbow. Despite the history of flexion-provoked paresthesias, the patient's sensory examination was unremarkable. This should prompt a consideration of processes that result in lower motor neuron dysfunction.

The differential diagnosis of a progressive lower motor neuron syndrome affecting a single arm is broad and includes the following[2]:

1. Structural cervical spine disease including radiculopathy, cervical spondylotic amyotrophy (CSA), Hirayama disease, or syringomyelia
2. Neurodegenerative disorders including progressive muscular atrophy and amyotrophic lateral sclerosis (ALS)
3. Infections affecting anterior horn cells including HIV, poliovirus, and West Nile virus.
4. Thoracic outlet syndrome
5. Idiopathic inflammatory conditions including multifocal motor neuropathy and brachial plexitis (e.g., Parsonage Turner syndrome)
6. Compressive ulnar, radial, or median mononeuropathy
7. Toxins including lead that can present with subacute radial neuropathy

In this case, given this patient's age and presentation, many of these possibilities can be easily eliminated. Given a progressive 2-year course isolated to one myotome, multifocal motor neuropathy seems unlikely. The pain in his elbow is inconsistent with the severe shoulder pain seen in brachial plexitis. The clinical picture is most concerning for compressive mononeuropathy or structural cervical spine disease.

Question for consideration:

1. What is the most appropriate next step in this patient's work-up?

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Section 3

The patient underwent a nerve conduction study (NCS) with EMG of his upper extremities. Sensory nerve conduction studies showed normal right median and ulnar sensory nerve action potentials. The motor nerve conduction studies of the right ulnar abductor digiti minimi showed low-amplitude compound motor action potentials (CMAPs) with normal latency and conduction velocity (CV). Right ulnar first dorsal interosseous (FDI) showed no slowing across the elbow. Right median abductor pollicis brevis CMAPs had normal latency, amplitude, and CV. Needle EMG showed normal insertional activity in both arms and in the cervical paraspinals. The motor unit action potentials in the right FDI, extensor indicis proprious, and extensor digitorum communis were large with reduced recruitment.

The above findings suggest chronic reinnervation with localization to C7–8 myotomes on the right. Despite the patient's symptoms of right elbow pain with positive Tinel sign, there was no evidence of ulnar neuropathy. MRI of the elbow was obtained to evaluate his focal pain and revealed a normal ulnar nerve, but did reveal a partial thickness tear of the ulnar collateral ligament, likely from pitching. Given the above explanation of his elbow pain and elimination of an ulnar neuropathy, the differential is narrowed to structural or functional cervical spine disease at the level of C7–8.

Given the patient's predominant motor weakness in a focal distribution referable to C7–8, more specific localization would implicate anterior horn cells at that same level. This distinctly narrows the differential to conditions that result in unilateral anterior horn cell dysfunction or loss. His clinical course of progressive focal weakness over 2 years is incongruent with an acute ischemic event or viral infection. Spinal muscular atrophy type 3 (Kugelberg-Welander disease) can present anywhere from childhood to adulthood, but would more typically involve the proximal muscle groups beginning in the lower extremities.[3] Despite his young age, it is imperative to consider ALS on the differential. Hirayama disease (HD) and cervical spondylotic amyotrophy should also be considered. These conditions can be differentiated using NCS. Thenar and hypothenar muscles are both innervated by C8–T1. ALS and HD tend to cause thenar and hypothenar atrophy, respectively. While cervical spondylotic amyotrophy tends to affect the cervical cord, it almost always manifests with shoulder abduction weakness, positive arm-drop sign, or wrist drop.[4] The ulnar to median CMAP ratio is lower in patients with HD, higher in patients with ALS, and normal in patients with CSA.[5] The ulnar to median CMAP ratio in this case was 0.25, which is low (normal 0.6–1.7).

Plain films of the patient's cervical spine revealed loss of cervical lordosis, but no evidence of a cervical rib or spondylotic disease. An MRI of the cervical spine did not reveal any lateral or central disc herniation, but did reveal subtle cervical cord atrophy in the lower cervical region.

Questions for consideration:

1. How does the MRI finding influence the differential?
2. What additional imaging is warranted?

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Section 4

The finding of cervical cord atrophy is suggestive of neuronal loss. Given the suspicion for HD, MRI of the cervical spine with dynamic flexion views was completed. The cervical cord again was mildly thinned at the level of C5–7. With flexion, there was anterior displacement of the dorsal dura in the mid to lower cervical segments, producing apparent cord flattening (figure 2). Thus, the clinical course, imaging, and electrodiagnostics were most consistent with HD.

Discussion

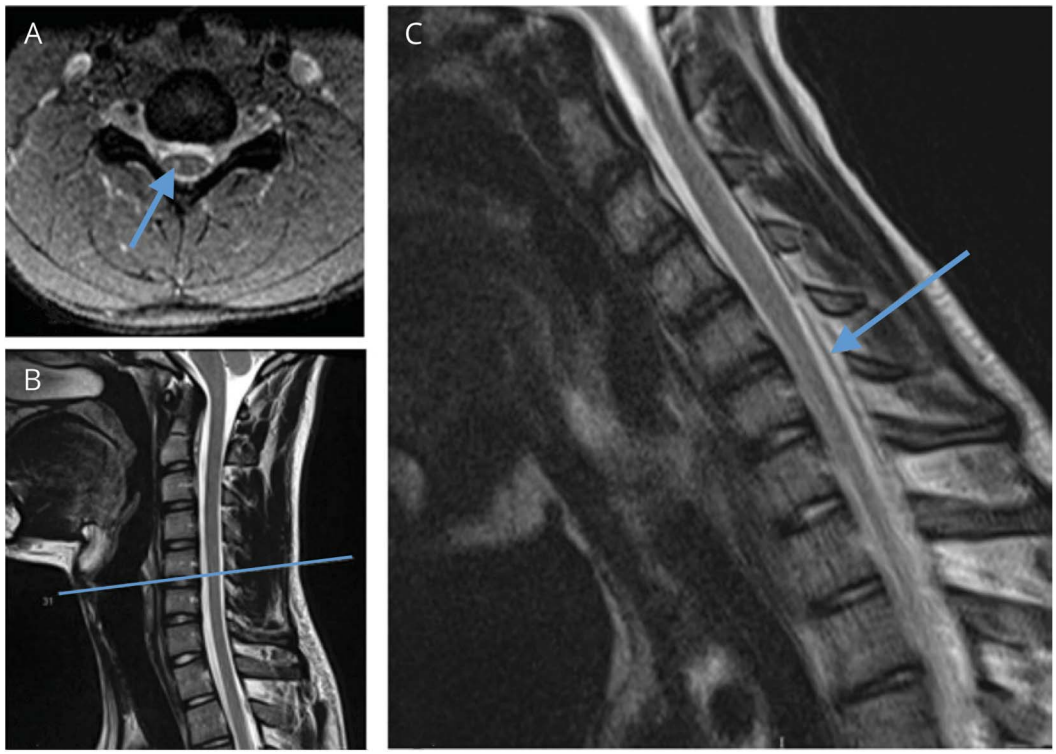
HD was first described in 1959 by Hirayama et al.[6] Other terms used in the literature include benign focal amyotrophy, monomelic amyotrophy, and benign juvenile muscular atrophy of the distal upper extremity.[7] The mean age at onset is around 18 with a range between 13 and 33.[8] It predominantly affects men in a 10–18 to 1 ratio.[8,9] The incidence of focal amyotrophy varies geographically. In Japan, the prevalence is estimated to be 1/30,000, whereas in Western countries it accounts for approximately 3% of motor neuron disease.[8]

The typical clinical features include insidious onset of focal atrophy and weakness in the C7–T1 segmental myotome. Patients have hypothenar and forearm atrophy sparing the brachioradialis, which results in a reverse split hand and oblique amyotrophy, respectively.[8] Though initially reported as an exclusively unilateral process, asymmetric bilateral upper extremity involvement has also been described.[7] Patients commonly experience "cold paresis," or worsening weakness when in cold temperatures. The prognosis is generally favorable, with a rate of severe disability less than 5%. The progression of symptoms typically lasts 2–5 years before spontaneously stabilizing.[8] There have been few cases that progress beyond 5 years, most of which have bilateral involvement.

EMG/NCS is necessary for precise localization and to obtain the ulnar to median ratio, which will be low (<0.6) in HD. It is imperative to obtain flexion views on MRI of the cervical spine. MRI findings supportive of HD include abnormal cervical curvature, T2-weighted imaging hyperintensities in the lower cervical cord, loss of dural attachment on flexion views, and thinning and compression of the lower cervical cord.[9] Boruah et al.[9] furthermore identified a postgadolinium enhancing posterior epidural crescent due to engorged epidural venous plexus in all 19 patients studied who had bilateral involvement.

The pathophysiology is likely forward displacement of the posterior cervical dural sac, which leads to spinal cord compression.[8] The dural displacement could be due to disproportionate growth of the cervical cord and spine in adolescence. This produces posterior venous congestion, which can result in ischemia to the vulnerable anterior horn

Figure 2 Radiographic findings



(A) Axial T2-weighted imaging shows punctate hyperintensity (short arrow) correlating to (B) region of cervical cord atrophy. (C) Flexion image with sagittal T2-weighted imaging shows loss of posterior dural attachment (long arrow) and cord flattening.

cells. The treatment is limiting cervical flexion until the disease process is believed to have stabilized. This can include wearing a cervical collar, which was recommended for our patient. In more severe cases, cervical spine arthrodesis can be considered.

Author contributions

C. Vachon: manuscript preparation. A. Abu Libdeh: identified pertinent clinical case, critical revision of manuscript.

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Disclosure

The authors report no disclosures relevant to the manuscript. Go to Neurology.org/N for full disclosures.

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