Olgu Sunumu/Case Report

# Travma Sonrası Hirayama Hastalığı: Olgu Sunumu

# Post-traumatic Hirayama Disease: Case Report

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***ÖZET***

*Hirayama hastalığı karakteristik klinik özellikleri arasında; genç başlangıç, sporadik oluşum, erkeklerde sık görülmesi, kraniyal sinirleri, piramidal yolları, duyusal, serebellar veya ekstra piramidal sistem tutulumu olmadan tek veya iki taraflı üst ekstremite sınırlı kaslarda erime ve güçsüzlük vardır. 22 yaşındaki bayan, alt motor nöron sendromu benzeri sağ tarafta belirgin üst ekstremitelerde yavaş ilerleyen güçsüzlük ile başvurdu. Bir yıl once trafik kazası geçirmişti. Son altı aydır güçsüzlük şikayeti artmıştı. İntervertebral disk hernisi yoktu, ancak; eski travmaya ait bulgular vardı. Burada, biz bir travma sonrası geliştiğini düşündüğümüz nadir bir Hirayama hastalığı olgusu sunduk.*

***Anahtar Kelimeler:*** *Travma, Hirayama*

***ABSTRACT***

*The characteristic clinical features of Hirayama disease are young age at onset, sporadic occurrence, male preponderance, wasting and weakness confined to a single or bilaterally upper limb without involvement of cranial nerves, pyramidal tracts, sensory, cerebellar or extrapyramidal systems. A 22-year-old girl was presented with slowly progressive weakness of her upper limbs especially on her right side like a lower motor neuron syndrome. She had a car accident one year ago. She had a complaint of weakness on her left side for last six months. No intervertebral disc herniation was present, but there was an evidence of old traumatic injury. Here, we presented a rare case of Hirayama disease after a trauma.*

***Keywords:*** *Trauma, Hirayama*

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**INTRODUCTION**

Hirayama disease is a rare neurological disease reported especially in Asia. Clinically, Hirayama disease is characterized by weakness of the hands and forearms in association with unilateral or bilateral, usually asymmetrical, muscular atrophy that develops gradually in young adults.1 Hirayama disease is a myelopathy related to flexion movements of the neck that produce ischemic damage in the anterior horn of the cervical cord.1 But here, we presented another rare case of Hirayama disease after a trauma.

**CASE**

A 22-year-old woman was presented with atrophy of the right upper limb. She had an accident and an operation about cervical vertebral fracture one year ago. The patient had noticed slowly progressive weakness of the fourth and fifth right fingers as well as atrophy of the right hand over the past year. Then, six months ago, the patient had weakness of the first three right hand fingers. Clinical examination showed an evident atrophy of the thenar, hypothenar and interosseous muscles of the bilaterally hands especially on the right side (Fig-1).

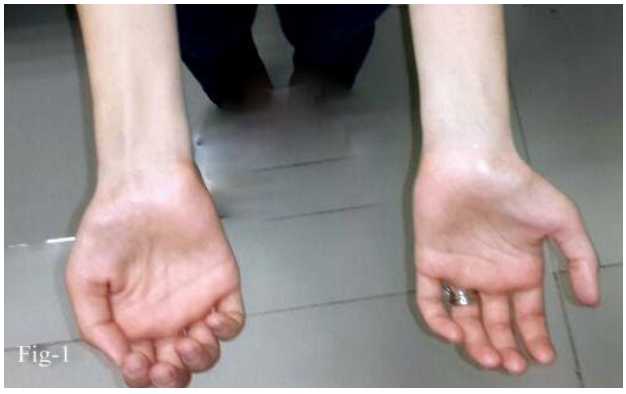


**Fig-1:Atrophy of the thenar, hypothenar and interosseous muscles of the bilaterally hands especially on the right side.**

Right ulnar claw was present (Fig-2).

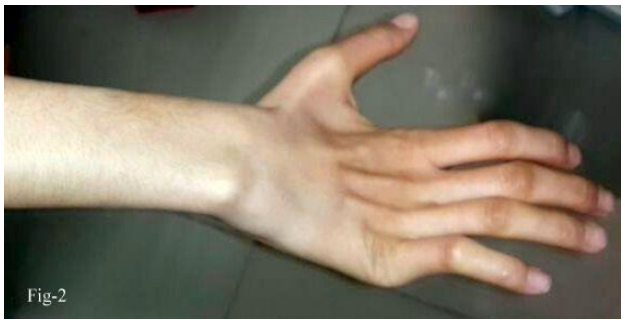


**Fig-2:Right ulnar claw.**

The right hand deficit was predominantly distal and involved the flexor digitorum, wrist flexors and interosseous muscles. The muscles of the upper arms and legs were completely normal. There was no proximal or lower limb strength deficit. Deep tendinous reflexes in all limbs were normal, and there was no Babinski sign or sensory deficit.

Electromyography showed denervation changes in C8-T1 muscles. No conduction blocks were identified. Nerve conduction studies showed normal sensory and motor velocities of the median and ulnar nerves. Cervical MRI study showed changes seconder to operation on the lower cervical vertebras at the level of C6-C7 (Fig-3).

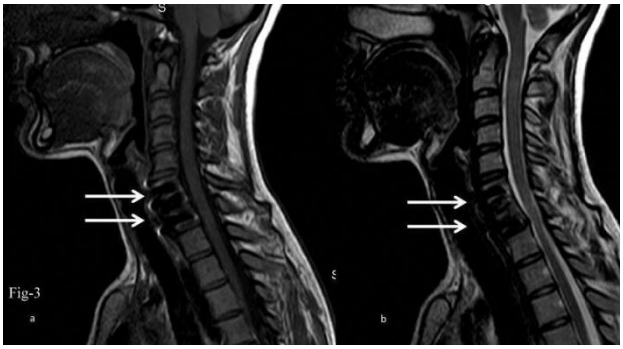


**Fig-3:Cervical MRI study shows changes seconder to operation on the lower cervical vertebras at the level of C6-C7.**

There was small increased signal intensity on T2-weighted images. No intervertebral disc herniation or degenerative change was present. No progression of symptoms was observed at the follow-up stage; thereby, no surgical treatment was performed.

**DISCUSSION**

Diagnostic criteria of Hirayama disease have included male predominance, weakness and atrophy predominantly in the C7-T1 myotomes in one upper limb or asymmetrically in both upper limbs, insidious onset in teenagers, initially rapid progression for 1-3 years, followed by arrest of the disease or a relatively benign course, electromyography evidence of chronic denervation in the clinically or subclinical affected muscles and absence of objective sensory loss.1 Our 22-year-old female case was presented with slowly progressive weakness of her upper limbs especially on her right side one year after a car accident.

The disease mechanism is unclear, but symptoms are attributed to the forward displacement of the posterior cervical dural sac when the neck is in flexion due to imbalanced

growth between the patient's vertebral column and spinal cord.2,3 Patients had flattening and atrophy of the lower cervical spinal cord. The causal mechanism behind such flexion-induced myelopathy was supported by focal ischemic damage in the anterior horn of the cervical cord attributable to arterial insufficiency during flexion and microcirculatory changes.4,5 Compression of the anterior cervical dural sac is associated with compression of the anterior spinal artery, resulting in focal ischemia. Ischemic damage in the anterior horn of the cervical cord attributed to arterial insufficiency during flexion and microcirculatory changes also appear.6 After MRI and electromyography studies we thought that patient look like a Hirayama case after trauma.

Differential diagnoses that should be excluded include syringomyelia, amyotrophic lateral sclerosis, cervical spondylotic myelopathy, expansile lesions, motor neuron disease and hereditary distal motor neuropathies.1 Although the etiology of sporadic motor neuron diseases remains unknown in the vast majority of cases, mechanical trauma has been suggested as a factor mostly regarding amyotrophic lateral sclerosis in adult patients.

To prevent functional disability, cervical-collar therapy is necessary. Cervical decompression with or without duraplasty as a surgical intervention has also good results.3 No progression of symptoms was observed at the follow-up stage of our case; thereby, no surgical treatment was performed. Only physical therapy was performed.

On conclusion; we presented another rare case of Hirayama disease to show that rarely cause may be a trauma. But case series necessary about this subject.

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