

Cervical Myelopathy Presenting with Positional Pseudomyotonia: a Case Report



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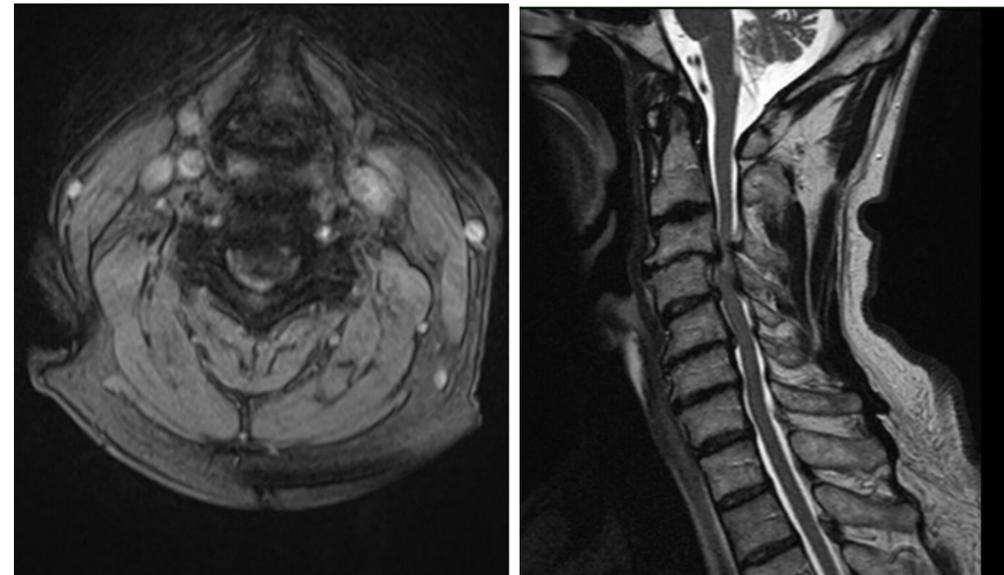
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Case Description

A 65-year-old male presented for electrodiagnostic evaluation for hand weakness. He reported 3 weeks of worsening extremity weakness, gait ataxia, and impaired hand relaxation. He was hyperreflexic with bilateral Hoffman sign and significantly impaired active finger extension. Bilateral upper limb strength and finger extension was notably worsened with neck extension. Sensation to light touch and pinprick was diminished in a length dependent fashion.

Needle EMG demonstrated bilateral symmetrically reduced recruitment with slightly large, polyphasic motor units in all tested muscles. Spontaneous activity could not be fully assessed due to impaired relaxation. Paraspinal EMG demonstrated one isolated prolonged myotonic discharge that was not repeatable and multiple CRDs.

Urgent cervical MRI demonstrated congenital spinal canal stenosis with superimposed posterior osteophyte complex formation and ligamentous infolding causing severe canal stenosis at C3-4 with acute on chronic cord compression and signal change. He subsequently underwent posterior cervical decompression and fusion.



Cervical spine MRI with severe canal stenosis at C3-4 with cord compression (axial and sagittal view)

EMG											
Side	Muscle	Nerve	Root	Fibs	Psw	MUPC Amp	Dur	Poly	Recrt	Comments	
Bilateral	Deltoid	Axillary	C5-6			Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Bilateral	Biceps	Musculocut	C5-6			Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Bilateral	Triceps	Radial	C6-7-8			Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Bilateral	Pronator Quad	Median (Ant Int)	C7-8			Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Bilateral	1stDorInt	Ulnar	C8-T1			Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Bilateral	Abd Poll Brev	Median	C8-T1	2+	2+	Incr	Nml	2+	Reduced	Inconclusive Fibs/ Psw due to persistent tone	
Paraspinal EMG											
Side	Muscle	Nerve	Root								Comments
Bilateral	C7 Parasp	Rami	C7-T1								CRD and one episode of myotonia

Needle EMG significant for one isolated prolonged myotonic discharge that was not repeatable and multiple CRDs with bilateral C7 paraspinal muscles

Discussions

Myotonia is delayed muscle relaxation after activation from increased excitability of muscle fibers resulting in repetitive discharge of action potentials. Clinical myotonia classically improves whereas pseudomyotonia worsens with repeated muscle activation. This patient demonstrated pseudomyotonia with impaired finger extension after forceful voluntary hand contraction. His impaired finger relaxation worsened with repeated efforts and cervical extension, improved with cervical flexion and extended periods of muscle rest. This is presumed secondary to maladaptive reorganization of the corticospinal tracts due to injury.

Conclusions

Classically, pseudomyotonia is characterized as delayed muscle relaxation after contraction without myotonic discharges on EMG. While this patient exhibited one myotonic discharge on paraspinal needle study, it was not repeatable within the muscle or in other muscle groups. Pseudomyotonia is rarely associated with cervical myelopathy but should be in the differential when physical exam and needle EMG demonstrates signs of impaired relaxation. It may also be a sign of chronic spinal cord compression.

References:

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- Satoyoshi E, Doi Y, Kinoshita M. Pseudomyotonia in cervical root lesions with myelopathy. A sign of the misdirection of regenerating nerve. *Arch Neurol.* 1972 Oct;27(4):307-13. doi: 10.1001/archneur.1972.00490160035005. PMID: 4341307.