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Case Report

Hirayama disease: An unusual case report

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Abstract

Hirayama disease is an uncommon and rarely seen lower motor neuron disease. It is usually seen during periods of rapid growth (15-25 years). Distal upper extremities characteristically show weakness and atrophy. In this paper a case is presented of this rare, and difficult to diagnose disease, not previously reported in this age group, and the findings are discussed in the light of information of earlier cases in Turkey.

A 53-year old male patient presented at Inonu University Neurology Polyclinic with complaints of motion tremor and atrophy in the left arm, which had been ongoing for 4 years. After electromyography, the deltoid, biceps, triceps, extensor digitorum communis, abductor pollicis brevis and abduction digiti minimi muscles were identified in the upper left limb. This case report presents information about the differential diagnosis, detailed symptoms and treatment methods of a Hirayama patient applying for physical therapy.

Keywords: Hirayama Disease; Lower Motor Neuron; Atrophy; Physiotherapy; Function.

INTRODUCTION

Hirayama disease (monomorphic amyotrophy), first described by Hirayama in 1959, is a benign sub-motor neuron disease that is five times more common in young males than in females (1,2). Bilateral muscular weakness and atrophy develop unilaterally or asymmetrically in the hands (tenar, hypotenar and interrosseous muscles) and slowly progresses in the anterior cruciate in 20% of patients. The disease is also known as juvenile muscular atrophy of the distal extremity distal. The disease has an insidious onset with slow progression in the first 2-5 years, then progresses more rapidly at approximately 5 years after onset. While more than half of the patients have unilateral weakness and atrophy, it appears bilaterally in a third of patients (2-6). It is usually seen during periods of rapid growth (15-25 years). Atrophy in the forearm occasionally ruptures the C6 innervated brachioradialis (oblique amyotrophy). There is no sensory loss, or the motor movement is delayed with respect to symptomatic findings. Muscle strength is also known as cold paralysis with poor prognosis with exposure to cold (3,4). Muscle weakness and atrophy are not usually seen on the face, neck, chest and stomach of patients with Hirayama disease (5). Weakness is seen when the involvement of the splenic extensor and ankle flexor muscles is dominant, affecting the flexor and extensor muscles in combination (6). In Hirayama patients, movement tremor is seen, triggered by light mobilization on the fingers while resting (7). Hirayama et al. (1996) reported that the posterior wall of the dura mater in in the lower cervical region was not sufficiently elastic, and that the taut dural portion was anteriorly displaced during the flexion movement, while dynamic compression and ischemic changes on the spinal cord were responsible (8). Hirayama disease is uncommon and the cause is unknown. The generally accepted hypothesis is cervical myelopathy induced by function (9,10). This disease is accompanied by non-specific ischemic lesions in the anterior horn cells. The increase in length observed in the loose dura mater flexion in healthy subjects may compensate. However, in patients with Hirayama disease, the length of the spinal column increases with flexion, and the dura mater is stretched and separated from the vertebral canal wall (11). This results

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in posterior dura mater anterior displacement and spinal cord compression. The compression of the lower cervical spinal cord may cause circulatory disturbances in the area of anterior spinal arterial irrigation (12). Chronic circulatory disturbances that occur with recurrent or continuous neck flexion can lead to gliosis and limited cord atrophy (13). In the radiological diagnosis of the disease, magnetic resonance imaging (MRI) examination is primarily applied. Various MRI findings have been described, include localized lower cervical cord atrophy, asymmetric cord flattening, parenchymal signal changes in the lower cervical cord, abnormal cervical curvature, posterior dural sac and ligation of the junction beneath the lamina below and posterior epidural extension of the posterior wall of the cervical dural canal (14,15). There is evidence of areas with signal loss due to flow, with a prominent epidural area suggesting a dilated epidural component and dilated epidural venous plexus. The changes are most common at the C6 vertebral level. Asymmetric cord atrophy in the lower cervical region has been described in routine MRI studies in Hirayama patients. When these findings are observed in a young patient with atrophy of the hands and / or forearm, Hirayama disease should be suspected and should be identified in the MRI study (16).

CASE REPORT

Presenting Concerns

A 53-year old male patient, of height 1.65m and weight of 82 kg presented at the neurology Polyclinic of Inonu University Turgut Ozal Medical Center in June 2017 with complaints of motion tremor and muscle weakness on the left side, which had been ongoing for 4 years.

Clinical Findings

In the neurological examination performed, low amplitude of BCAP and prolongation in distal latencies in the left musculocutaneus, axillary nerve were recorded and further neurological motor unit potential tests were applied. Subsequently, cervical cord atrophy, asymmetric cord flattening, parenchymal signal changes in the lower cervical cord, and abnormal cervical curvature were observed on magnetic resonance imaging. In addition, atrophy was observed in the left upper limb in the deltoid, biceps, triceps, extensor digitorum communis, abductor pollicis brevis and abductor digiti minimi, tenar, hypotenar, and interosseous muscles (Figure 1). Left hand distal and proximal muscle loss and joint contracture also developed (Figure 2).

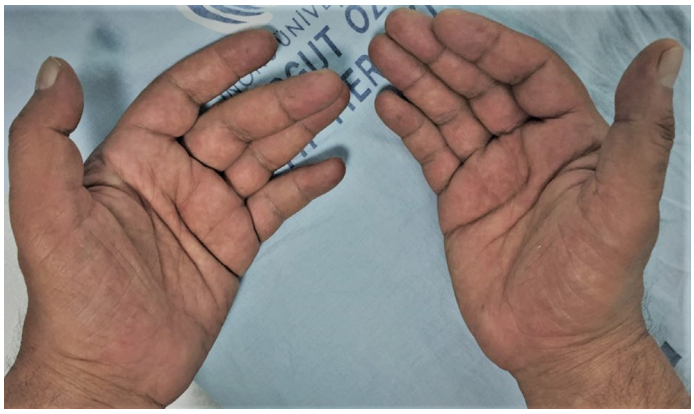

Figure 1. Left hand thenar and hypothenar region muscle atrophy

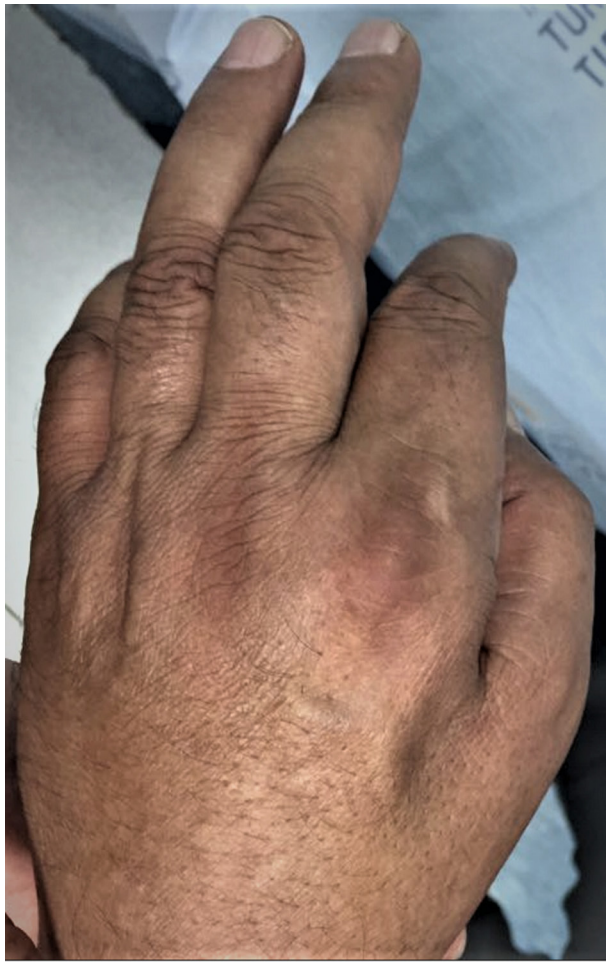

Figure 2. Left hand distal and proximal atrophy and joint contracture

Diagnostic Focus and Assessments

The distance of anterior head tilt was 18.5 mm, the C7 corpus vertebral angle (C7 angle) was 37^0, the posterior tangent method was C2-3 C7 with 21^0-34^0 cervical lordosis while patients diagnosed with Hirayama disease usually have a 43.7^0 angle of cervical lordosis. (Figure 3).

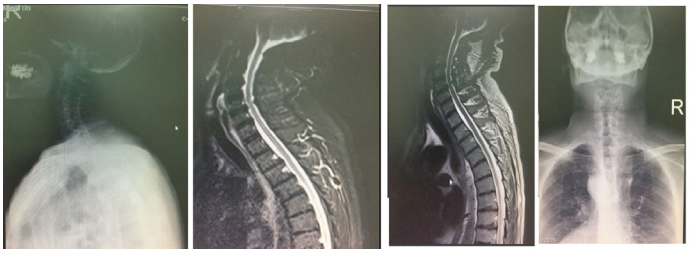

Figure 3. MRI image of hinging cervical involvement

Normally the scapular adduction angle is 17^0 while it was determined as 14^0 in the current patient, and scapular abduction is normally 17^0 and the current patient was measured as 20^0. Upper extremity range of motion (ROM) resuls of patient with Hirayama disease are shown in Table 1. In this patient, evaluation was made of the psychosocial status, history, background, cervical region range of motion (ROM), cervical region extensor muscle strength test (levator scapula-trapezius-sternocleidomasteoidus (scm)-related spina muscular evaluations), and carpal

intrinsic and extrinsic muscles. (Table 2). Table 3 presents more detailed anthropometric circumference evaluation (ACE) results of the upper extremities.

Metocarpo-phalangeal, and interphalangeal ROM were also assessed. The Box and block test (BBT), Nine-hole peg test (NHPT), Bournemouth neck survey, Beck depression inventory and Functional indicator of Revel were used to assess hand skills. Anterior and lateral clinical posture analysis was applied. A rough grip (grip meter), and finger grip (pinch meter) were used for distal muscle strength assessment. The evaluation of daily living activities in Hirayama patients is applied using the Barthel index (BI).

In the anterior postural analysis of the current patient, the right shoulder was higher and left-side lateral flexion was detected. In the lateral postural analysis, anterior head tilt, increased total kyphosis and increased lumbar lordosis were observed. VAS was used to evaluate upper extremity neck pain. The values reported by the patient were a maximum pain intensity of 10 and the lowest of 5. Rough and finger grip strength (pinch) values were measured (Table 4) using a Jamar hand dynamometer to measure rough grip strength and a pinch meter to measure the pinch finger grip strength.

In the measurement of neck pain, 69 points from a maximum of 70 were recorded on the Bournemouth Neck Survey. A total of 40 points were recorded on the Neck Disability Questionnaire, in which ≥35 points is defined as complete disability.

With a score of 42 points on the Beck Depression Inventory, the patient was diagnosed as having a severe depressive condition. In the assessment of the functional status of the hand, the score recorded was 16/20 points on the Revel's Functional Indicator.

In the box-block test, which measures hand grip ability, the number of strokes of box blocks in 1 minute is recorded. The test was repeated 3 times and the highest scores recorded were 34 for the right extremity and 11 for the left.

The nine-hole peg test (NHPT), which measures hand strength, consists of 9 wooden bars of equal dimensions and a wooden block with 9 holes. The patient places the bars in the holes and the time is recorded. The average of 3 trials is used for the evaluation. The current patient had no movement in the left arm and the average placement time of the blocks was 35 seconds.

Table 1. Upper Extremity Range of Motion (ROM) Measurement Results

	ROM of Right Extremity	ROM of Left Extremity
Abduction of Shoulder	60^0	7^0
Flexion of Shoulder	70^0	5^0
Flexion of Elbow	135^0	15^0
Flexion of Wrist	65^0	20^0
Extension of Wrist	60^0	15^0
Supination of Wrist	48^0	50^0
Pronation of Wrist	60^0	45^0
Ulnar Deviation of Wrist	25^0	20^0
Radial Devation of Wrist	30^0	15^0
Flexion of MCP	60^0	10^0

Table 2. Evaluation of Muscle Strength of the Upper Extremities

	Muscle Strength of Right Extremity	Muscle Strength of Left Extremity
Abduction of Shoulder	3	1
Flexion of Shoulder	4	1
Adduction of Shoulder	3	1
External Rotation of Shoulder	4	2
Medial Rotation of Shoulder	3	1
Flexion of Elbow	5	3
Extension of Elbow	4	2
Flexion of Wrist	4	2
Extension of Wrist	5	1
Radial Deviation of Wrist	3	2
Ulnar Deviation of Wrist	4	2
Flexion of MCP	4	2

Table 3. Anthropometric Circumference Evaluation (ACE) Results of the Upper Extremities

	ACE of Right Extremity	ACE of Left Extremity
Wrist (on styloid process of ulnae)	17.5 cm	18 cm
Forearm (widest part of forearm)	25 cm	22 cm
Biceps (widest part of biceps)	27 cm	25 cm

Table 4. Hand and Finger Grip Strength Measurement Results

	Right Extremity	Left Extremity
Pinch Grip Strength	2.33 kg	0.33 kg
Rough Grip Strength	13 kg	4.33 kg

DISCUSSION

This paper presents the detailed symptoms and treatment methods of physical therapy applied to a patient with Hirayama disease, and information about the differential diagnosis. To the best of our knowledge, this is the first study to have investigated detailed symptoms, the differential diagnosis, and treatment methods used in the rehabilitation of Hirayama disease.

Early detection of the disease is extremely important as the prevention of neck flexion can significantly reduce the progression of the disease. As the currrent patient presented at the polyclinic after 4 years of disease progression, almost complete findings of the disease late period were observed.

According to previous studies, 20% of patients with Hirayama disease (in thenar, hypothenar and interrosseous muscles) and bilateral muscular weakness and atrophy develop slowly with unilateral or asymmetric progression in the anterior cortex (14-18). The current patient was

seen to be consistent with these findings in literature. The patient had a marked degree of atrophy in the left upper extremity, thenar and hypothenar muscles. Unlike previous reports in literature, in the tests and clinical evaluations applied to the current patient, neck pain and limitation of movement were observed, with severe narrowing of the cervical canal in neck flexion seen on MRI.

Tokumaru et al. reported that even if the disease is non-progressive, myelopathy should be treated when symptomatic. Early recognition of the disease contributes to the prevention of neck flexion. To evaluate the effect of cervical collar treatment in Hirayama patients, a collar orthosis for cervical stabilization was applied to 38 patients with progression over a period of 5 years, and these were compared with an untreated control group of 45 patients. Spinal cord atrophy and flattening of the neck flexion before and after treatment were evaluated on computed tomography (CT) and MRI. After treatment, the duration of disease progression was seen to be 1.8 years in the experimental group and 3.2 years in the control group. In the experimental group, 15 patients were not only stabilized but also recovered from cold paralysis and muscle weakness. The use of cervical collar (Cervical Collar Orthosis, Philedephia cervical orthosis, Somi cervical orthotics and Cervical Brace) has been reported to prevent flexion and progression of the disease (17). Surgery can be performed when a cervical collar is ineffective. Surgical intervention should be the first choice in patients with an inelastic posterior aspect of the dural canal and passive absence of venous occlusion because of the presence of the anterior surface of the canal and no benefit has been gained from conservative treatment and the complaints are progressive (17,18). Surgical options, such as cervical decompression and fusion, have shown favorable results (19). Another aim is to prevent anterior flaccidity with anterior fusion as another surgical option (19). When Hirayama disease is diagnosed, physical therapy exercises and a cervical collar are recommended with clinical follow-up.

In physiotherapy applications, the exercise program should aim to prevent movement inhibition of tremor, thenar, hypothenar and interrosseous muscular atrophy, reduce distal and proximal joint contractures, and increase muscle strength (18, 20). In parallel with literature, the current patient was applied with exercises such as cervical smoothing exercises, neck splenius and respiratory muscle strength augmentation exercises and neck extension exercises.

CONCLUSION

This paper can be considered to contribute to literature as it reports in detail the physical therapy and orthopedic evaluations of the 53-year old patient with explanations of different treatment methods for Hirayama disease. Detailed physical and orthopedic evaluations should be made of these patients for the implementation of appropriate physiotherapy and rehabilitation programs.

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