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A rare case of Hirayama disease

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Abstract
Introduction: Hirayama disease is a form of cervical myelopathy common in male adolescents that affects distal upper extremities & anterior horn cells of spinal cord. It is a symmetrical lower motor weakness that is self-contained in hands & forearms.
Materials and Methods: Observational type of case study using MRI. 16 years male presented with inability to move his right index finger & decreased sensations for 3 months
Results: T2 sagittal images showed mild atrophy with flattening of cervical cord & displacement of posterior dura with compression on cord on dynamic flexion. Cord diameter was reduced suggestive of compression.
Conclusion: Based on the patient's clinical, electrophysiological, & radiological characteristics, "Hirayama disease" was diagnosed. In addition to physiotherapy, a cervical collar was used to treat him in order to avoid neck flexion & thus surgery was avoided.

Keywords: Hirayama disease, MRI, cervical myelopathy, compression of cervical cord by dural sac, necrosis of anterior horns of lower cervical cord, chronic microcirculatory alterations in area of anterior spinal artery by prolonged flexion

Introduction
Hirayama disease is a form of cervical myelopathy common in male adolescents that affects distal upper extremities & anterior horn cells of spinal cord. It is a symmetrical lower motor weakness that is self-contained in hands & forearms [1]. Asymmetric compression of cervical cord by dural sac is considered cause of this disorder [2]. Hallmark pathology being necrosis of anterior horns of lower cervical cord, is caused by chronic microcirculatory alterations in area of anterior spinal artery brought on by repeated or prolonged flexion[3]. Aim of this case report is to describe role of magnetic resonance imaging (MRI) as an effective diagnostic imaging tool in evaluation of Hirayama disease for early & effective management.

Materials and Methods
Observational type of case study using MRI.16 years male presented with inability to move his right index finger & decreased sensations for 3 months. There was no history of trauma, prior hospitalization or any other comorbidities. A clinical diagnosis of Hirayama disease was suspected & patient advised MRI examination of cervical spine.

Results
T2 sagittal images showed mild atrophy with flattening of cervical cord at C4-C5 levels (figure 1).T2 sagittal dynamic flexion study showed ventral displacement of posterior dura with compression on cord & attenuated ventral adjacent subarachnoid spaces from C4 lower end to lower end of C6 (figure 2).Cord diameter on sagittal section at level of C5 was 0.72 cm on neutral position (figure 3) & 0.5 cm on flexion (figure 4) there by suggestive of reduced diameter due to compression. Crescenteric area seen on flexion at the same site posteriorly.

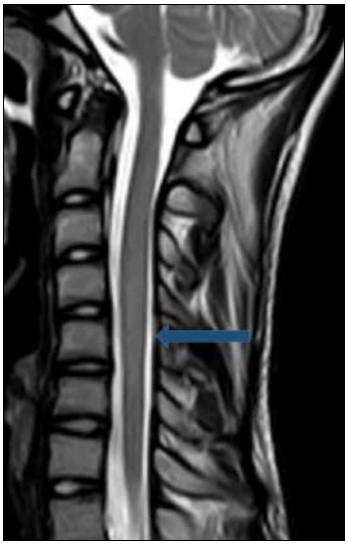

Fig 1: T2 sagittal images showed mild atrophy with flattening of cervical cord at C4-C5 levels

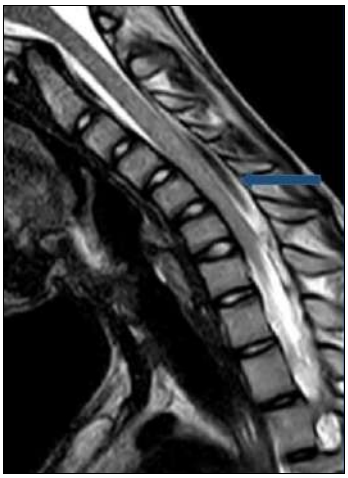

Fig 2: T2 sagittal dynamic flexion study showed ventral displacement of posterior dura with compression on cord & attenuated ventral adjacent subarachnoid spaces from C4 lower end to lower end of C6

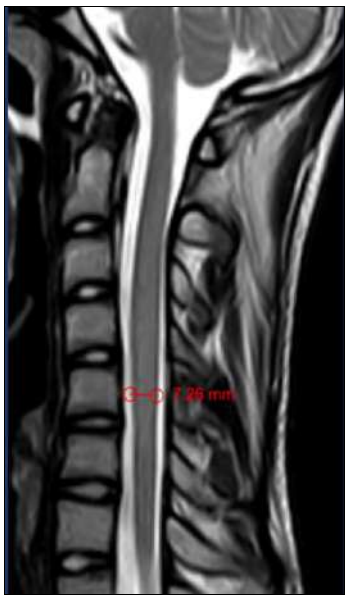

Fig 3: Cord diameter on sagittal section at level of C5 was 0.72 cm on neutral position

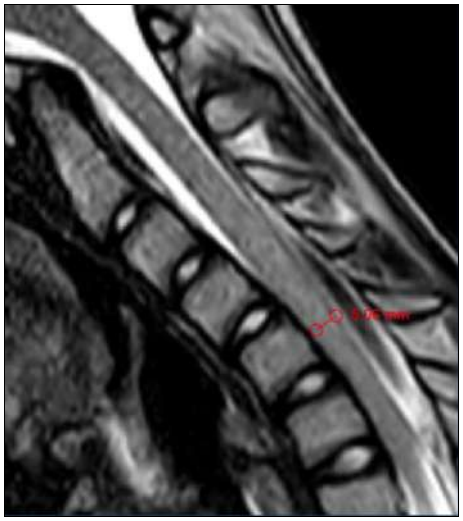

Fig 4: Cord diameter was 0.5 cm on flexion there by suggestive of reduced diameter due to compression. Crescenteric area seen on flexion at the same site posteriorly.

Discussion

It is evident that MRI with flexion study can effectively depict compression with displacement of cervical cord responsible for this pathology. An early diagnosis allows cervical collars to be used to manage this condition & reduce need for lengthy surgical procedures [4].

Conclusion

Based on the patient's clinical, electrophysiological, & radiological characteristics, "Hirayama disease" was diagnosed. In addition to physiotherapy, a cervical collar was used to treat him in order to avoid neck flexion & thus surgery was avoided.

Acknowledgments

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Conflict of Interest

Not available

Financial Support

Not available

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