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The importance of dynamic flexion in the assessment of hirayama disease: A case series

Vignesh Muthukumaran[1], Mohammad Hassan[2*], Karunya Lakshmi[2], Sivasubramaniyan KM[2], Sukrit Deo[1]
Department of Radiodiagnosis, Sri Lakshmi Narayana Institute of Medical Sciences, Puducherry, India
Assistant Professor, Department of Radiodiagnosis, Sri Lakshmi Narayana Institute of Medical Sciences, Puducherry, India

Abstract
Introduction: Hirayama disease is a non-progressive juvenile spinal muscular atrophy of the distal upper limbs. It is a cervical myelopathy caused due to flexion movements of the neck. This is a case series of six young males presenting with atrophic changes of muscle groups involving distal upper limbs, and with Hirayama disease as their clinical diagnosis.
Methods: We retrospectively reviewed cases of Hirayama disease in 6 patients referred for MR imaging to our department. We evaluated the clinical presentation and etiology of Hirayama disease based on multimodality imaging findings. Dynamic MR imaging was done in all patients to assess the degree of disease involvement of the disease.
Results: In our study of 6 cases, all revealed anterior displacement of the detached posterior dura from the underlying lamina with varying degree of compression of the thecal sac. Widening of the posterior epidural space with flow voids were seen better on 3D-CISS images. Conservative management and follow up imaging were suggested in all patients. MRI is the modality of choice in establishing the diagnosis of Hirayama disease. Specifically dynamic flexion imaging is of paramount importance in the accurate depiction of imaging findings.

Keywords: hirayama, dynamic flexion, non-progressive juvenile spinal muscular atrophy

Introduction
It was first described by Keizo Hirayama *et al*. in their study of Hirayama disease, which is also termed as non-progressive juvenile spinal muscular atrophy of the distal upper limbs, is a rare type of cervical myelopathy caused due to flexion movements of the neck. It is considered a benign motor neuron disorder with a stationary stage after a progressive course and it is a disease of young males between the ages of 15-25 years and is seen more commonly in Asian countries.

Material and Methods
This is a case series done on 6 patients with findings suggestive of Hirayama disease with proven or almost certain etiologies and were referred for MRI study to the Department of Radio-diagnosis in our hospital during the period of June 2020 to June 2021. All our patients were imaged in our institution. MRI was done using Siemens 1.5 T Magnetom Essenza MRI scanner. Patients were evaluated and appropriate imaging was done of the cervical spine in Neutral, Extension, Semi-flexion and Flexion positions. We evaluated the radiological findings giving importance to dynamic imaging to substantiate our findings.

Discussion
The etiopathogenesis of this disease remains unclear. The disease was first reported in 1959 by Hirayama *et al* [1]. This is a pure motor focal amyotrophy caused by the atrophy and the weakness at spinal segments C7, C8 and T1 innervated muscles due to dynamic compression in the spinal cord.

The clinical features of Hirayama disease are progressive muscular weakness of one or both hands and forearms without compromising the brachioradialis muscles [2].

(a) (b)

Fig 1: (A), (B) – 22 year old male patient with clinical diagnosis of Hirayama disease shows atrophic changes involving the thenar and hypothenar muscles of the hand, predominantly on the right side.

Patients presented with unilateral or asymmetrical bilateral atrophy of the upper limb thenar, hypothenar or forearm muscles and the sensory examination of these patients was normal. There was no significant past history of trauma or any chronic illness in these patients.

Imaging Features

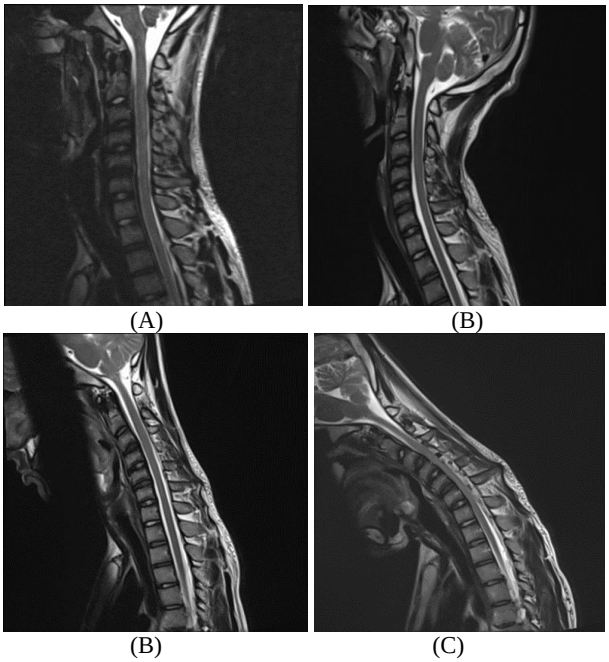

Fig 2: (A), (B), (C), (D) – 22-year-old male patient with clinical suspicion of Hirayama disease. (A) in neutral position (B) in extension, (C) in semi flexed position and (D) dynamic flexion image, progressing narrowing of spinal canal, widening of the posterior dural sac and compression on the cervical spinal cord.

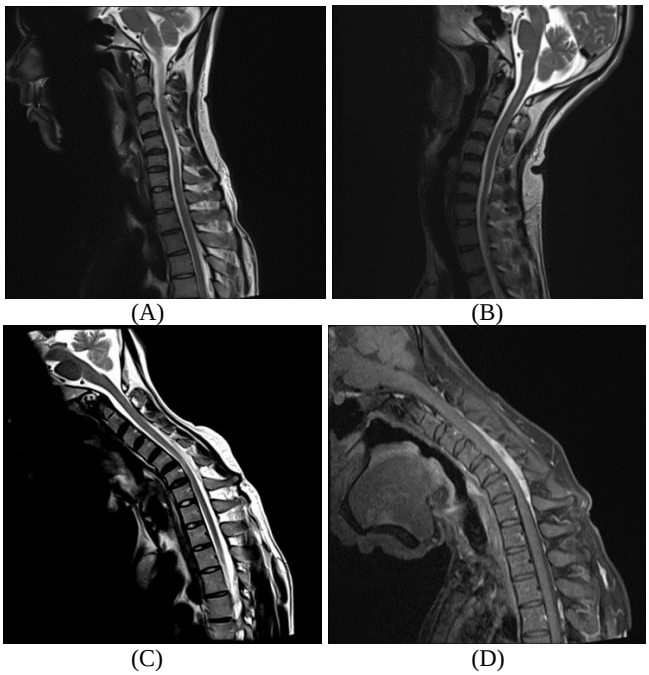

Fig 3: (A), (B), (C), (D),(E) – 25 year old male patient with clinical suspicion of Hirayama disease shows focal thinning of cervical spinal cord at C5-C6 level. (A) neutral position (B) in extension (C) flexion image (D) T1 Post contrast images show widening of the posterior dural sac enhancement of the posterior dural plexus. Focal thinning of the cervical cord is also seen at the region of compression.

The imaging modalities used to differentiate Hirayama disease from other causes of amyotrophy of distal upper limb included radiographs and dynamic magnetic resonance imaging (MRI) of cervical spine. The findings obtained radiographically were nonspecific and showed only loss of cervical lordosis. The better tissue visualization makes MRI the imaging modality of choice. Bede *et al*. [3] in a recent report described about a patient of Hirayama disease in whom there was segmental cord atrophy in neutral neck position in axial and sagittal views in the affected segment and there was normal architecture above and below the affected segments. "sand-watch" appearance on sagittal views was seen on the e focal flattening of the cord. high index of suspicion should be there on routine MRI of cervical spine in neutral position. Chen *et al*. [4] described another important imaging aspect in neutral position that showed loss of attachment between the posterior dural sac and subjacent lamina on axial MRI imaging in neutral position is the important finding in Hirayama disease diagnosis. Differential diagnoses of amyotrophy of distal upper limb muscles includes Amyotrophic lateral sclerosis, cervical cord tumor, myelopathy due to trauma or spondylosis, and syringomyelia. Since routine MRI of cervical spine is done only in neutral position, considerable degree of suspicion is necessary for ruling out Hirayama disease and a Dynamic imaging MRI cervical spine is of paramount importance. Among these patients, we performed T2 weighted (T2W), (Pre- and post-contrast) T1W-fat saturated in sagittal, axial planes, and 3D-CISS/FIESTA sequence; Raval *et al* [5] suggested the inclusion of -CISS/FIESTA sequence in flexion in their study done in 2010. Magnetic resonance imaging done in neutral position did not help in ruling out Hirayama disease. Neutral position MRI shows normal diameter of the spinal canal with no neural foraminal stenosis.

Conclusion

Hirayama disease is a rare self-limiting clinical condition presenting with weakness in the upper limb muscles. The diagnosis of this condition has to be done early as it is most commonly seen young age people in their second and third decades. Flexion studies are not routinely evaluated in a typical cervical MR spine study and hence the likelihood of missing this rare condition is high. As per our study, even semi flexed positions did not adequately demonstrate the positive imaging findings in Hirayama disease. Hence it is of utmost importance that any suspected case of Hirayama disease should be appropriately imaged in complete dynamic flexion. The usage of cervical collar has been useful as it stops the further progress of the disease. The use of hard cervical collar for preventing cervical flexion has shown to reduce the progress of the disease as a conservative management [6].

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