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# Hirayama disease: A case of an Asian male with asymmetric bilateral amyotrophy of upper limbs

Jun Wang; Tingting Shen; Qianchang Wu; Ping Ni; Weifeng Luo*

***Corresponding Author: Weifeng Luo**

Department of Neurology and Clinical Research Center of Neurological Disease, The Second Affiliated Hospital of Soochow University, Suzhou 215004, China

Email: lwfwxx@126.com

## Abstract

**Introduction:** Hirayama disease (HD), also known as monomelic atrophy, is a rare clinical condition with preference for Asian males [1]. It usually involves the unilateral upper extremity. Here, we present a case of asymmetric bilateral muscular atrophy and explore the cause.

**Methods:** The patient underwent medical history, clinical investigation, neuroimaging and electromyogram.

**Results:** The weakness and hyposthenia of upper limbs are observed. MRI images of cervical flexion showed anterior displacement of the posterior dural sac. Electrophysiological examination showed the magnitude of the compound motor action potentials (CMAPs) is decreased.

**Discussion:** Hirayama disease should be highly considered in patients who have an insidious onset in adolescence and a slowly progressive course. Weakness and muscular atrophy of upper limbs are frequent [2].

**Keywords:** Hirayama disease; Hyposthenia; Cervical myelopathy; Monomelic atrophy; Muscular atrophy.

## Case Report

MJH is an 18-years-old Chinese male. He had a wholesome childhood and has no mental illness. Computer games were his favorite. He has no family history of genetic disorders either. 3 years ago, he began to have hyposthenia of left upper limb. The manifestation was that he gradually had difficulty extending his left thumb. Dorsal flexion of the fingers was also problematic. 1 year ago, weakness of the left upper extremity got worse and passed to the right, mainly manifested as hyposthenia of the right forearm. Hypotrophy of palmar and thenar muscles bilaterally begun to appear, more evident on the left. Besides, his weakness got worse on exposure to cold environment, and fasciculation occurred when the affected

muscles contracted, limiting his daily activities.

Physical examination of the patient's upper limbs showed asymmetric bilateral amyotrophy involving thenar muscles, hypothenar muscles, interossei muscles, wrist flexors and extensors muscles (Figure 1). The atrophy of muscles was worse on his left side. Other symptoms such as cranial nerve signs, pyramidal signs, abnormal reflexes and sensory disturbance are not observed. Besides, his laboratory examination was normal as well.

MRI in a neutral cervical position revealed the loss of normal cervical lordosis and an antero-posterior flattening of the spinal cord between C5 and C6. In MRI of cervical flexion, it showed anterior displacement of the posterior dura and high signal intensity between C5 and C6 (Figure 2). In his electrophysiological examination, nerve conduction velocities were normal but it showed attenuation of CMAPs of bilateral ulnar nerve. In needle electromyography, the denervation of the atrophied muscles was notable.

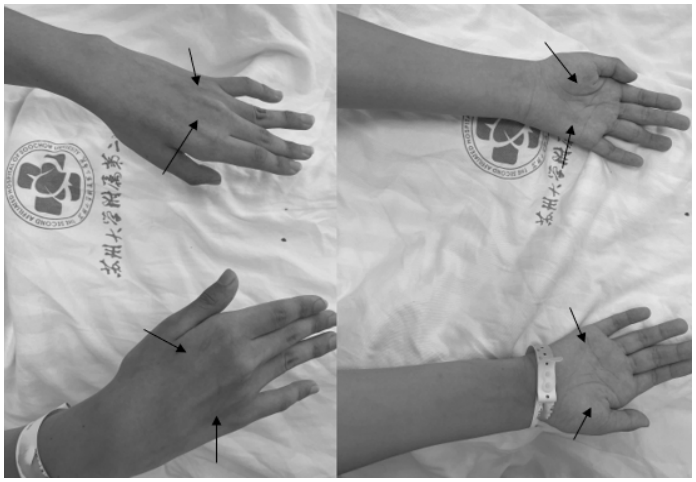

**Figure 1:** Atrophy of thenar muscles, hypothenar muscles, interossei muscles (black arrows).

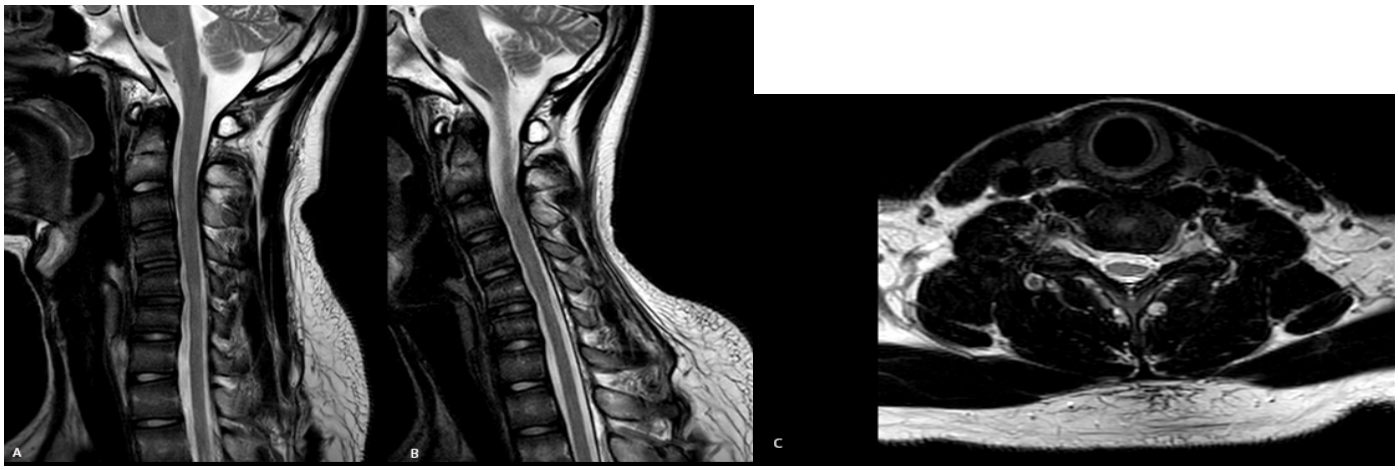


**Figure 2:** Local abnormal signals in the spinal cord **(A)**; Cord flattening with loss of attachment of dura from subjacent lamina **(B)**; Asymmetric cord flattening on neutral transverse T2WI **(C)**.

## Discussion

Hirayama disease is a cervical myelopathy that mainly affects Asian males [1]. It was first described in 1959 by Hirayama and is a rare clinical condition, which has an insidious onset and a progressive course, with subsequent clinical stabilization in a few years. It usually appears with unilateral distal weakness and amyotrophy [1-3]. In this case, we showed an Asian male with bilateral asymmetrical weakness and muscular atrophy of the hands and forearms. This example of bilateral form may be a more serious type of HD. He also had cold paresis and fasciculations during extension.

The etiology and pathogenesis of Hirayama disease are still unclear currently [1]. The leading theory is that the displacement of the posterior dural sac compress the lower cervical spinal cord, causing ischemia of anterior horn of the cervical spinal cord [1,2,4,5]. MRI images revealed compression of the lower cervical cord, the reason of which is the forward displacement of the cervical dural sac at the time of bending the neck. This mechanism is more credible in males because, in adolescence, the increase in height can lead to an imbalance between the length of the spine and that of the spinal cord and dural sac. Therefore, the displacement will be more apparent when bending the neck [6].

The diagnosis of Hirayama disease is mainly based on medical history, clinical investigation, MRI images and electromyography. History may suggest that there are triggers for the onset of the disease such as poor posture during the course. Neurological examination reveal weakness and muscle atrophy in upper limbs. The needle electromyography revealed the denervation of the atrophied muscles. Nerve conduction studies revealed moderate attenuation of the compound motor action potentials with normal motor conduction velocities. MRI scan of cervical flexion showed anterior displacement of the posterior dura, the reduced antero-posterior diameter of lower cervical cord and high signal intensity lesion which is mostly obvious at the C5-C6 level [4,7,8].

Currently, there is no effective medicine for Hirayama disease. Due to its self-limiting disease course, the treatment of Hirayama disease mainly takes conservative treatments such as wearing cervical collar and physical therapy. For patients whose conservative treatment is ineffective or even worsening, decompressive surgery is indicated. However, since the pathogenesis is unknown, surgery is controversial.

Up to date, robotic devices are new technologies used for rehabilitation of various neurological diseases [9,10].

## Conclusion

In conclusion, Hirayama disease is a rare disease presented with progressive weakness and amyotrophy of the distal upper limbs in males. Therefore, HD should be considered in any patient with an insidious onset and progressive course of hyposthenia. HD usually presents with single upper extremity weakness, we report the case of an Asian Male with asymmetric bilateral weakness and amyotrophy of upper limbs. Based on the medical history and clinical tests, it is a case of HD.

## Declarations

**Conflicts of interests:** All contributing authors report no conflicts of interest in this work.

**Approval of the research protocol:** The protocol for the research project has been approved by a suitably constituted Ethics Committee of the institution within which the work was undertaken and that it conforms to the provisions of the Declaration of Helsinki (as revised in Brazil 2013).

**Informed consent:** A statement that the subject gave informed consent and patient anonymity are preserved.

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**Authors Information:** Jun Wang; Tingting Shen; Qianchang Wu; Ping Ni; Weifeng Luo*
Department of Neurology and Clinical Research Center of Neurological Disease, The Second Affiliated Hospital of Soochow University, Suzhou 215004, China.

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