

Hirayama disease – MR imaging in neutral and flexion positions.

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Section: Neuroradiology

Area of Interest: Neuroradiology spine

Imaging Technique: MR

Case Type: Clinical Cases

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Patient: 19 years, male

Clinical History:

A 19-year-old male patient presented with unilateral upper limb weakness for the last eight months. Neurological examination revealed weakness and muscle wasting in right forearm and hand (Fig. 1), without sensory disturbance. No sensory or motor deficit elicited in left upper limb or in bilateral lower limbs.

Imaging Findings:

Cervical MRI plays an important role in the diagnosis of this rare disease. Imaging in our case is done with 1.5 T MRI (Philips Multiva). Imaging protocol includes T1-and T2-weighted imaging in both axial and sagittal planes and in both neutral and flexion positions. MR imaging in neutral position demonstrates the subtle focal cervical spinal cord atrophy (Fig. 2 & 3), especially in the lower cervical region. Flexion of neck is achieved by placing the custom built positioning sponges under head. Imaging in this position, additionally elicits the anterior displacement of dura with resulting widening of the posterior epidural space (Fig. 4) and more pronounced cord thinning, which was not obvious in neutral position (Fig. 5).

Discussion:

Hirayama disease is a juvenile form of spinal muscular atrophy characterised by unilateral or asymmetric bilateral involvement of hand and forearm muscles (C7-T1 myotomes) [1]. This non-progressive focal amyotrophy predominantly affect males in their second decade of life [2,3]. Relative sparing of sensory system and brachioradialis muscle are characteristic of this oblique amyotrophy [1]. Although the exact mechanism is uncertain some believe that disproportionate growth of vertebral column and spinal canal contents is responsible for the short and tight dural sac, resulting in cord compression on flexion [1]. This chronic process leads to microcirculatory disturbance in the anterior spinal artery territory [1]. MRI findings in neutral position include loss of cervical lordosis, localised cord thinning or flattening and rarely intramedullary signal changes [1,2,4]. Imaging in flexion position reveals forward displacement of the dura with resultant enlargement of posterior epidural space. Contrast MRI shows crescent shaped enhancing epidural space with flow voids [2,4]. Findings may be subtle, in routine MRI, in neutral position as in our case. Hence, dynamic MR imaging of cervical spine in neutral and flexion positions is important in diagnosis especially in patients with classical clinical presentation. Early diagnosis is essential as early application of cervical collar decreases the morbidity of this condition. Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Juvenile asymmetric segmental spinal muscular atrophy (JASSMA) or Hirayama disease., Spinal muscular atrophy , Amyotrophic lateral sclerosis, Toxic neuropathy, Post polio syndrome

Final Diagnosis: Juvenile asymmetric segmental spinal muscular atrophy (JASSMA) or Hirayama disease.

References:

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Figure 1

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Description: Wasting of muscles in right hand when compared to normal muscle bulk in left hand.

Origin: Department of Radiology and Imaging Sciences, Meenakshi hospital, Tanjore, India.

Figure 2

a



Description: Cervical T1W-Sagittal MR image in the neutral position shows focal thinning of the lower cervical cord at the C6-7 vertebral level. **Origin:** Department of Radiology and Imaging Sciences, Meenakshi hospital, Tanjore, India.

Figure 3

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Description: Cervical T2-Sagittal MR image in the neutral position shows focal thinning of the lower cervical cord at the C6-7 vertebral level (arrow) without any abnormal intramedullary signal intensity.
Origin: Department of Radiology and Imaging Sciences, Meenakshi hospital, Tanjore, India.

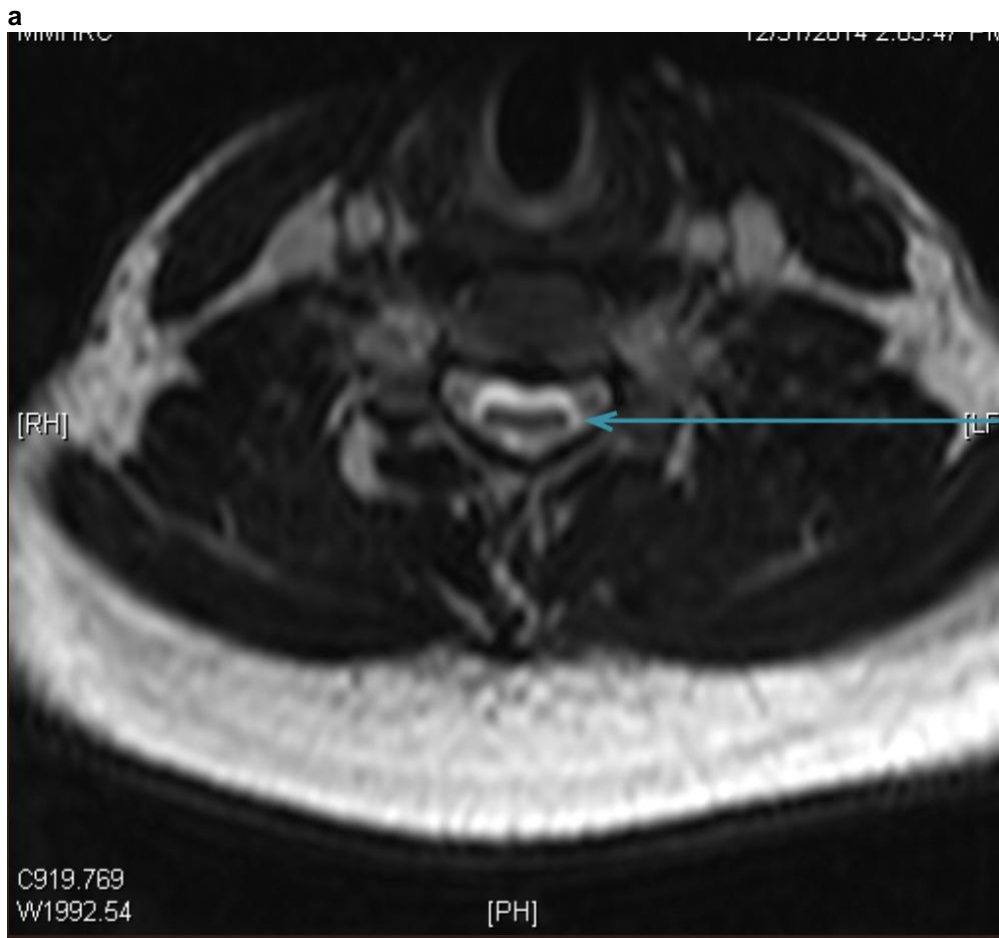
Figure 4

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Description: Cervical T2-Sagittal MR image in the flexion position shows the anterior shifting of posterior wall of the dural sac between C5 and C7 vertebral levels and the cervical cord compressed over the posterior surface of the vertebra (arrow). **Origin:** Department of Radiology and Imaging Sciences, Meenakshi hospital, Tanjore, India.

Figure 5



Description: Cervical T2-Axial MR show the asymmetric and marked flattening of cervical cord (arrow).

Origin: Department of Radiology and Imaging Sciences, Meenakshi hospital, Tanjore, India.