

Hirayama Disease

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KEYWORDS

- Hirayama disease • Cervical myelopathy • Neck flexion
- Adolescent

Hirayama disease is a benign, self-limiting cervical myelopathy first brought to attention by Hirayama¹ in 1959. During the past half century, researches have established this disease as a new entity that differs from motor neuron disease because of spinal cord compression by the posterior dural sac during neck flexion. Primarily affecting male adolescents, it is clinically characterized by a progressive course of distal upper-limb muscular weakness and atrophy, followed by a spontaneous arrest within several years. Early recognition of the disease allows early intervention, which has been shown to stop disease progression. This article reviews the clinical presentation, pathophysiology, image findings, and treatment of Hirayama disease, because a thorough understanding of the disease is essential for early recognition and treatment.

In 1959, Hirayama and colleagues² published 12 cases distinguishing a new clinical entity from what was originally thought to be a type of progressive and degenerative motor neuron disease. The term “juvenile muscular atrophy of unilateral upper extremity” was applied. This new clinical entity was further consolidated by following reports of 20 patients in 1963³ and 38 patients in 1972.⁴ The patients are generally in their teens and early twenties, predominantly male, with insidious onset of weakness and muscle wasting involving unilateral distal upper limb, followed by variable period of progression and spontaneous arrest within several years.

Since then, there have been many reports of Hirayama disease, mostly from Japan and other Asian countries, as well as few from other countries.^{5–7} However, early reports in the 1960s and 1970s emphasized clinical features and electrophysiological evaluations. It was not until 1982 that the first autopsy case was obtained by Hirayama and

colleagues,^{8,9} which revealed anterior-posterior flattening of the lower cervical cord associated with ischemic and atrophic changes of the anterior horn cells. Pathologic evidence of focal ischemic cervical poliomyelopathy of the disease prompted neuroradiologic investigation in search of the anatomic pathophysiology of the spinal cord and canal. Earlier studies were conducted using myelography and CT myelography.^{10,11} Since MR imaging equipment became available and widely accessible in the late 1980s,^{11–15} several publications have provided valuable findings in neck-flexion and neutral-position MR imaging of the cervical spine that have assisted the diagnosis of this disease. Imaging findings revealed dynamic changes of the cervical spine, spinal canal, and spinal cord during neck flexion, mainly in the forward displacement of posterior dural sac, causing anterior-posterior flattening and asymmetric atrophy of the lower cervical cord, which corresponded with the pathologic findings.

Several names have been used in publications to describe this disease, including benign juvenile brachial spinal muscular atrophy, juvenile asymmetric segmental spinal muscular atrophy, juvenile muscular atrophy of the distal upper extremity, monomelic amyotrophy, and oblique amyotrophy.^{1,5,9,16,17} Hirayama disease is still the most commonly used named throughout publications in neurology and neuroradiology, as well as related fields.

DEMOGRAPHICS

Hirayama disease occurs mainly in people in their teens and early twenties with a significant male predominance. The mean age of onset ranges from 15 to 20 years of age in different

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studies.^{5-7,17-21} The degree of male preponderance reported in large series ranged from purely male to a male/female ratio of 2.8:1.^{6,22,23} A delay of age of onset after the peak age of normal growth curve was observed in several studies.⁵⁻⁷ Hirayama⁵ observed that the peak of onset age is approximately 2 years later than the peak of longitudinal growth curve of juveniles in Japan. Based on this finding, Hirayama^{1,5} speculated that disproportional growth between the vertebral column and the contents of the spinal canal, especially the dural sac, during the juvenile growth spurt underlies this phenomenon. Tashiro and colleagues²⁴ suggested that the difference in the male and female incidence of the disease could be explained by a more rapid increase in height of males at puberty compared with females.

Hirayama disease is nonfamilial in most patients. There are, however, a few reports of familial cases. Nine familial cases have been reported in the English literature.²⁵ So far, there are no published reports regarding genetics information that maybe related to familial Hirayama disease. The explanation for the familial occurrence is indeed interesting and needs further studies.

As Hirayama disease became more recognized, several cases and large series have accumulated. There are an exceedingly large number of cases in Japan and more cases reported from other Asian countries in the past 3 decades, particularly in China, Taiwan, Malaysia, India, and Sri Lanka, but relatively fewer cases from Europe and North America.⁵⁻⁷ Whether there might be an ethnic factor in this disorder is still unclear.

CLINICAL FEATURES OF HIRAYAMA DISEASE

Patients with Hirayama disease present with insidious onset and slow progression of muscle weakness and muscular atrophy of the distal upper limb, including thenar, hypothenar, interossei muscles, and wrist flexors and extensors, with sparing of brachioradialis muscles (Fig. 1). The border of muscular atrophy runs obliquely over the volar and dorsal surfaces of the forearm, therefore it is also referred to as oblique amyotrophy.⁴ Both extensor and flexor muscles of the fingers and wrists are involved, particularly the finger extensors and wrist flexors. Some investigators reported asymmetric atrophy was more severe on the ulnar than the radial side in the flexor and extensor compartments.^{7,16} The phenomenon of cold paresis is widely observed in the patients, characterized by exacerbating of finger weakness on exposure to cold environment.^{5,6} Kijima and colleagues²⁶ proposed that this is caused by conduction block of the muscle fiber membrane

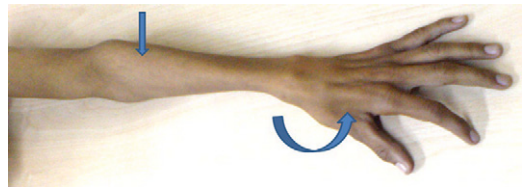


Fig. 1. Photograph of a 19-year-old man, note the wasting of first dorsal interosseous (*curved arrow*) and other small muscles of the hand with preservation of brachioradialis muscles (*straight arrow*) giving an appearance of oblique amyotrophy. (From Pradhan S. Bilaterally symmetric form of Hirayama disease. *Neurology* 2009;72:2083-9; with permission.)

in reinnervated muscles after active denervation. When cold paresis is induced, compound muscle action potentials show delayed latencies and decreased amplitude after high-frequency repetitive nerve stimulation. Resting fasciculation is not observed, but contraction fasciculation is well documented.^{7,27,28} It is characterized by fascicular twitching in the extensor side of forearm muscles or tremor-like movement of the fingers during stretching. Fatigue is the most common symptom (33%), followed by cold paresis, atrophy, and tremor.²⁹ Although most patients reported absence of subjective or objective sensory disturbances, a few patients show slight hypoesthesia in a localized area of the hand.^{5,6,30} The muscle stretch reflexes of the arms and legs are within normal range. There are no accompanying neurologic symptoms such as cranial nerve involvement, pyramidal signs, sensory disturbance, and urinary disturbance. Muscle biopsy and needle electromyography show denervation of the atrophied muscles.^{5,27} Nerve conduction velocity and analysis of the cerebrospinal fluid are almost normal.

Patients with Hirayama disease commonly show initial unilateral monomelic atrophy, but it is asymmetrically bilateral in some and, occasionally, symmetric.⁵ The right upper limb is more frequently involved in Hirayama disease, regardless of the handedness.^{3,19,23,30} Huang and colleagues⁶ reported more conspicuous manifestation in right upper limb with a right/left ratio of 3:1. Often, the disease progressed to involve the other side in an asymmetric manner.^{6,16} A bilateral symmetric form of Hirayama disease has occasionally been reported. The incidence was 3.1% by a National Japan Survey²⁴ and 10% by Pradhan,³¹ who reported 11 cases of bilateral involvement in a series of 106 patients. He proposed that the "bilateral symmetric Hirayama disease" is a more severe variant of Hirayama disease based on more severe and significant clinical and

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