CASE REPORT

Five Years of Follow-up after Posterior Cervical Fusion Surgery for Hirayama Disease : A Case Report

Kosuke Sugiura[1], Toshinori Sakai[1,2], Masatoshi Morimoto[1], Hiroaki Manabe[1], Fumitake Tezuka[1], Kazuta Yamashita[1], and Koichi Sairyo[1]

[1]Department of Orthopedics, Institute of Biomedical Sciences, Tokushima University Graduate School, Tokushima, Japan, [2]Department of Orthopedics, Tokushima Prefectural Miyoshi Hospital, Tokushima, Japan

Abstract : Background : Hirayama disease (HD) is characterized by slow progression of muscle atrophy without sensory disturbance in a single upper extremity in adolescent boys. HD can be treated using both conservative measures and surgery. However, the optimal treatment remains controversial. Case Presentation : We have encountered an 18-year-old man with HD who presented to us with a 2-year history of progressive muscle atrophy and weakness of the left upper extremity. He underwent posterior cervical fusion surgery in the extended position without decompression. As of 5 years postoperatively, there has been no deterioration of his muscular atrophy and weakness and his condition has mildly improved. He was able to return to daily life and work with no difficulty. Conclusions : Although cervical fusion surgery has several risks, including adjacent segment disease, posterior spinal fusion surgery without decompression in the short segment can be considered as a surgical option for HD. J. Med. Invest. 71 : 298-302, August, 2024

Keywords **:** *Hirayama disease, lower cervical spine, flexion position, muscle atrophy, posterior cervical fusion surgery*

INTRODUCTION

Hirayama disease (HD) is a rare but well-known disorder in South and East Asia, particularly in Japan. Typically occurring in adolescent boys and young men, HD is characterized by slow progression of muscle weakness and atrophy in a single distal upper extremity without sensory disturbance or pyramidal tract signs (1, 2). The underlying pathology in HD is suspected to be ischemic necrosis or atrophy of the anterior horn at the lower cervical levels due to compression by the posterior wall of the vertebral body or intervertebral disc during repeated cervical flexion (2, 3). A popular hypothesis for this pathological forward shift of the spinal cord is a difference in growth rate between the spinal column and dura mater, which leads to a forward shift of the posterior surface of the dura mater with tension in cervical flexion (4).

HD can be treated using conservative measures or by surgery. Typical conservative treatment consists of external fixation with an orthosis such as a neck collar (5, 6). Several surgical treatments, including anterior or posterior spinal fusion with/without decompression, laminoplasty, duraplasty, and combined procedures have been reported (1, 7-10). However, the optimal treatment remains controversial (11).

We have successfully treated an 18-year-old Japanese man with a diagnosis of HD by posterior cervical spinal fusion without a decompression procedure. In this report, we present the clinical results and magnetic resonance imaging (MRI) findings in this patient after 5 years of follow-up.

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Address correspondence and reprint requests to Kosuke Sugiura, MD, PhD, Assistant Professor, Department of Orthopedics, Institute of Biomedical Sciences, Tokushima University Graduate School, 3-18-15 Kuramoto-cho, Tokushima 770-8503, Japan and Fax : +81-88-633-0178. E-mail : sugiura_of_tokushima@yahoo.co.jp

REPORT OF THE CASE

History of present illness

An 18-year-old man with no significant past medical history noticed occasional tremor in the fingers of his left hand when he was a junior high school student. After entering high school, he complained of slowly progressive muscle weakness of the left upper extremity over a period of approximately 2 years. His family history included a diagnosis of dermatomyositis in his father and multifocal motor neuropathy in a sister that was treated by repeated intravenous immunoglobulin infusions.

Examinations

On first presentation, we noted mild muscle atrophy in his left ulnar side of forearm, hand, and shoulder girdle (Fig. 1). Neurological examinations revealed muscle weakness at the level of the C7-T1 myotomes in the left upper extremity but no obvious sensory disturbance. Although the brachioradialis and triceps tendon reflexes were hypoactive on both sides, there were no pyramidal tract signs in either lower extremity. Tremor was apparent in the fingers of the left hand in the extension position. Plain cervical radiographs showed decreased neck lordosis without abnormal instability of intervertebral joint (Fig. 2). Cervical magnetic resonance imaging (MRI) in the neutral position showed flattening of the spinal cord at the C5-C6 levels despite adequate sparing of the subdural space. However, cervical MRI in the flexion position revealed forward shift of the dura mater with an asymmetrically flattened spinal cord and swelling of the epidural venous plexus and flow void in the lower cervical spine (Fig. 3). A computed tomography myelogram showed forward shift of the dura mater, narrowing of the subdural space, and flattening of the spinal cord at the levels of C4-C5 to C6-C7 in the flexion position with slight kyphotic change at C5-C6 level (Fig. 4).

Based on these clinical manifestations and radiological findings, the diagnosis was HD. Although some studies have suggested that the symptoms of HD are likely to resolve spontaneously

over 3-4 years (5, 6), the patient was experiencing progressive muscle atrophy and hoped to undergo surgery to prevent further progression of his symptoms.

Surgical procedure

We performed posterior cervical spinal fusion without decompression at the C4-C6 vertebral levels using an autogenous iliac bone graft. Posterior cervical spinal fusion was performed in a mildly extended position under fluoroscopic guidance with the patient's head held by a Mayfield clamp. Screws were inserted in each lateral mass using the Roy-Camille technique (12). Bilateral rods were connected with a transverse connector (Fig 5).

Postoperative course

In the month following surgery, the patient's neck was immobilized using a soft collar. Thereafter, his clinical course was followed for 5 years. Although he continued to have atrophy of the hypothenar and dorsal interosseous muscles, and tremor in the fingers of the left hand in the extension position, there was improvement in his grip strength. Postoperative plain radiographs in cervical flexion position showed slight posterior angulation at the C6-C7 intervertebral level (Fig. 5). However, widening of the anteroposterior diameter of the spinal cord at the C5-C6 and C6-C7 intervertebral levels was apparent without the progression of disc degeneration in MRI at 5 years (Fig. 6). The patient suffers from no difficulty in daily life and work.

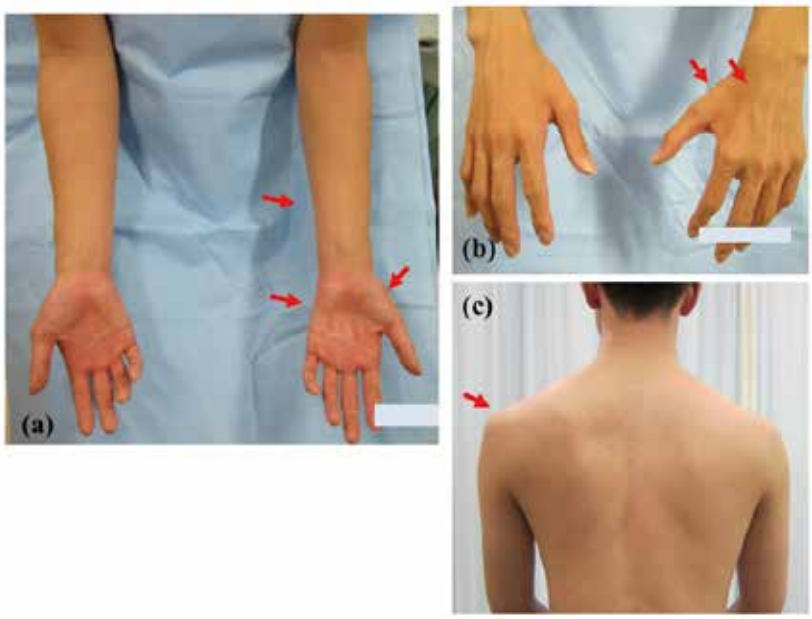

Fig. 1. Photographs showing unilateral muscle atrophy in the left upper extremity. (a) Ulnar regions of the forearm, thenar, and hypothenar muscles, (b) hand intrinsic muscles, and the (c) serratus anterior muscle. In each image, arrows show atrophied muscle.

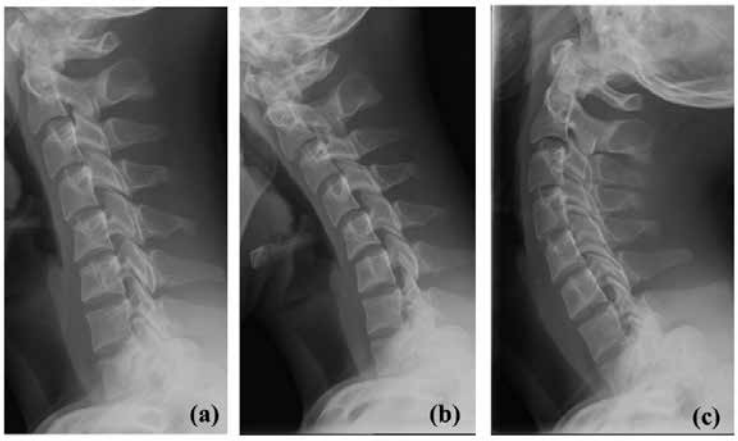

Fig. 2. Preoperative radiographs of the cervical spine. (a) Neutral, (b) flexion, and (c) extension positions showing decreased cervical lordosis without abnormal instability.

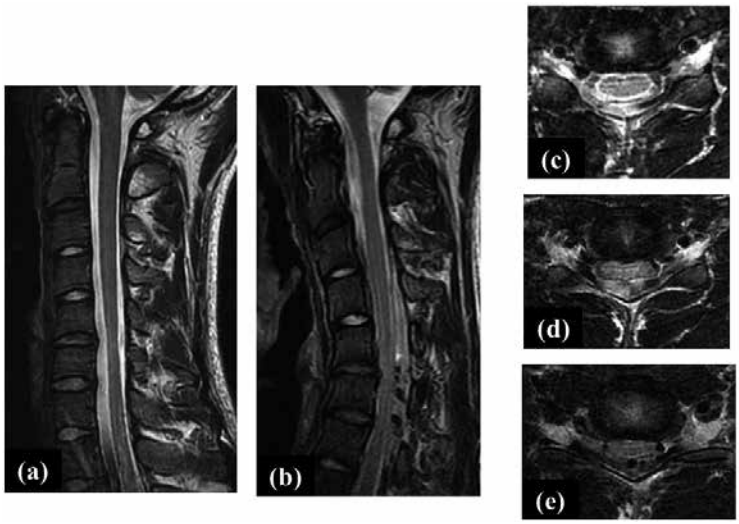

Fig. 3. Preoperative dynamic magnetic images of the cervical spine. (a) Neutral position, (b) flexion position, and axial views in the flexion position at the (c) C4-C5, (d) C5-C6, and (e) C6-C7 levels showing forward shift of the dura mater with an asymmetrically flattened spinal cord and swelling of the epidural venous plexus and flow void in flexion position.

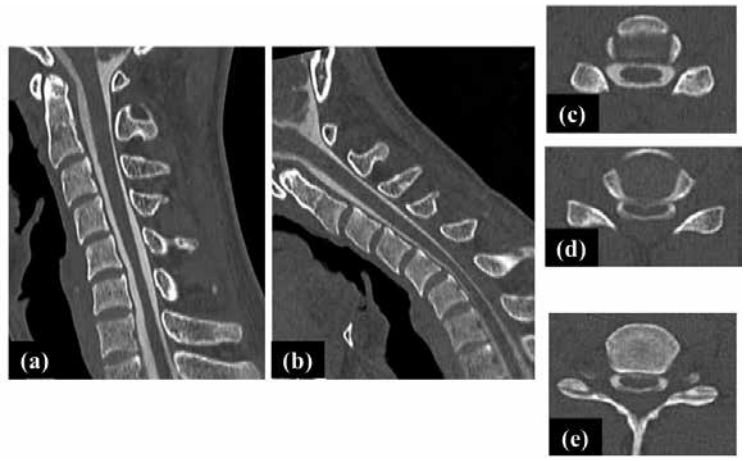

Fig. 4. Preoperative computed tomography myelogram of the cervical spine showing sagittal views obtained in a (a) neutral or (b) flexion position and axial views of (c) the C4-C5, (d) C5-C6, and (e) C6-C7 levels showing forward shift of the dura mater, narrowing of the subdural space, and flattening of the spinal cord at the levels of C4-C5 to C6-C7 in the flexion position.

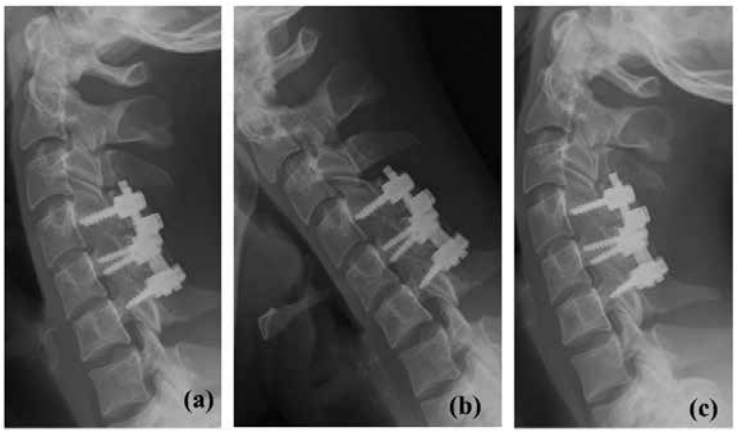

Fig. 5. Five years postoperative radiographs of the cervical spine. (a) Neutral, (b) flexion, and (c) extension positions showing bony fusion at C4-6 intervertebral levels and slight posterior angulation at the C6-C7 intervertebral level without abnormal instability and slippage.

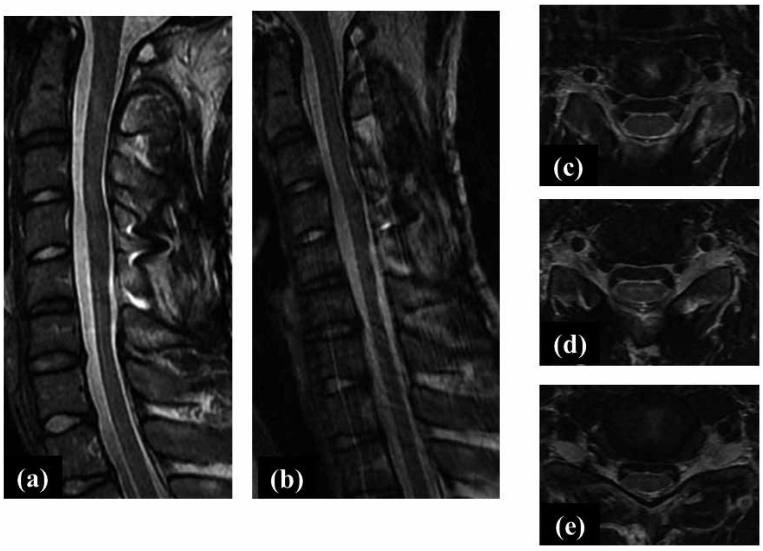

Fig. 6. Five years postoperative dynamic magnetic resonance images of the cervical spine. (a) Neutral position, (b) flexion position, and axial views in the flexion position at the (c) C4-C5, (d) C5-C6, and (e) C6-C7 levels showing widening of the anteroposterior diameter of the spinal cord without the progression of disc degeneration.

DISCUSSION

HD is a selective motor neuron disease in which there is ischemic necrosis or atrophy of the anterior horn cells in the spinal cord at the lower cervical levels due to repeated compression by the posterior wall of the vertebral body or intervertebral disc in cervical flexion (2, 3). The pathology of HD is thought to involve a forward shift of the spinal cord during flexion of the neck, arising from the difference in growth rate between the spinal column and dura mater during adolescence. The dura mater is occasionally tense and shifts forward in neck flexion when the growth rate of the dura mater is slower than that of the spinal column during growth periods (4). Therefore, an important component of treatment for HD is prevention of repeated compression of the lower cervical cord in neck flexion until the dura mater is adequately mature.

Both conservative and surgical treatments are indicated for HD. Typical conservative treatment includes external fixation of the neck with an orthosis such as a soft neck collar (5, 6). Although some reports have suggested that conservative treatment with a neck orthosis achieves good clinical outcome, we believe that prevention of repeated flexion stress is essential when treating patients with HD. However, young patients who wear a neck orthosis for a long period of time may lose their ability to perform activities of daily living.

Several surgical treatments, including anterior or posterior spinal fusion with or without decompression, laminoplasty, duraplasty, and combined procedures have been reported in the past (1, 7-10). Although the optimal treatment continues to be controversial (11), many operators choose anterior fusion or duraplasty and few choose posterior cervical fusion (1, 7-10). The possibility of adjacent segment disease (ASD) must be considered in young patients who undergo spinal fusion surgery. Reported risk factors for cervical ASD include age younger than 60 years, fusion adjacent to the C5-C6 and/or C6-C7 levels, presence of disc herniation and/or dural compression secondary to spinal stenosis with a mean anteroposterior diameter spinal canal of 13 mm or smaller, and anterior cervical fusion surgery at more than one level (13, 14).

In this case, we chose posterior cervical fusion surgery without decompression for the following reasons. First, posterior cervical fusion surgery allows earlier return to normal daily life without the need for a neck orthosis. By performing fusion surgery, the duration of wearing a neck orthosis was shortened. Second, as mentioned earlier, an important aim of treatment for HD is prevention of repeated compression of the spinal cord in neck flexion. Therefore, we believe that a decompression procedure, such as laminoplasty or duraplasty, is not always necessary if fusion surgery is performed. By performing fusion surgery alone, we were able to shorten the operation time and decrease some of the surgical risks, including those of epidural hematoma, spinal cord injury, and postoperative cerebrospinal fluid leak. Third, anterior procedures are associated with some critical complications, including major vascular injury, laryngeal edema, recurrent laryngeal nerve palsy.

Given that the majority of patients with HD are adolescent boys or young men and most of the pathological lesions are at the lower cervical levels, patients with HD who undergo cervical fusion would have some risk factors for ASD. In our case, although there was slight posterior angulation at the C6-C7 intervertebral level in the flexion position after C4-C6 fusion, MRI did not show progressive disc degeneration during 5 years of postoperative follow-up. Because there have been few reports of long-term follow-up such as 5 years after the posterior fusion surgery for HD, we consider this report to be valuable.

CONFLICTS OF INTEREST DISCLOSURE

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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