

# Bimelic Symmetric Hirayama's Disease: Outgrowing its 'Brachial Monomelic Amyotrophy' Origins

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## Description

A 18 year old boy presented to us with history of insidious onset, gradually progressive weakness of bilateral upper limbs which started 2 years back but had been static for the past one year. He had difficulty in buttoning his shirt and holding a pen. There was no difficulty in raising his arms while wearing his clothes. He also noticed gradual thinning of both his hands and distal part of forearm near his wrist. There was no history suggestive of lower limb, trunk or cranial nerve involvement. There was no sensory loss and no bowel bladder involvement. There was history of pain over neck on prolonged flexion. There was no history of similar disease in family. On examination bilateral wasting of small muscles of hand and muscles over the medial aspect of forearm. Tone was reduced in bilateral wrist and elbow joints. Bilateral biceps, triceps and supinator jerks were absent. All other central nervous system examination was within normal limits. Muscle enzymes CPK and LDH were normal. NCV of upper limbs showed no response in bilateral ulnar and median nerves. EMG showed denervation potentials and fasciculations. MRI cervical spine showed loss of dural attachment posteriorly with increase in posterior epidural space and engorged venous plexuses at lower cervical and upper dorsal spine. There was antero-posterior flattening of spinal cord from C5 to D3 segment. Based on the clinical presentation and MRI findings a diagnosis of bimelic symmetric Hirayama's disease was made. Patient was prescribed a cervical collar. At 6 months follow-up patient showed no progression of weakness or atrophy. Hirayama's disease is a benign non-progressive illness affecting young males causing lower motor neuron type of upper limb weakness without sensory involvement due to forward displacement of the posterior wall of the lower

cervical dural canal during neck flexion [1]. The disease was thought to be unilateral or asymmetric in onset if bilateral leading to it being called brachial monomelic amyotrophy. But it is recognised now that up to 10% of patients may have bilateral symmetric involvement [2]. Syringomyelia, spinal cord tumors, multifocal motor neuropathy, and toxic neuropathies are close differentials in cases with bilateral symmetric onset. Early diagnosis and use of a cervical collar prevents progression of disability in Hirayama's disease [3].

## Learning Points

1. Hirayama's disease can present with bilateral symmetric onset of upper limb involvement.
2. A dynamic contrast enhanced MRI of cervical spine showing loss of posterior dural attachment, anterior dural shift with flexion and antero-posterior flattening of lower cervical and upper thoracic spinal cord is diagnostic of Hirayama's disease.
3. Early diagnosis and use of a cervical collar prevents progression of motor weakness.

## References

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