

## **A LATE ONSET OF HIRAYAMA DISEASE WITH LATE PROGRESSION IN A MALAYSIAN MAN: CASE REPORT**

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**Introduction:** Hirayama disease, also known as monomelic amyotrophy (MMA), is a rare cervical myelopathy that manifests between the age of 15-25 years, in asymmetrical pattern, mainly causing atrophic weakness of forearms and hands. Forward displacement of dural sac during neck flexion causing ischemic changes over the anterior part of spinal cord has been postulated to be its pathology. Disease progresses first few years of onset, then stabilises in 3-5 years

**Discussion:** A 54 years old gentleman, presented 6 years prior, with history of right index, middle and ring finger weakness, occurring spontaneously with no history of trauma. Clinical assessment showed right hand thenar muscle atrophy with finger drop of the index, middle and ring finger. Initially, C7 and C8 power was grade 4, T1 was grade 2 over right side. Scapulohumeral reflex was positive both sides. Otherwise, there was no cervical tenderness and sensation was intact. MRI Cervical revealed multilevel prolapse intervertebral disc with narrowed neural foramen at level of C6/C7. This did not explain the cause of the finger drop, therefore, nerve conduction studies done, revealing diffuse upper limb chronic neurogenic myotomal involvement severely affecting the right C7 myotome. Patient was then referred to neuromedical team and sent for physiotherapy. Repeated MRI cervical after 2 years revealed cervical spondylosis with C5/C6 myelomalacia. Disease did not progress until this year, 6 years after onset, where his weakness progressed to all fingers. His power dropped to grade 3 at C7 and C8 level. Patient was advised to avoid neck flexion and physiotherapy continued to avoid further muscle weakness due to disuse atrophy

**Conclusion:** Hirayama Disease onset can be at a later age. Therefore, it should not be excluded in the older age group. Long term follow up is also crucial as course of the disease may progress longer.