Weak hands from a flexed neck

An 18-year-old man presented with 2 years of progressive hand weakness. Two routine cervical MRIs were normal. Examination demonstrated weakness and atrophy in left C7–8 myotomes, and weakness in right C7–8 myotomes. The examination was otherwise normal. EMG demonstrated fibrillation potentials and neurogenic motor unit potentials in C7–T1 myotomes bilaterally. The clinical features were characteristic of juvenile distal segmental muscular atrophy (Hirayama disease). Cervical MRI with neck flexion was typical of this disorder, exemplifying the importance of flexion images1 (figure). Treatment includes observation, cervical collar, or cervical fusion. Response is controversial; however, progression typically ceases spontaneously after several years.

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