http://jmscr.igmpublication.org/home/
ISSN (e)-2347-176x ISSN (p) 2455-0450
crossref DOI: https://dx.doi.org/10.18535/jmscr/v11i4.17

IGM Publication
Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

Monomelic Amyotrophy - A Rare Case Report

Authors
Dr M. Karuna Sagar[1], Dr Ramchandra Prasad. K[2], Dr Katturoju Himalay[3], Dr Syed Imran[4*]
[1]Professor and Head of the Department of General Medicine, Mediciti Institute of Medical Sciences, Ghanpur, Telangana
[2]Assistant Professor Department of General Medicine, Mediciti Institute of Medical Sciences, Ghanpur, Telangana
[3]Senior Resident Department of General Medicine, Mediciti Institute of Medical Sciences, Ghanpur, Telangana
[4]Third Year Post Graduate Department of General Medicine, Mediciti Institute of Medical Sciences, Ghanpur, Telangana
*Corresponding Author
Dr Syed Imran

Abstract
Monomelic amyotrophy (MMA),is a rare benign disorder, also referred to as Hirayama's disease Juvenile non progressive amyotrophy, Sobue disease. It is a focal, lower motor neuron type of disease. Mainly young males in their second and third decades of age are most commonly affected. In majority of people cause of this disease is unknown. MRI of cervical spine in flexion will reveal the cardinal features of Hirayama disease.

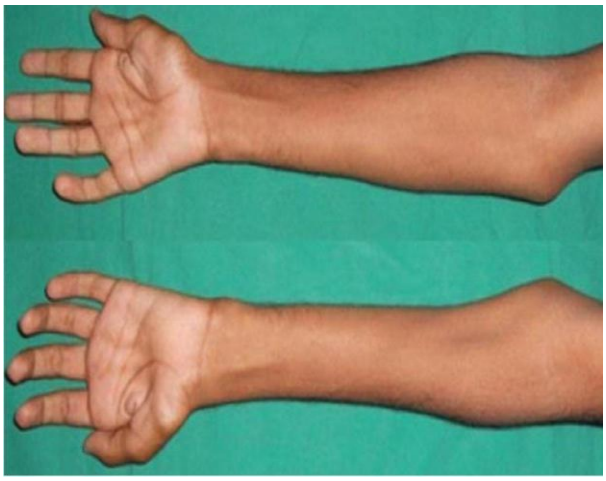
Case Report

This is a case of 28-year gentleman who is a software employee came with a history of insidious onset of weakness in both the hands, left side followed by right of 4 year duration. He noticed that the weakness in the left hand muscles which was gradually progressed to the fore arm muscles. Within 6 months he noticed similar complaints in the right hand also, which was progressed gradually to the fore arm muscles. He had also noticed atrophy of muscles of hand and fore arm which was gradually progressive in nature. 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.

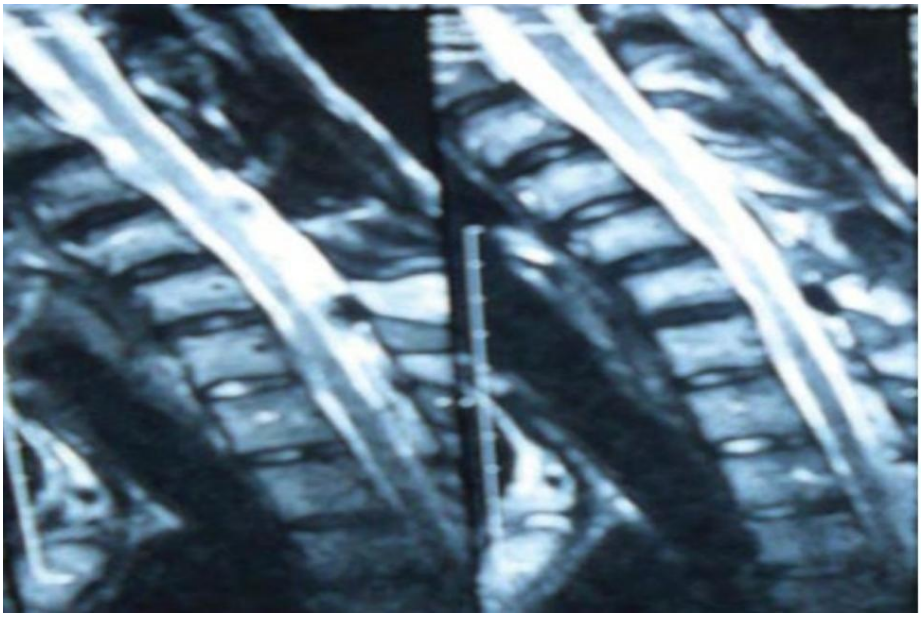
On physical examination he was conscious oriented to time, place and person. His cranial nerves and sensory examination was normal. Motor examination showed bilateral complete clawing of fingers with gross atrophy and weakness of thenar, hypothenar, inter osseous, fore arm muscles, biceps, triceps and deltoid muscle of both upper limbs except Brachioradialis, which was spared on both the

sides. Deep tendon reflexes were not exaggerated. Co ordination and gait were normal.

Blood routine investigations, blood urea, serum creatinine, serum electrolytes, liver function tests, thyroid function tests, erythrocyte sedimentation rate and creatine phosphokinase were within normal limits. Plain cervical spine X - ray was normal except for loss of cervical lordosis. Electromyogram showed evidence of denervation in the form of fibrillation and fasciculations in C7 and C8 distribution in both upper limb muscles. Left side was more predominant compared to right. Both Brachioradialis were spared. There was no involvement of the lower limb muscles.MRI of the cervical spine in neutral position showed thinning of cervical cord from C4 to C7 level, suggestive of cord atrophy. There was mild irregularity of the cord at C5-C6 level. On forward flexion of the cervical spine, there was anterior displacement of the posterior dura from C4 to T1 levels with maximal shift at C6 and C7.



MRI in Flexion



Based on the clinical features and the characteristic findings on flexion MR images diagnosis of Monomyelic Amyotrophy was made. Patient was put on a hard cervical collar to prevent neck flexion and followed up clinically at regular intervals. At the end of 12-month follow-up the patient was doing well, with no further progression of symptoms.

Discussion

Hirayama disease is characterised by focal amyotrophy with unilateral or asymmetric bilateral weakness and wasting of muscles innervated by C7, C8, and T1. It's an insidious onset, chronic, often self-limiting disorder with male preponderance, seen between the ages of 15 and 25 years. The pathogenesis of the disorder is unknown – probable causes suggest that an imbalanced growth between the patient's vertebral column and spinal canal contents. This imbalanced growth will cause disproportional length between the patient's vertebral column and the spinal canal contents, which will cause a “tight dural sac” or “overstretch of the cord” in the neutral position and an anteriorly displaced posterior dural wall when the neck is flexed.

Myelography may show the forward movement of the posterior dural wall when the neck is flexed. MRI studies in neck flexion, which are easy to obtain, will show the forward displacement of the posterior wall and a well-enhanced crescent-shaped mass in the posterior epidural space of the lower cervical canal. Diagnosis of Hirayama disease is mainly based on flexion MRI of cervical spine. Asymmetry is one of the most characteristic findings of this disease, both clinically and radiologically.

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