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Case Report

Radiology Section

Unique Manifestation of Hirayama Disease: Bilateral Radial and Ulnar Nerve Palsy with Hypothenar Atrophy, and Atypical Snake Eye Appearance

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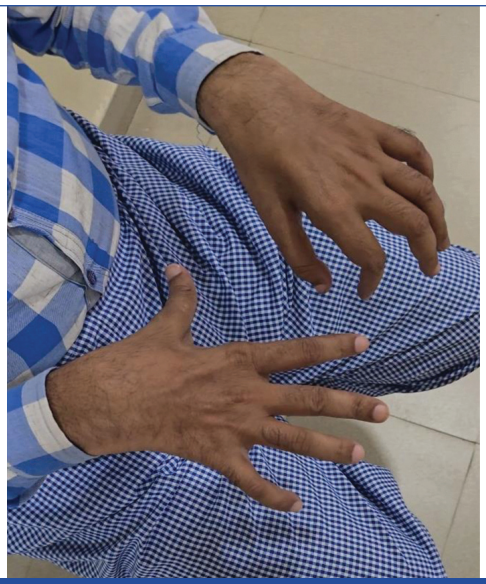
ABSTRACT

A rare cervical myelopathy called Hirayama disease usually manifests as muscle atrophy and distal upper limb weakness that develops gradually in young males. A 42-year-old male presented with progressive weakness and atrophy in both upper limbs for over 12 years, accompanied by fine tremors. Neurological examination revealed weakness in wrist extension and finger movements, profound atrophy of the hypothenar and interosseous muscles, and sensory deficits consistent with radial and ulnar nerve involvement. Cervical spine Magnetic Resonance Imaging (MRI) in a flexed position revealed anterior displacement of the posterior dural sac (from C3 to C7 level), with compression of the lower cervical cord, confirming the diagnosis of Hirayama disease. This case highlights a rare and severe manifestation of Hirayama disease, emphasising the bilateral involvement of multiple peripheral nerves, a finding uncommon in classical presentations. Early diagnosis, aided by dynamic MRI imaging, is crucial to prevent further neurological deterioration. Treatment options, including cervical collar immobilisation, aim to halt disease progression. This report underscores the importance of considering Hirayama disease in the differential diagnosis of atypical peripheral neuropathy presentations, particularly in young patients and, in this case, in an older age group (40-45 years). It also provides insight into the varied clinical spectrum of this condition.

Keywords: Anterior displacement of posterior dural sac, Cervical cord thinning, Claw hand, Radial nerve palsy, Ulnar nerve palsy, Wrist drop

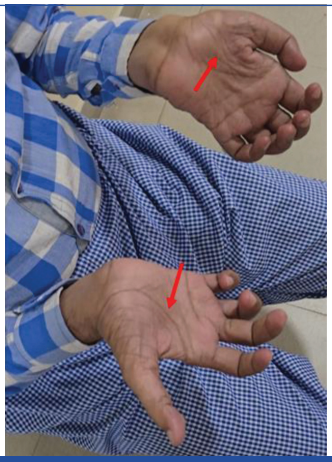
CASE REPORT

A 42-year-old male patient came to the neurology department with complaints of bilateral upper limb weakness that has been gradually progressive for the last 12 years. The patient is a farmer by occupation, and there is no history of trauma. Initially, there was weakness in the right upper limb, which progressed to the left upper limb. Initially, the patient did not encounter any difficulty in his work. Later, the condition worsened, and gradually he presented with tremors in the hand (as shown in [Video]) and subsequently developed wrist drop [Table/Fig-1], claw hands [Table/Fig-2], and atrophy of the hypothenar muscles [Table/Fig-3], which hampered his daily life activities. No gait abnormalities were present.



[Table/Fig-1]: Depicting bilateral wrist drop- radial nerve palsy.

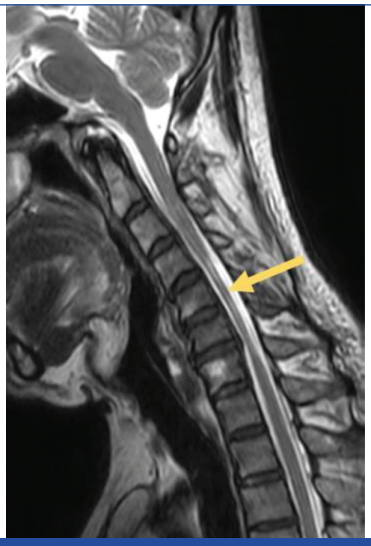
[Table/Fig-2]: Depicting bilateral claw hands- ulnar nerve palsy.



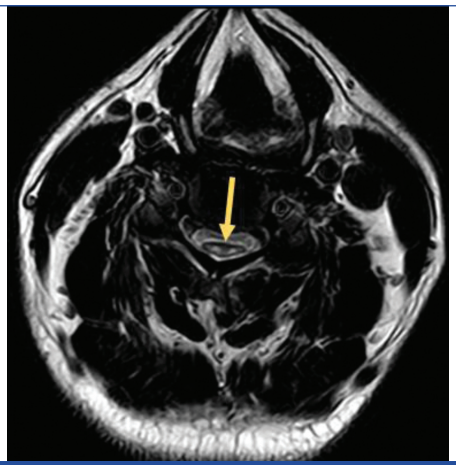
[Table/Fig-3]: Bilateral hypothenar eminence muscles atrophy (red arrow).

On general examination, the patient's vital signs were stable. Following the laboratory work-up, the following information was discovered: White Blood Cells (WBC) 7,600/mm^3, platelets 250,000/mm^3, and haemoglobin 14.2 g/dL (all within normal ranges). The Erythrocyte Sedimentation Rate (ESR) was 12 mm/hr; serum electrolytes showed calcium (Ca^{2+}) at 9.3 mg/dL, magnesium (Mg^{2+}) at 2.0 mg/dL, potassium (K^+) at 4.1 mEq/L, and sodium (Na^+) at 140 mEq/L (all within normal range); Alanine Aminotransferase (ALT): 22 U/L, Aspartate Aminotransferase (AST): 24 U/L, and Alkaline Phosphatase (ALP): 75 U/L (normal) for Liver Function Tests (LFTs); urea: 28 mg/dL, creatinine: 0.9 mg/dL (normal) for Renal Function Tests (RFTs); Thyroid function tests showed Thyroid Stimulating Hormone (TSH) at 2.1 µIU/mL, T4 at 7.8 µg/dL, and T3 at 1.2 ng/mL (all within normal ranges); Vitamin B_{12} was 450 pg/mL (normal); serum copper: 110 µg/dL; serum ceruloplasmin: 25 mg/dL; Fasting Blood Sugar (FBS): 92 mg/dL; and HbA1c: 5.4% (normal).

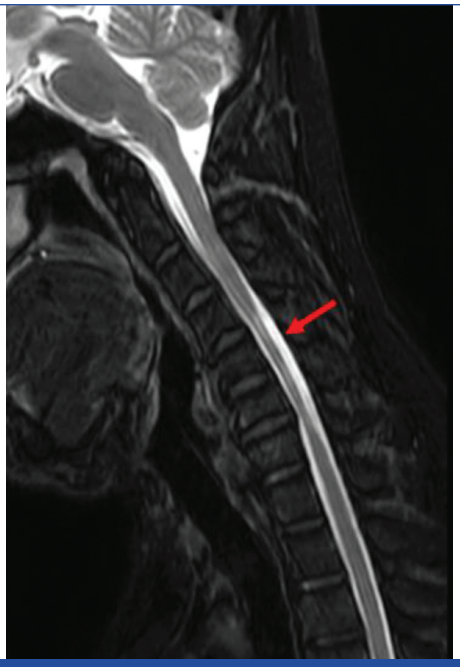
The patient underwent an MRI of the cervical spine, which revealed thinning of the spinal cord with T2 hyperintensity noted in the spinal cord extending from the lower endplate of C4 to the lower endplate of C7 vertebra [Table/Fig-4], with a Snake Eye Appearance (SEA) [Table/Fig-5], suggestive of myelomalacia. In the flexed position, there is anterior displacement of the dura from the C3 to C7 vertebra with an expanded posterior epidural space [Table/Fig-6]. Straightening of the cervical spine was also noted [Table/Fig-7]. A lipohaemangioma was noted in the body of the C7 vertebra with a Schmorl's node at the upper endplate [Table/Fig-7]. The radiological findings are consistent with the diagnosis of Hirayama disease, along with mild degenerative changes of the cervical spine.



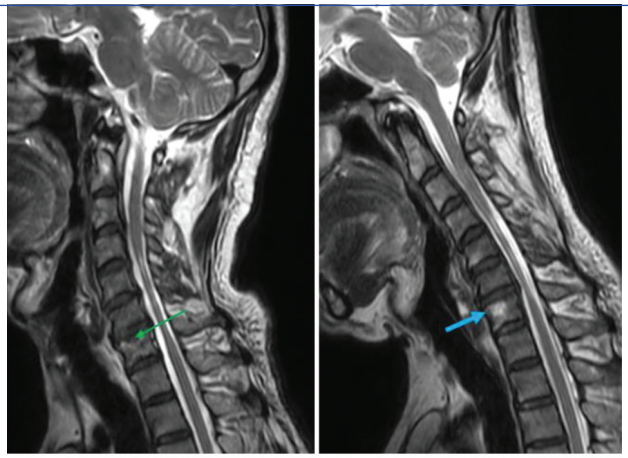
[Table/Fig-4]: Sagittal T2WI- in flexed position shows thinning of spinal cord with T2WI hyperintensity noted in spinal cord extending from lower endplate of C4 to lower endplate of C7 vertebra (yellow arrow).



[Table/Fig-5]: Axial T2WI diffuse disc bulge indenting over anterior thecal sac causing spinal canal narrowing there is T2 hyperintensity noted in spinal cord showing characteristic SEA s/o compressive myelopathy.



[Table/Fig-6]: Sagittal- T2FSWI In the flexed position, there is anterior displacement of the dura from C3 to C7 vertebra with expanded posterior epidural space.



[Table/Fig-7]: Sagittal T2WI- Lipohaemangioma (blue arrow) noted in body of C7 vertebra with Schmorls node (green arrow) noted at upper-end plate also cervical spine straightening noted (a- normal position).

The patient was later managed conservatively with a cervical collar for immobilisation and was counseled about the progression of the disease, as well as the need for surgical management in the event of a worsened condition. This may include multilevel cervical fixation, which involves fixation of the atlantoaxial joint and the facetal fixation method, and could be performed in the future.

DISCUSSION

Hirayama disease, also called Juvenile Muscular Atrophy (JMA), is a very uncommon form of cervical myelopathy that affects young individuals and leads to motor dysfunction of the C7 to T1 myotomes [1,2]. Hirayama disease, sometimes referred to as oblique amyotrophy or JMA of Distal Upper Extremity (JMADUE), is a rare neurological condition that primarily affects young boys between the ages of 20 and 30. It results in progressively worsening upper limb weakness, tremors, and fasciculations, often in an asymmetrical manner. The disorder leads to dynamic compression during neck flexion, caused by a misalignment between the spinal cord and the vertebral column. Flexion-extension MRI is essential for early diagnosis and successful treatment [3].

As evidenced by severe presentations, bilateral-symmetrical involvement in Hirayama disease may be regarded as a severe form, even though the diagnosis remains clear [4]. The pathology of the cervical spine, which may include kyphosis or loss of normal

curvature, is evident on MRIs. It is associated with anteroposterior cord flattening and C5-C6 cord atrophy, both of which worsen with flexion. Posteriorly, disc protrusion is often observed, and there may be anterior signal intensity changes in the cord. Lastly, the dural sac's contact with the spinal laminae is not effaced posteriorly [5].

The lower cervical cord is compressed during cervical flexion, which causes the posterior dural sac vertebral wall to move anteriorly, resulting in varicose veins and an increase in the posterior epidural space. A cervical flexion range of 30 to 40 degrees is advised [6].

The "SEA" is a reversible condition, but it has an adverse prognosis in cases of degenerative cervical myelopathy. A 2020 case series found that SEA is a poor surgical prognostic marker in 22-28% of patients. However, the baseline neurological condition remains crucial for predicting patient outcomes [7].

The differentials considered for our case scenario included compressive myelopathy, which was ruled out as there was no disc bulge at that level causing compressive myelopathy; it was causing multilevel cord changes. Hirayama disease and Amyotrophic Lateral Sclerosis (ALS) were also considered. ALS was ruled out as the patient does not exhibit any alterations in behaviour, hyperreflexia, bulbar signs (dysphagia and slurring of speech), or gait abnormalities.

The disorder is characterised by dynamic alterations in the cervical spinal cord during neck flexion, localised spinal cord atrophy, and an insidious onset. During neck flexion, imaging investigations show compression of the spinal cord and anterior displacement of the posterior dura mater. Early diagnosis is essential to prevent the condition from worsening. In addition to cervical collar treatment, surgical procedures such as spinal fusion or duraplasty may be necessary in extreme cases [8].

CONCLUSION(S)

This report emphasises the importance of considering Hirayama disease in patients presenting with atypical peripheral neuropathy, even in older age groups. Early diagnosis and appropriate management can significantly improve outcomes in severe cases. In our patient, the atypical presentations of claw hand and wrist drop warrant further investigation and analysis to better understand their pathophysiology and implications.

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