

Hirayama Disease-Atypical Presentation

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Section: Neuroradiology

Area of Interest: Neuroradiology spine

Procedure: Imaging sequences

Imaging Technique: MR

Special Focus: Pathology Case Type: Clinical Cases

Authors: DR PRIYANK S CHATRA, DR VARUN BABU

Patient: 36 years, male

Clinical History:

A 36-year-old male patient presents to the clinician with neck pain & right upper limb weakness which has increased over the last few years. Neutral MRI reported as degenerative discs. EMG showed denervation patterns in multiple small muscles of the right hand localized to C5-T2 dermatomes, then a flexion MRI was performed.

Imaging Findings:

On neutral MRI, degenerative disc disease is seen with myelomalacia changes with focal cord atrophy at C5/6 (Figure -1, 2). On flexion MRI, there is anterior shift of the posterior dura extending from C4 to D2 (Figure-3, 4). Maximum indentation was noted at the level of C5 on the right side (Figure-5). In addition, there was atrophy and thinning of the cervical cord at this level. Multiple flow voids were seen posterior to the dura on T2W images (Figure-3). On post contrast study, there is enhancement in the venous plexus posterior to the displaced dura (Figure-6). Findings representing Hirayama disease.

Discussion:

Hirayama disease was first reported by Hirayama and colleagues in 1959 in Japan. Since then multiple cases have been reported in the Indian subcontinent [1]. Hirayama disease usually involves young adults in the age group of 15 to 25 years with characteristic male predominance. Although the disease might have started in adolescence, the presenting age can range from 15- 41 years [2].

As flexion MRI is not performed on all cervical spine studies, identifying the abnormality on neutral position becomes more important. Several signs have been described in the literature regarding findings seen on neutral position, which should alert the radiologist to perform a flexion MRI. Loss of attachment (LOA) of the posterior dura to the lamina is described by many authors as the most accurate finding on neutral MRI [2]. Other findings include abnormal cervical curvature, focal localized cord atrophy, and non-compressive intramedullary T2W high signal representing myelomalacia [3]. Loss of attachment (LOA) was however not seen in this patient and is likely to go unnoticed to an untrained eye. Other findings like focal cord atrophy and loss of cervical lordosis and focal intramedullary T2W hyperintense signal was attributed to degenerative disc on the neutral examination.

Dynamic MRI is the only way to differentiate between these conditions when the clinical examination is inadequate or inconclusive. Our patient was given a cervical collar and was followed up every 6 months. There was no further

progression of the symptoms after 2 follow-ups. Definitive treatment in the form of cervical decompression with fusion is reserved for non-responsive cases.

Strong clinical suspicion in a young male patient with disproportionate atrophy of the cord should alert the radiologist and alter the examination in favour of a flexion study.

Differential Diagnosis List: HIRAYAMA DISEASE, DEGENERATIVE DISC DISEASE, MYELOMALACIA

Final Diagnosis: HIRAYAMA DISEASE

References:

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- Lehman VT, Luetmer PH, Sorenson EJ, Carter RE, Gupta V, Fletcher GP, Hu LS, Kotsenas AL (2013) Cervical Spine MR Imaging Findings of Patients with Hirayama Disease in North America: A Multisite Study. Am J Neuroradiol 34:451-456 (PMID: [22878010](#))
- Kyoung Hee Lee, Dae Seob Choi, Young Suk Lee, Dong Ho Kang (2016) Clinical Experiences of Uncommon Motor Neuron Disease: Hirayama Disease. Korean J Spine 13(3): 170–172.

Figure 1

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Description: Figure 1: T1W sagittal image showing focal cord atrophy at C5-6 level (arrow). **Origin:** Department of Radiology Little flower hospital Angamaly, Kerala, India

Figure 2

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Description: T2W sagittal image showing degenerative discs at multiple levels with focal cord atrophy at C5-6 (arrow) and cord signal changes extending from C4 to C6 level. **Origin:** Department of Radiology Little Flower Hospital Angamaly, Kerala, India

Figure 3

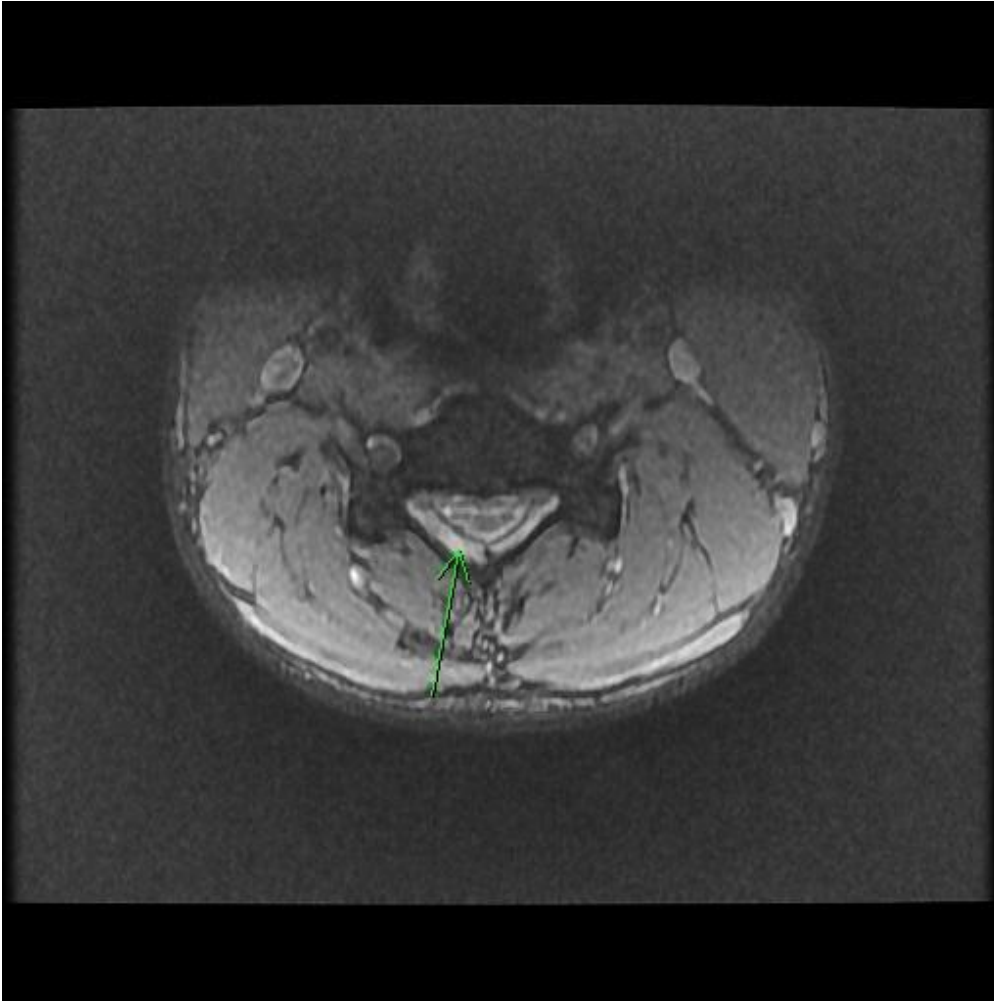
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Description: T2W sagittal flexion image showing anterior indentation of the posterior dura with flow voids (arrow) in the displaced extradural compartment. **Origin:** Department of Radiology Little Flower Hospital Angamaly, Kerala, India

Figure 4

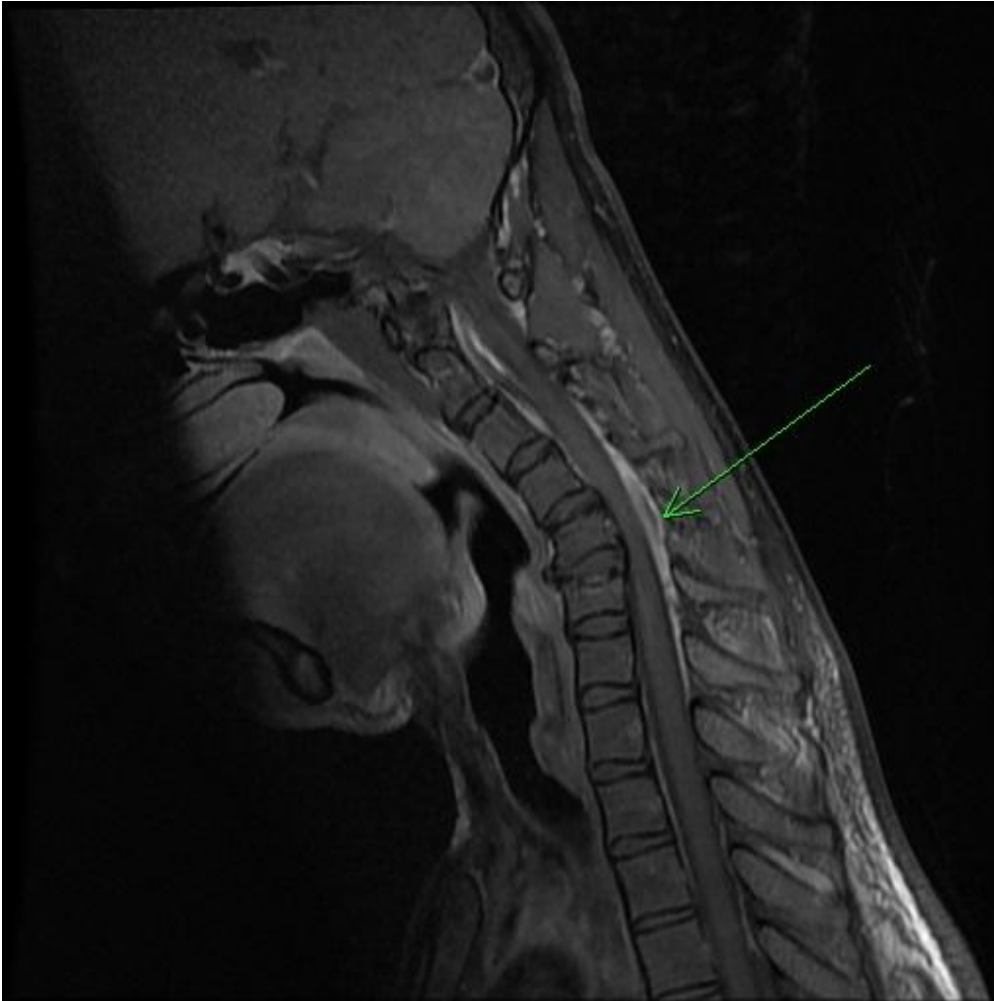
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Description: GRE axial flexion image showing anteriorly displaced dura more prominent on the right side (arrow). **Origin:** Department of Radiology Little Flower Hospital Angamaly, Kerala, India

Figure 5

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Description: Post contrast T1W FS sagittal flexion image showing enhancement of the engorged venous plexus posterior to the displaced dura (arrow). **Origin:** Department of Radiology Little flower Hospital Angamaly, Kerala, India