International Journal of Science and Research (IJSR)
ISSN: 2319-7064
SJIF (2022): 7.942

Hirayama's Disease: A Radiological Camouflage

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Abstract: ***Hirayama's disease is a rare disorder. It is also referred as juvenile non progressive amyotrophy, Sobue disease. It is a lower motor neuron type of disease. It commonly affects young males in their second and third decades. It is seen most commonly in Asian countries like India. In majority of the cases cause of this disease is unknown. We reported a case of Hirayama's disease in 16 year old male patient who presented with the complaints of progressive weakness in bilateral upper limbs since 5 months. On MRI cervical spine, it revealed the cardinal features of Hirayama disease.***

Keywords: Hirayama's disease

1. Introduction

Hirayama's disease is a very rare and benign disorder. It is also referred as Juvenile non progressive amyotrophy, Sobue disease. It is an untreatable lower motor neuron type of disorder. It affects mainly the young males in their second and third decade of life. It is seen most commonly in Asian countries like India. There is insidious in onset, gradually progressive muscular dystrophy of distal part of the upper limbs due to flexion movements of the cervical spine. The etiopathogenesis behind this disease is imbalance between the growth of vertebral column and spinal canal contents leading to abutment of spinal cord on the vertebrae anteriorly and detachment of posterior dura.Due to repeated flexion there is chronic microcirculatory changes in the territory of anterior spinal artery and at last leading to necrosis of anterior horn cells.

2. Case Report

16 year old male patient presented with the complaint of progressive weakness in the distal part of B/l upper limbs with atrophy of B/l thenar muscles and muscles of the forearm. He did not have any pain, loss of sensation, diplopia, dysphagia, ptosis, muscle cramps, fasciculations, headache or neck pain. There was no history of trauma, febrile illness, poliomyelitis or exposure to toxins or heavy metals in the past. There was no family history of similar complaints or neuromuscular disease.

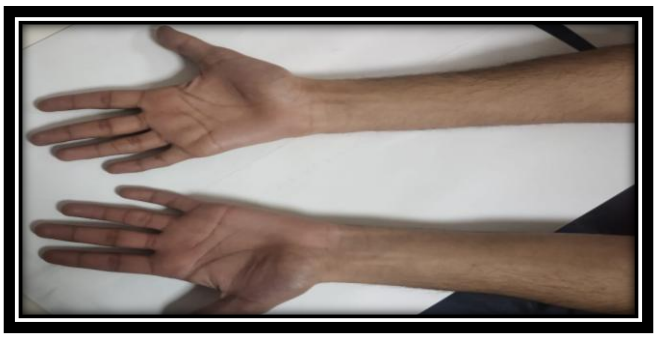

Figure 1: Atrophy of B/L thenar and forearm muscles.

MRI cervical spine was requested. There was loss of cervical lordosis with atrophy of spinal cord in the cervical region (predominately at C6-C7 level).On the basis of clinical history and findings n the initial MRI scan of cervical spine in neutral position ,MRI cervical spine in flexion position was acquired. On flexion, there was increase in the laminodural space, engorgement of vertebral venous plexus and anterior displacement of cord. So, diagnosis of Hirayama's disease was reached based on the clinical scenario and imaging findings.

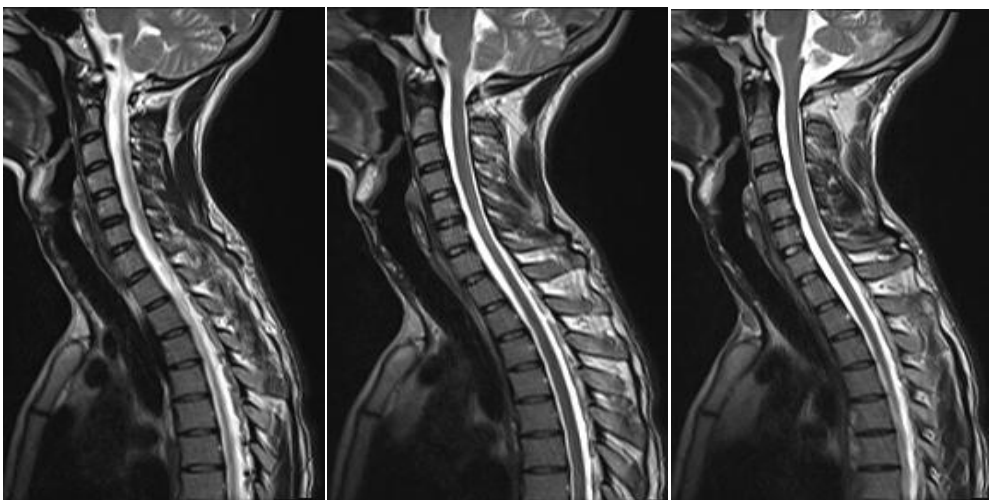

Figure 2: Cord atrophy at lower cervical region (predominately at C6-C7 vertebral level)

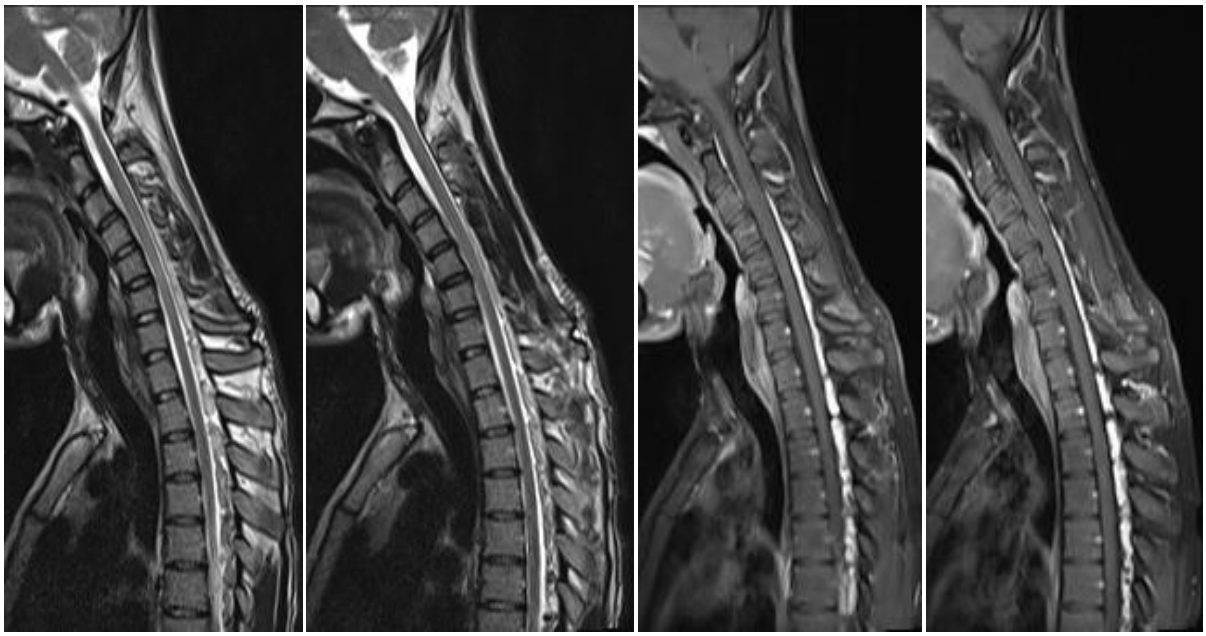

Figure 3: On Flexion. There is increase in laminodural space with engorgement of vertebral venous plexus and anterior displacement of spinal cord.

3. Discussion

There is focal amyotrophy with unilateral or asymmetric bilateral weakness and wasting of muscles innervated by C7, C8, and T1. It is insidious in onset, gradually progressive, chronic, self-limiting disorder. It has male preponderance, seen between the ages of 15 and 25 years. The lesions are found only in the anterior horns of the spinal cord from C-5 to T-1, particularly marked at C-7 and C-8. Most commonly seen in Japan and other Asian countries like India and Malaysia [7]. The probable causes suggest an imbalanced growth between the vertebral column and spinal canal contents. This imbalanced growth will cause disproportional length between the vertebral column and the spinal canal contents leading to tight dural sac/overstretching of the cord in the neutral position and anteriorly displaced posterior dural wall when the neck is flexed.

MRI shows atrophy of the lower cervical cord in a neutral position, loss of cervical lordosis in neutral position and loss of attachment between the posterior dural sac and subjacent lamina, engorgement of vertebral venous plexuses in flexion MRI of cervical spine.

4. Conclusion

Early diagnosis is necessary in the cases of Hirayama's disease to prevent the progression of the disease. Use of cervical collar can prevent the neck flexion and halt the progression of the disease. Diagnosis of Hirayama disease needs a high index of suspicion by radiologist in order to acquire flexion MRI of cervical spine. Therefore familiarity with this disorder is vital to diagnose the disorder as earliest as possible.

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