

E-Pearl of the Week

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Cervical MRI with anterior flexion in juvenile distal segmental muscular atrophy

Juvenile distal segmental muscular atrophy (also referred to as Hirayama disease) is an insidious condition causing weakness and atrophy in muscles of the hand and forearm. It is often seen in young men. Most patients have unilateral disease, and some have weakness and atrophy that is bilateral but asymmetric. Finger tremor can be seen during movement, due to fascicular twitching of forearm muscles. An initial progressive clinical course is followed by an arrest of progression after a few years.

Cervical MRI shows atrophy of the lower cervical spinal cord in about half of patients. A more dramatic finding on cervical MRI with anterior flexion is anterior displacement of the dural sac and cord resulting in the appearance of an enlarged posterior epidural space in which engorged epidural veins are characteristically seen. This highlights the importance of requesting MR sequences with anterior neck flexion in patients with progressive weakness and atrophy of hand and forearm muscles.

Dimberg EL, Rubin DI. Weak hands from a flexed neck. *Neurology* 2009;73:996.

Hirayama K, Tokumaru Y. Cervical dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. *Neurology* 2000;54:1922–1926.

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Disclosure: Dr. Overman is a member of the Resident & Fellow Team and Podcast Committee for the *Neurology* Journal.