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CASE REPORT

Medicine Science 2017;6(3):560-1

Medicine Science International Medical Journal

# Hirayama Disease (monomelic amyotrophy)

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Received 01 February 2017; Accepted 21 February 2017
Available online 16.03.2017 with doi: 10.5455/medscience.2017.06.8598

**Abstract**
Hirayama disease (HD) or monomelic amyotrophy (MA) is a rare muscular atrophy that affects young Asian males, usually occurs in one of the upper limbs that progresses slowly. It is diagnosed by means of electromyographic/electroneurographic conduction speed studies and by magnetic resonance imaging (MRI) of the spinal cord. In this paper two different HD case is reported.

**Key words**: Hirayama Disease, motor neuron disease, electromyography, late on-set.

## Introduction

Hirayama disease (HD), also known as monomelic amyotrophy (MMA) was first reported by Hirayama et al. in 1959 as a benign focal motor neuron disease characterized by unilateral atrophy of the forearm ulnar muscle [1]. This rare disease affects mostly unilateral second motor neuron of upper limbs. Atrophy and weakness are mostly seen on distal muscles of the limb. This disease is more prevalent in east Asian countries. It mainly develops in the late teens and early twenties with a male preponderance. Symtoms progresses initially and is followed by spontaneous arrest several years after the onset [2]. Here we report two different cases of HD.

## Case 1

A 80-year-old man presented with a 6 months' history of weakness and atrophy with rarely happening numbness that started in the left hand and forearm. The weakness was first observed during carrying weights with affected arm. His and his family past medical history was unremarkable.

Neurologic examination revealed thenar atrophy of the left hand. The weakness of the forearm muscles on the left was 4/5. The weakness of the first distal interphalangeal joint, first proximal interphalangeal joint, second distal interphalangeal joint and interphalangeal joint were 2/5, 3/5, 3/5 and 3/5 respectively.

Routine laboratory tests, including full blood count, thyroid function tests and serum creatine kinase were within normal limits. Magnetic resonance imaging of the servical spine resulted with normal findings. Nerve conduction studies (NCS) showed no amplitude of compound muscle action potential (CMAP) in left median and ulnar nerves. Sensory NCS was normal in both nerves. The needle examination showed chronic partial denervation signs like high amplitude and duration of motor unit potentials, fibrillation potentials and dilution of contraction pattern.

## Case 2

This 23-year-old women presented with 2 years story of slowly progressive distal weakness, atrophy and numbness of the both hand and forearms. The majority of the symptoms were right sided which she was unable to carry weights with. She had a medical history of surgirical operation for carpal tunnel syndrome of right wrist. Neurologic examination revealed thenar atrophy of the both hands. The weakness of the intrinsic hand muscles on the left was 4/5.

Routine laboratory tests, including full blood count, thyroid function tests and serum creatine kinase were within normal limits. Magnetic resonance imaging of the servical spine resulted with normal findings. Nerve conduction studies were normal in both side median and ulnar nerves. The needle examination showed high amplitudes of motor unit potentials, giant potentials and resting denervation potentials of both side abductor pollicis brevis and abductor digiti minimi muscles. These findings correlated with involvement of anterior horn of spinal cord.

## Discussion

Hirayama disease which is rare in western countries, is more prevalent in Asian countries. As we could search a few cases repoted from Turkey [3-5]. Development in the second or third decade is one of the main characteristic of the HD. Like our first case late-onset HD is so rare [6].

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Magnetic resonance imaging could help diagnostic research for HD. Lower servical cord atrophy, asymmetrical cord flattening, loss of attachment between the posterior dural sac and subjacent lamina, abnormal servical curving and posterior epidural venous widening are some of the MRI findings of HD [4]. Atypically there were no pathological MRI finding in our patients.

Electromyographic studies are needed to diagnose for HD. Motor NCS present with low amplitudes of motor unit potentials while sensory NCS of the median and ulnar nerves result normal in most of the patients. As it is devised in our patients findings for involvement of anterior horn of spinal cord are the charactheristic of the needle studies in HD patients [7]. But atypically motor NCS resulted normal in our second case.

In conclusion her we report two rare cases of HD with atypical presence. HD is a progressive disease, and should be considered in patients presenting with weakness and atrophy of upper limbs.

## References


1. Jin X, Jiang JY, Lu FZ, Xia XL, Wang LX, Zheng CJ. Electrophysiological differences between Hirayama disease, amyotrophic lateral sclerosis and cervical spondylotic amyotrophy. BMC Musculoskelet Disord. 2014;15:349.
2. Anuradha S, Fanai V. Hirayama Disease: A Rare Disease with Unusual Features. Case Rep Neurol Med. 2016;2016:5839761.
3. Yoldaş TK, Çoruh Y, Keklikoglu HD, Polat HG, Güven B. Hirayama Hastalığı. Fırat Tıp Dergisi 2008;13(4):274-6.
4. Yuksel M, Kalemci O, Yuksel KZ, Erguden C, Yucesoy K. Hirayama hastalığı ve tanıda Manyetik Rezonans görüntülemenin önemi. Sinir Sistemi Cerrahisi Derg. 2009;2(4):191-5.
5. Ozturker C. Kara K, Incedayi M, Sonmez G, Mutlu H.. Hirayama disease. Spine J. 2016;16(5):299-300.
6. Patel DR, Knepper L, Jones HR Jr. Late-onset monomelic amyotrophy in a Caucasian woman. Muscle Nerve. 2008;37(1):115-9.
7. Basil T. Darras, Jennifer A. Markowitz, Umrao R. Monani, Darryl C. De Vivo. Spinal Muscular Atrophies In: Neuromuscular Disorders of Infancy, Childhood, and Adolescence: A Clinician's Approach. 2nd edition, Elsevier, London, 2014: 117-45.