International Journal of Research in Medical Sciences
Kumar N et al. Int J Res Med Sci. 2014 Aug;2(3):1232-1235
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012
DOI: 10.5455/2320-6012.ijrms20140885

Case Report

Response of Hirayama disease to surgical intervention: case report

Navneet Kumar[1]*, Sumedha Mukherjee[2], Mukesh P. Patel[1], K. B. Shah[1], Sunil Kumar[1]

[1]Department of Neurosurgery, NHLMMC, VSGH, Ahmedabad, Gujarat, India
[2]Department of Anaesthesiology, DR BC ROY PGIPS, Kolkata, West Bengal, India

Received: 2 July 2014
Accepted: 13 July 2014

***Correspondence:**
Dr. Navneet Kumar,
E-mail: nvntkumar5@gmail.com

ABSTRACT

Hirayama disease also known as monomelic amyotrophy, primarily involves distal upper limb extremities. It differs from the known types of motor neuron diseases because of its nonprogressive behavior and pathologic findings of focal ischemic changes in the anterior horn of the lower cervical cord. We present a young male with Hirayama disease who had a left upper extremity involvement which was progressive in nature. He didn't respond with initial treatment of cervical collar. Consequently surgical intervention improves muscle weakness and decrease the neurological deficit.

Keywords: Hirayama disease, Monomelic amyotrophy, Juvenile muscular atrophy

INTRODUCTION

Hirayama Disease (HD) involves benign focal amyotrophy of the distal upper limbs and is often misdiagnosed as motor neuron disease. Even routine Magnetic Resonance Imaging (MRI) may be reported normal. Besides, due to the long plateau phase in the disease there is no definitive treatment modality. Limiting prolonged neck flexion, conservative physiotherapy and cervical collars are some of the temporary relievers with varying results. Surgical intervention may be a permanent solution when the above methods fail. It improves muscle strength and neurological deficits.

CASE REPORT

An otherwise healthy 21 year old man presented to us with insidious onset of weakness in the left hand for the past 4 years. He had fasciculations in his forearm and hand, along with a cramping sensation for the same duration. He showed wasting and atrophy of the same hand leading to disparity of both upper limbs. He denied having any complain regarding the other limb. He did not have any history of trauma or family history suggestive of neuromuscular disease.

On physical examination we found atrophy and weakness of the left volaris compartment with moderate atrophy of the left interossei dorsalis manus muscles. Spontaneous fasciculations were visible in the left flexor carpi ulnaris muscle. Deep tendon reflexes were normal. Sensation to pin prick, vibration and joint position were intact. The patient did not demonstrate any extra pyramidal signs, ataxia, sensory disturbances or abnormalities in sweating or urination. Motor conduction study showed reduced CMAP amplitude of left median and ulnar nerves with normal distal latency and conduction velocity. F wave study showed prolonged latency of left median and ulnar nerves. Sensory studies showed normal parameters of bilateral median, ulnar and sural nerves. Needle EMG studies from bilateral FDI showed abnormal spontaneous activity in the form of fasciculation potentials with neurogenic MUAPs and full recruitment pattern. MRI cervical spine flexion and extension showed anterior shifting of posterior Dural canal at lower cervical spinal cord. This resulted in compression of the cord in flexion

movement of the neck whereas in neutral position there was no cord compression. These findings pointed to flexion myelopathy. We initially tried cervical collar due to the stationary nature of the disease. But as the disease progressed we decided for a surgical intervention. Patient underwent laminectomy from C4 to C6 with posterior pedicular screw fixation

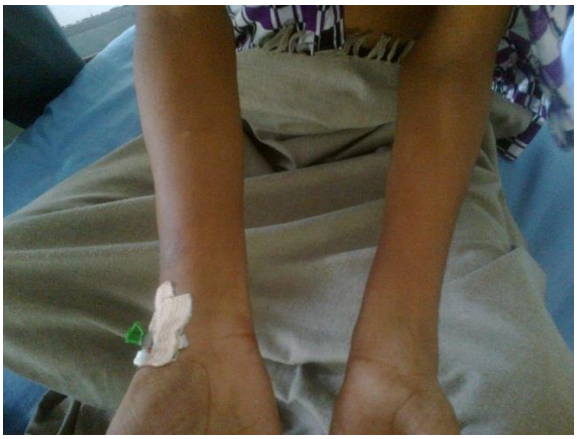

Figure 1: Atrophy & wasting of left upper extremity.

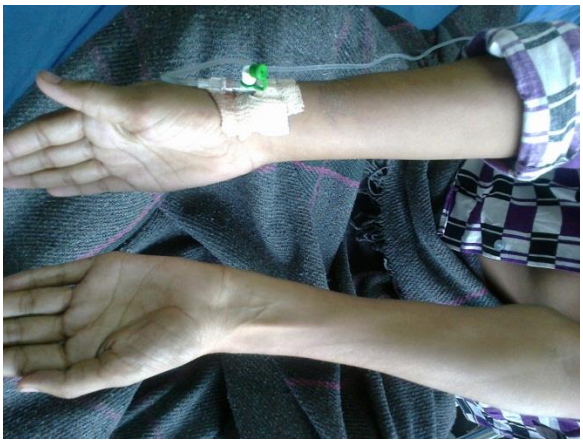

Figure 2: Atrophy & wasting of volar aspect and ulnar side of forearm.

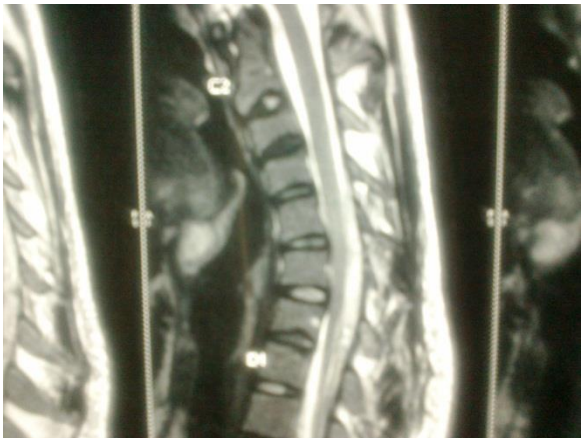

Figure 3: MRI scan of cervical spine in flexion showing anterior shifting of Dura.

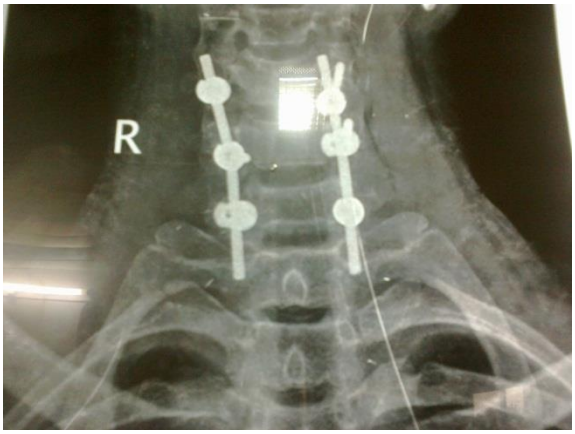

Figure 4: Posterior pedicular screw fixation of cervical spine.

DISCUSSION

We inferred from the history, physical examination and radiological findings that our patient could have a restricted form of motor neuron disease (monomelic muscular atrophy), a limited form of multifocal motor neuropathy, and chronic left sided radiculopathy. We easily ruled out any traumatic or ischemic insult causing focal spinal cord atrophy by eliciting a detailed history from the patient.

In a young patient with clinical evidence of hand and/or forearm muscle atrophy and asymmetric thinning of the cervical spinal cord, a diagnosis of Hirayama disease (juvenile muscular atrophy of the distal upper extremity) should always be considered. Hirayama, a benign focal amyotrophy of the upper limbs commonly occurs in young males between the age of 15-25 years.[1] The pattern of forearm involvement is also referred to as an oblique amyotrophy as brachioradialis muscle is spared.[1] There is unilateral involvement in the majority of patients, but asymmetric and symmetric bilateral involvement are also observed. Our patient (21 years) had primarily a left forearm involvement. The disease has an insidious onset and remains in a plateau phase for a considerable period. A slowly progressive variant of the disease is known as the O Sullivan MacLeod syndrome. The patients commonly complain of painless wasting of upper limbs with hand tremors and loss of fine motor control movements. Our patient had a similar manifestation.

Detection of focal spinal cord atrophy in such a patient should prompt one to perform dynamic evaluation of the cervical spine with flexion and extension. Though radiographic views may be normal, forward migration of the posterior surface of the Dura mater with compression of the spinal cord obtained with flexion of the cervical spine is a characteristic of Hirayama disease. Our MRI findings corroborated with the above findings.

The pathogenesis of this entity is debated. Tokumaru concluded that dynamic spinal cord compression at neck

flexion with forward displacement of the posterior Dura is an unequivocal finding in the progressive stage of this disease.[1] As this finding is absent in elderly patients; it suggests that dynamic compression of the spinal cord may be an important finding in the diagnosis of this disease. Kikuchi et al. believe that a "tight" Dural canal during flexion of the neck is due to a disproportional length between the vertebral column and the Dural canal.[2,3] During neck extension, the normal cervical Dura mater is slack and consists of transverse folds. During neck flexion, the length of the cervical canal increases. In healthy subjects, the Dural slack compensates for the increased length in flexion and stays in apposition with the bony canal. Patients with Hirayama disease, however, may have an imbalance in growth of the vertebrae and the Dura mater. This results in an anterior shift of the posterior Dural wall, thereby causing spinal cord compression.

The mechanism of myelopathy may involve ischemic changes or chronic trauma inflicted by repeated neck flexion. In an autopsy study of a patient with this disease, ischemic changes were demonstrated in the anterior horn cells, along with asymmetric spinal cord thinning.

There is no definitive treatment for this disease. The primary goal is to limit neck flexion. Posture with long term neck flexion is to be avoided. The use of low pillows is recommended. Ando T et al. found conservative physiotherapy helpful in a 16 year male with reversible muscular weakness of right proximal upper limb.[4]

A neck collar is generally advised for 3-5 years. Several studies recommend the early use of cervical collar to arrest the progression of the disease as it has a long stationary phase. Tokumara et al. conducted a study where they prescribed cervical collars in 14 patients while the rest 18 were allowed to follow natural course of the disease. They concluded that the muscle weakness of the affected limb improved and the disease progression was negligible in cervical collar group. Tokumara studied 38 cases where he found premature arrest of the disease when cervical collars were used early in the disease.[5,6] Our patient had used neck collar for 12 months but the disease was progressing. So we decided to go in for a surgical intervention.

Surgical intervention has been a choice of treatment when disease worsens despite conservative therapy or cervical collar usage. Imamura H et al. successfully treated a 16 year Hirayama male by anterior C5 vertebrectomy followed by fixation of C4-6 vertebral bodies using iliac bone and plate system.[7] The patient showed marked improvement of muscle strength within six months. They proposed that surgical intervention causes permanent stable fixation with much shorter period of external cervical immobilization as long term collar therapy may be unbearable in most cases. It also improves neurological deficits leading to better quality of life. Masaki T et al. did a spinal immobilization by wiring and bone graft implantation of C2-6 vertebrae to limit the range of anterior nuchal flexion in a case of Hirayama disease.[8] This resulted in improvement of muscle strength along with disappearance of dysthesia on neck flexion. Kohno et al. surgically treated five young males with cervical flexion myelopathy.[9] Three of them underwent anterior decompression and fusion of the cervical spine with long bone grafting. Rest two had posterior fusion of five laminas (C3-7) using Rogers method. They concluded that anterior method is preferred as it allows the removal of anterior structures (vertebral bodies, intervertebral discs or osteophytes) which compresses cervical spine in flexion condition. Posterior approach is advocated where wide range of anterior cervical structures need to be decompressed. But both results in improvement of muscle weakness and decrease of sensory disturbances.

CONCLUSION

Hirayama disease though have a stationary phase, surgical intervention have shown to improve the neurological deficits and muscle strength leading to better quality life. Surgery seems to be beneficial for patients who do not respond to conservative treatment, as this gives permanent stable fixation with much shorter period of external cervical immobilization compared with cervical collar therapy in which long-term application is frequently unbearable in many patients.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Hirayama K, Tokumaru Y. Cervical Dural sac and spinal cord in juvenile muscular atrophy of distal upper extremity. Neurology. 2000;54:1922-6.
2. Kikuchi S, Tashiro K, Kitagawa K, Iwasaki Y, Abe H. A mechanism of juvenile muscular atrophy localized in the hand and forearm (Hirayama's disease): flexion myelopathy with tight dural canal in flexion (in Japanese). Clin Neurol (Tokyo). 1987;27:412-9.
3. Kikuchi S, Tashiro K, Kitagawa K, Iwasaki Y, Abe H. A mechanism of juvenile muscular atrophy localized in the hand and forearm (Hirayama's disease): flexion myelopathy with tight dural canal in flexion. Rinsho Shinkeigaku. 1987;27:412-9.
4. Ando T, Fukatsu H, Kameyama T, Takahashi A, Yamada H. A case of flexion myelopathy presenting with reversible muscular weakness and atrophy of the unilateral proximal upper limb Rinsho Shinkeigaku. 1993 May;33(5):575-8.
5. Tokumaru Y, Hirayama K. Cervical collar therapy for juvenile muscular atrophy of distal upper extremity (Hirayama disease): results from 38 cases. Rinsho Shinkeigaku. 2001;41:173-8.

6. Tokumaru Y, Hirayama K. A cervical collar therapy for nonprogressive juvenile spinal muscular atrophy of the distal upper limb (Hirayama's disease). Clin Neurol (Tokyo). 1992;32:1102-6.
7. H. Imamura, S. Matsumoto, M. Hayase, Y. Oda, H. Kikuchi, M. Takano. A case of Hirayama's disease successfully treated by anterior cervical decompression and fusion. Brain Nerve. 2001;53(11):1033-8.
8. Masaki T, Hashida H, Sakuta M, Kunogi J. A case of flexion myelopathy presenting juvenile segmental muscular atrophy of upper extremities: a successful treatment by cervical spine immobilization. Rinsho Shinkeigaku. 1990 Jun;30(6):625-9.
9. S. Konno, S. Goto, M. Murakami, M. Mochizuki, H. Motegi, H. Moriya. Juvenile amyotrophy of the distal upper extremity: pathologic findings of the Dura mater and surgical management. Spine. 1997;22(5):486-92.

DOI: 10.5455/2320-6012.ijrms20140885
Cite this article as: Kumar N, Mukherjee S, Patel MP, Shah KB, Kumar S. Response of Hirayama disease to surgical intervention: case report. Int J Res Med Sci 2014;2:1232-5.