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# Bilateral Involvement in Three Patients with Hirayama Disease

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## Abstract

We report three patients with Hirayama disease with bilateral involvement. Case 1 developed weakness and wasting of bilateral hands at the age of 15 years. Case 2 developed unilateral weakness and wasting of the hand at the age of 18 years, and gradually exhibited progression to the other side within 1 year. Case 3 was a female who developed weakness and wasting of one hand at the age of 17 years, and showed dysesthesia and weakness in the other side, with pyramidal tract signs 31 years after the onset of the disease; she was diagnosed with complication of cervical disc hernia. We notice that bilateral involvement represents a severe form of this disease. We also need to differentiate incidental complications when the patients show worsening after a long stationary period.

**Keywords:** Hirayama disease, Cervical disc hernia, MRI

## Introduction

Hirayama disease is a benign focal amyotrophy of the distal upper limbs. This disease was first reported in 1959 as "juvenile muscular atrophy of the unilateral upper extremity" [1]. Since then, similar cases have been described under a variety of names not only in Japan, but also in other Asian countries, as well as in Europe and North America [2-4]. The disease is characterized by the insidious onset of unilateral or asymmetric atrophy of the hand and forearm, with sparing of the brachioradialis muscle. After a period of deterioration of 3-5 years, a stable stage is reached. The pathophysiology of this disease is considered to be dynamic cord compression during neck flexion [2-4]. A bilaterally symmetric form of the disease is reported rarely [5,6]. Recently, we experienced three patients with Hirayama disease and bilateral involvement, one of whom received anterior decompression and fusion for a cervical disc hernia 31 years after the initial onset of the condition.

## Case Reports

### Case 1

A Japanese 37-year-old male developed weakness and wasting of bilateral hands at the age of 15 years. He was diagnosed as having Hirayama disease at the local university hospital. The weakness and wasting of the hands ceased within a few years. He visited our hospital for consultation 22 years after the onset of the condition. His past medical history was unremarkable. He played volleyball during junior high school. On neurological examination, weakness and atrophies of the bilateral upper extremities were confined. Ulnar-side dominant weakness atrophies were apparent in the forearms and in the intrinsic hand muscles (Figure 1a). Handgrip power was 6.1 kg on the right side and 0 kg on the left side. Postural hand tremor was observed. Sensation was intact. Tendon reflexes on the upper and lower extremities were almost normal, and Babinski sign was absent. Routine laboratory data were mostly normal. Cervical T2-weighted Magnetic Resonance Imaging (MRI) in sagittal and axial planes disclosed cervical cord atrophy, with a maximum at the C6 level and a hyperintense signal in the parenchyma (Figure 2a and 2b).

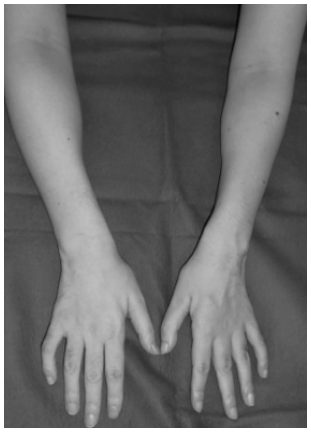

**Figure 1a:** Muscular atrophies of bilateral forearms and intrinsic hand muscles in Case 1. The brachioradialis muscles were spared and showed oblique atrophies, and the little fingers were slightly flexed.

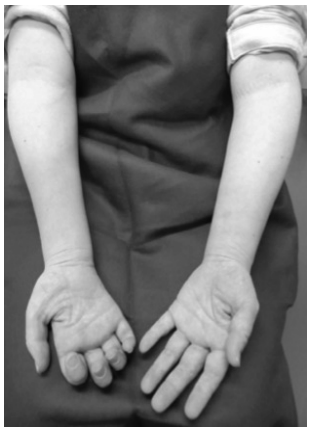

**Figure 1b:** In Case 3, bilateral weakness and atrophies were observed in the forearms and hands, predominantly at the right side, and 2–5 fingers exhibited a flexed position.

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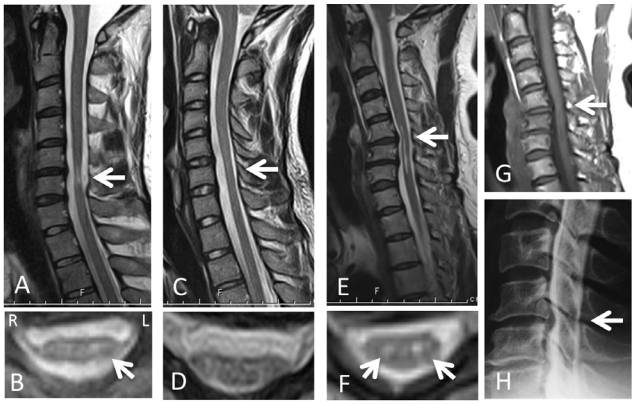


**Figure 2:** MRI findings of Case 1 (A, B), Case 2 (C, D), and Case 3 (E, F, G), and myelography findings of Case 3 (H). T2-weighted MRI showed localized cervical cord atrophy, with a maximum at C6–C7 (arrows) (A, C, E) and intramedullary hyperintensities (arrows) (A, E). Axial T2-weighted MRI disclosed hyperintense signals in the anterior horns (B, F). T1-weighted MRI (G) and myelography (H) disclosed a disc hernia at the C5/C6 level (arrows).

### Case 2

A Japanese 22-year-old male developed weakness and wasting of the right hand at the age of 18 years and exhibited gradual progression of the condition. He noticed weakness and wasting of the left hand at the age of 19 years. He visited our hospital 4 years after the onset of the disease. His past medical history was unremarkable. He played football during junior high school and high school. On neurological examination, weakness and atrophies were confined to the bilateral upper extremities. Ulnar-side dominant atrophies in the forearms and in the intrinsic muscles of both hands. Postural hand tremor was observed, but sensation was intact. Tendon reflexes on the upper and lower extremities were almost normal, and Babinski sign was absent. Routine laboratory data were mostly normal. Motor nerve conduction studies of the median and ulnar nerves showed no delay. Cervical T2-weighted MRI disclosed atrophy of the cervical cord, with a maximum at the C6-C7 level, without disc herniation (Figure 2c and 2d). The use of a cervical collar was recommended.

### Case 3

A Japanese 47-year-old female developed weakness and wasting of the right hand at the age of 17 years and exhibited gradual progression of the condition. She was diagnosed as having Hirayama disease at the local neurological clinic. The progression of the disease ceased within a few years. She played basketball during junior high school. Her past medical history was unremarkable. Recently, she noticed dysesthesia and weakness of the left hand, and was referred to our hospital 31 years after the onset of weakness in the right hand. Bilateral weakness and atrophies were observed in the forearms and hands, predominantly on the right side (Figure 1b), and she could not extend the fingers of the right hand. She showed dysesthesia on the left hand. Tendon reflexes on the upper and lower extremities were hyperactive, with both Babinski signs. Cranial nerves and bladder function were intact. Fasciculation was not observed. Preoperative laboratory data were mostly normal. T2-weighted MRI showed localized cervical cord atrophy, with a maximum at C6–C7 (Figure 2e), and disclosed the presence of a symmetrical hyperintense signal in the anterior horn ("snake eyes" appearance) in the axial plane (Figure 2f). T1-weighted MRI (Figure 2g) and myelography (Figure 2h) disclosed a disc hernia at the C5/C6 level. She received anterior decompression and fusion at the C5/C6 level.

## Discussion

Hirayama disease is a benign lower motor neuron disorder of the young, with male predominance, insidious onset of weakness and atrophy, and slow progression over 3-5 years, followed by a stationary course. The pathological and MRI findings suggest that repeated or sustained neck flexion cause an anterior shift in the cervical dural sac, which is compressed against the posterior margin of the vertebral body [2,3,7]. The compressed cervical cord at the C6 vertebral level, which corresponds to the C7 and C8 cord segments, results in microcirculation disturbances in the anterior horn. There are many reports of the MRI findings of this disease, including localized lower cervical atrophy, asymmetric cord flattening, loss of cervical lordosis, anterior displacement of the dorsal dura on flexion, and intramedullary hyperintensity [8,9]. Based on this hypothesis, the use of a cervical collar is recommended [3,9].

Judging from the onset of age, clinical course, pattern of atrophy (namely, oblique atrophy), and neurological and MRI findings, the three patients were diagnosed as having Hirayama disease. Originally, the disease was thought to affect unilateral distal upper extremities, but bilateral involvement has been reported, ranging from 15% to 61.8%, and asymmetric involvement is a consistent feature in the reported studies [3,4]. According to the long-term follow-up of 44 patients with brachial monomelic amyotrophy, eight patients (18.2%) showed minimal involvement of the contralateral upper limb, with gross asymmetry [4]. The bilateral symmetric form of this disease has been reported rarely [5,6]. Pradhan [5] reported that 11 patients (approximately 10%) among 106 patients with Hirayama disease had bilaterally symmetric involvement, and that nine of them had a history of unilateral onset. The most important characteristics of this type of presentation are severe weakness and wasting; thus, the author concluded that bilaterally symmetric Hirayama disease is a severe form of the classic disease. Jain et al. [6] reported a young male with symmetrical bilateral weakness of the hands and forearm over the last 2 years, and emphasized the pivotal role of MRI in the diagnosis of this condition. Cases 1 and 2 have this form of the disease. Case 2 showed a history of unilateral onset, but exhibited an almost similar involvement in both upper limbs after 4 years. The progression of weakness and wasting observed in Case 2 did not cease at the examination, and the use of a cervical collar was recommended. The condition of this patient warrants a careful follow-up.

Case 3 was female, and it has been reported that females are affected about 3% - 13% in this disease [3]. After attaining a stationary course, none of these patients developed fresh symptoms or signs during the long follow-up, and there was no evidence of the involvement of pyramidal tracts or cranial nerves during the follow-up assessment [4]. However, Case 3 showed dysesthesia, weakness, and wasting in the contralateral side after a long stationary period. She also exhibited pyramidal tract signs, and MRI and myelography disclosed complication with cervical disc hernia. Cervical spinal canal stenosis and cervical disc hernia are popular in middle-aged people and the elderly; thus, we need to differentiate these incidental complications when the patients show worsening after a long stationary period.

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