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Case Report

Hirayama Disease: a Case Report with Late Onset

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Abstract: The etiology of Hirayama's disease (HD) is still controversial, but the most accepted mechanism is the repetition of sudden neck flexion movements, which causes a flattening of the cervical spinal cord. It reaches the cells of the anterior tip of the medulla in the C7-T1 segments. 45-year-old patient with HD, diagnosed 1 year ago with typical atrophy of the left upper limb (LUL) associated with fasciculation and decreased osteotendinous reflexes in upper limbs (ULL). Due to the rarity of the disease, there are many underdiagnosed cases, which results in low epidemiological data reported. We are aware that the prevalence in males is explained by accelerated growth at puberty.

Keywords: Hirayama disease; Upper limbs; Cervical spinal cord.

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1. Introduction

The Hirayama Disease (HD) was first described by Kenzo Hirayama in 1959 and it was first designed as juvenile muscular atrophy. Hirayama disease is coined in 1991 [1]. It is always described as a rare condition, but we were not able to find its real incidence searching for it in the literature. In a report in 1991 Chan estimated 150 cases for Japan, 37 for Indian and 102 for Sri Lanka [2]. It affects predominantly young males in their teens and twenties [1, 3], especially Asians. It was observed anterior displacement of posterior dura matter is series of 15 cases and cervical flexion would cause compression and ischemic changes to the anterior horn cells of the spinal cord [3].

It is a self-limited condition, and manifestations include distal weakness of the upper limbs, asymmetrically, worsening on exposure to cold and generally absence of sensory disturbances. Differential diagnosis should include Amyotrophic lateral sclerosis, syringomyelia and cervical compression among others. Treatment is conservative with cervical collar but some cases may require surgical treatment, with posterior lateral mass instrumentation [4] or cervical anterior discectomy and fusion [5].

2. Case Report

A black man 35 years old, bus driver who complains that for the last 2 years he has been observing distal left upper limb muscle atrophy and weakness which rapidly affected also his right upper limb (Figure 1). At the same time, he had some difficulty to do flexor and extensor movements with both hands and fingers and he was not able to drive anymore. So, he was examined by an orthopedist who asked a consultation with a neurologist. When he came to our office, we were able to observe bilateral muscle upper limb

atrophy, fasciculations and diminished deep tend reflexes. There is no medullar compression on MRI (Figure 2). Electromyography demonstrates fibrillations potentials and active denervation of distal muscles. He was treated with cervical collar and physiotherapy.

Figure 1. Distal muscles atrophy.

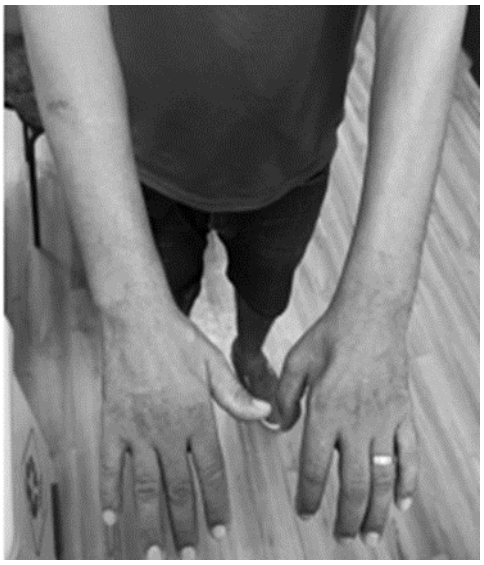
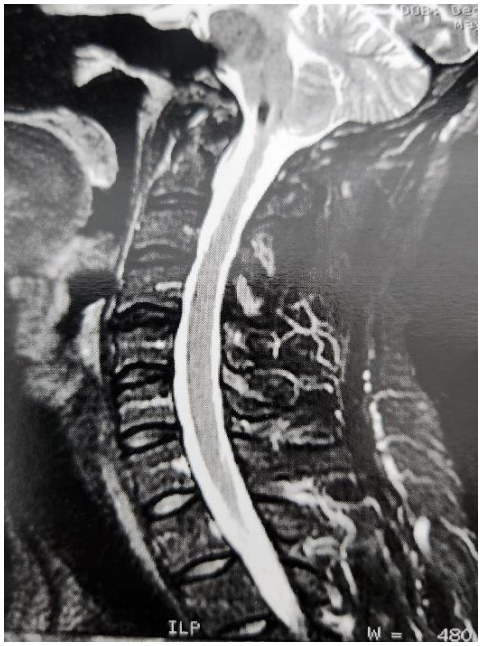


Figure 2. Cervical MRI.



3. Discussion

HD is more common in Asian population, but an increasing incidence is being observed in North America, Europe, and Australia in the last decades [1]. A male: female ratio varies 20:1 in Japan; 2.8:1 in India; and 11:1 and 31.6:1 in China [2, 3]. The right upper limb is much commonly affected initially however in this patient the left upper left limb was initially affected and then both were affected. This patient presented weakness, fasciculations, bilateral muscle atrophy, insidious onset, that are consistent with clinical diagnostic criteria for HD. These criteria are essentially insidious onset, weakness, atrophy,

fasciculations, unilateral or bilateral predominantly asymmetrical and self-limited progression. Rarely sensory disturbance may occur as well as pyramidal signs [6].

Computed tomography (CT), Magnetic resonance imaging (MRI) as well as electromyography are very important to confirm the diagnosis. On CT and MRI forward displacement of posterior dural sac when performing a cervical flexion can be observed and this would cause compression and injury of spinal cord damaging the anterior horn cells. However, the exact mechanism of HD is not exactly understood [4]. Areas of cervical spinal cord atrophy loss of cervical lordosis are observed in MRI. In Myelography is difficult to retain contrast in the subarachnoid space but is not used anymore [5].

4. Conclusions

HD presents self-limited progression with better prognosis than other motor neuron diseases. Treatment is usually conservative, but some cases may require surgical procedures.

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