International Journal of Research in Orthopaedics
Palla A et al. Int J Res Orthop. 2017 Jul;3(4):886-889
http://www.ijoro.org

Case Report

DOI: http://dx.doi.org/10.18203/issn.2455-4510.IntJResOrthop20172891

Hirayama disease: a rare neurological condition of cervical cord pathology

Abhilash Palla*, Vinay Jain K., Naveen Kumar L., Rangaswamy B. T.

Department of Orthopaedics, M S Ramaiah Hospitals, Bengaluru, Karnataka, India

Received: 04 May 2017
Accepted: 30 May 2017

***Correspondence:**
Dr. Abhilash Palla,
E-mail: pallaabhilash@gmail.com

ABSTRACT

The disease is characterized by a progressive muscle weakness and wasting of distal upper limb muscles sparing brachioradialis. The pathology is in the lower cervical cord due to forward displacement of dural sac and spinal cord by neck flexion. Repeated neck flexion may result in ischemia of the lower cervical anterior horns. The diagnosis can be confirmed by MRI, EMG. It requires a high degree of suspicion in a young patient presenting with unilateral upper limb distal muscle weakness with no sensory involvement, as early diagnosis will help in preventing the progression of the disease by employing simple measures like usage of a cervical collar. We report a case of a 25 year old man presenting with progressive distal upper limb muscle wasting, on investigating further diagnosed to have Hirayama disease.

Keywords: Hirayama, Muscular atrophy, Cervical cord

INTRODUCTION

Hirayama disease is a rare neurological condition characterized by muscular atrophy of upper extremities unilaterally. It is sporadic, seen in early twenties with a male preponderance, commonly seen in the Asian countries. It predominantly affects the lower cervical cord. The weakness has an insidious onset and gradual progression for initial couple of years followed by a plateau of muscular atrophy, but no sensory or autonomic involvement.[1]

CASE REPORT

A 25 year old man presented with a 2 year history of slowly progressive weakness of left hand and forearm following a fall from bike. He is not able to lift heavy objects and does not have a good grasp of the left hand. He has neck pain following the fall. No sensory impairment or involvement of any other limb. No dysphagia, diplopia, weakness of lower limbs or bowel & bladder involvement. No significant past history or exposure to toxins or intake of drugs or any allergies. No significant family history.

On examination, left forearm and hand muscles wasting (Figure 1 and 2) was noted with sparing of brachio-radialis. Thumb abduction (Figure 3) and palmar grasp impaired. No autonomic features. Proximal muscles were normal and no cranial nerve, cerebellar or posterior column involvement.

Figure 1: Dorsum of both hands showing wasting of intrinsics.

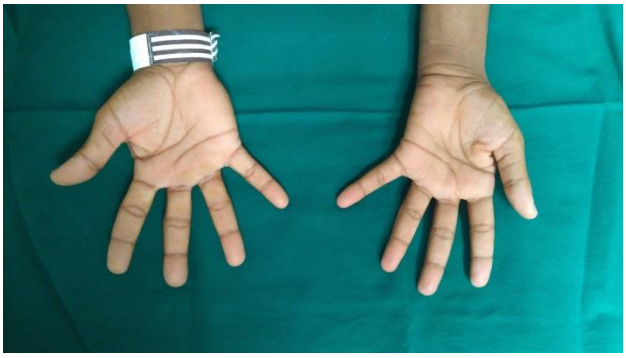

Figure 2: Palmar aspect of both hands showing wasting of intrinsics, thenar and hypothenar eminences.

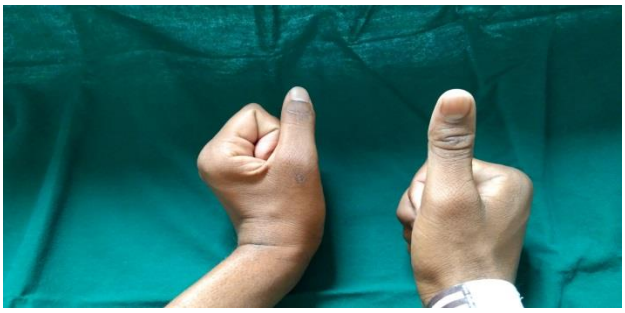

Figure 3: Inability to abduct left thumb.

Cervical spine X-rays showed no gross abnormality (Figure 4 and 5). Multiplanar T1, STIR, GRE and T2 sequences through the cervical spine imaged in neutral, flexion and extension of the neck show atrophy and flattening of the spinal cord at C6 and C7 levels (lower cervical cord atrophy) with a minimum AP diameter of about 4.5 mm. The rest of the cord is normal in morphology and signal intensity. But there is no evidence of widening of the posterior epidural space on flexion manoeuvre. Rest of the vertebral structures, joints, disc, ligaments were normal (Figure 6 and 7).

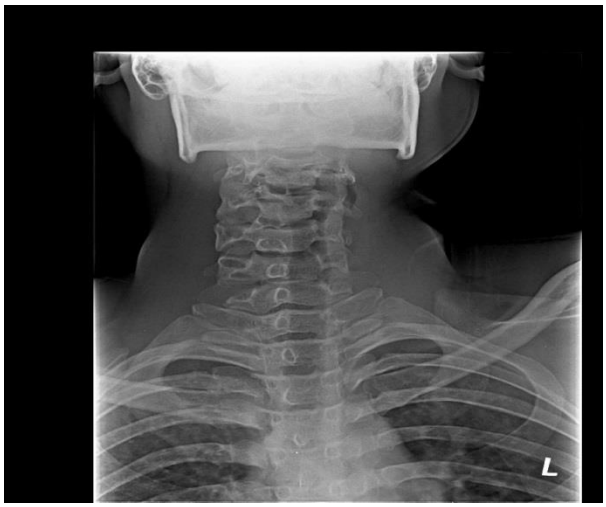

Figure 4: X-ray AP view of cervical spine.

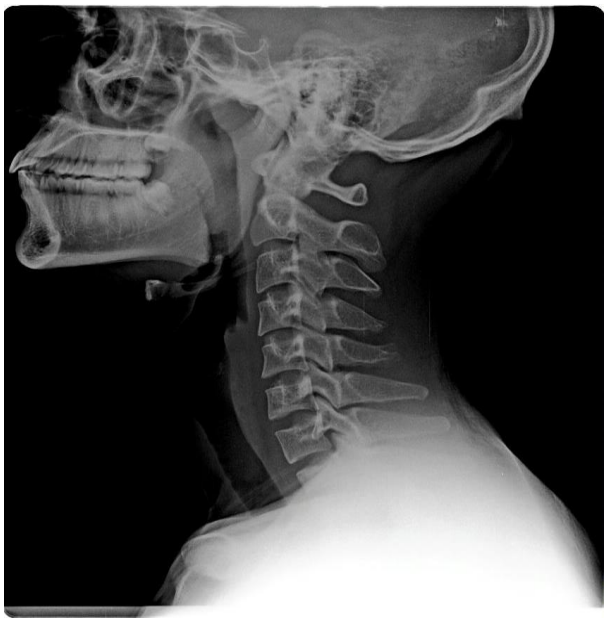

Figure 5: X-ray lateral view of cervical spine.

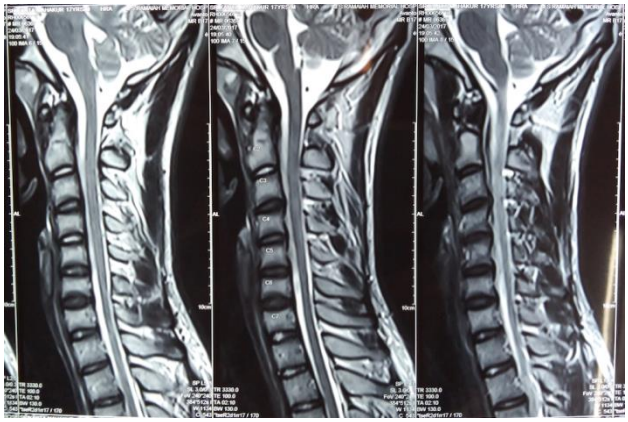

Figure 6: Sagittal cuts of C-spine MRI.

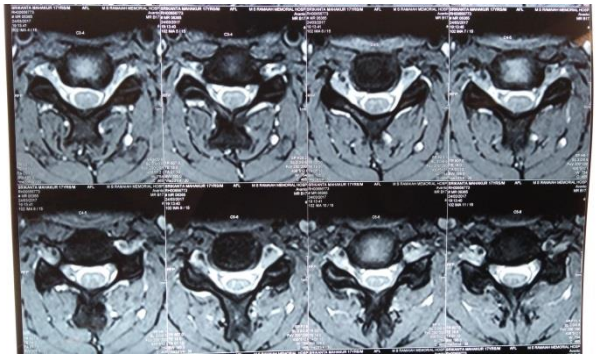

Figure 7: Axial cuts of C-spine MRI.

Nerve conduction study was performed on bilateral median, radial, ulnar, common peroneal and posterior tibial nerves. Motor study showed reduced CMAP amplitude and normal motor conduction velocity on left median and ulnar nerves. Mildly reduced CMAP amplitude in left radial nerve. Normal distal motor

latency in all the nerves. Sensory latency, SNAP amplitude and sensory conduction velocity was normal in all the nerves. Absent F-wave on left median and ulnar nerves. Thus giving an impression of axonopathy of left median and ulnar nerves.

Electromyography by using concentric disposable needle was done on bilateral first dorsal interossei and left triceps, biceps brachii and brachioradialis muscles. Left first dorsal interosseus and triceps muscles show spontaneous activities in the form of positive sharp waves with occasional fibrillations, large amplitude motor unit potentials and reduced recruitment with incomplete interference pattern, suggesting neurogenic pattern in left C7, C8 and T1 innervated muscles.

Complete blood counts, renal and hepatic function tests, acute phase reactants were within normal range and serological tests were negative.

DISCUSSION

Hirayama et al in 1959 reported 12 patients of this disease, who were clinically distinguished from other progressive motor neuron diseases like amyotrophic lateral sclerosis, spinal progressive muscular atrophy.[1] It is a benign cervical myelopathy associated with neck flexion.[2]

It is commonly seen between 15 and 25 year old males, characterized by insidious onset of asymmetrical weakness and wasting of upper limb muscles in C7, C8 and T1 myotomes with a progression in the initial couple of years followed by a plateau. The features are like minipolymyoclonus aggravated by cold temparatures. Cranial nerve examination, autonomic system, Cerebellar functions, pyramidal tracts of lowerlimbs are usually normal. No sensory or reflex involvement.[3] Brachioradialis is generally spared.[4]

The exact pathogenesis is still a matter of debate. But it is postulated to be associated with a tight dural canal during neck flexion.[5] The short and tight duramater seen in these patients cannot cope up with the stretching of the vertebral canal during neck flexion. Hence on neck flexion, it tightens and shifts anteriorly causing canal compression.[6] Repeated neck flexion in the initial stages causes episodes of ischemia resulting in chronic myelopathy evidenced by asymmetric lower cervical cord thinning.[7] The pathological study conducted by Hirayama et al showed a circulatory insufficiency of the anterior horn cells of the spinal cord particularly at C7 and C8 levels.[8]

Imaging findings of MRI particularly are important in diagnosing this disease.[9]

The differential diagnoses include spinal muscular atrophy, amyotrophic lateral sclerosis, post-polio syndrome, multifocal motor neuropathy with conduction block, toxic neuropathy and syringomyelia. They can be differentiated by their clinical, radiological and electrophysiologic features.[1]

Hirayama disease is a self-limiting disorder and no published consensus on definitive treatment. But early diagnosis helps in preventing progression by limiting neck flexion by instituting the usage of a cervical collar. Physiotherapy helps in preventing joint stiffness and muscle wasting.[10] Certain studies have advocated surgical interventions like decompression and fusion in selective patients with anterior effacement and severe cervical kyphosis on MR imaging during knee flexion and have shown promising results.[9].

CONCLUSION

Hirayama disease requires a high degree of suspicion in a young patient presenting with unilateral upper limb distal muscle weakness with no sensory involvement, as early diagnosis will help in preventing the progression of the disease by employing simple measures like usage of cervical collar.

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

REFERENCES

1. Hirayama K, Tsubaki T, Toyokura Y, Okinaka S. Juvenile muscular atrophy of unilateral upper extremity. Neurol. 1963;13(5):373.
2. Sitt JC, Fung EL, Yuen EH, Ahuja AT. Hirayama disease in a 17-year-old Chinese man. Singapore Med J. 2014;55(6):87.
3. Hassan KM, Sahni H, Jha A. Clinical and radiological profile of Hirayama disease: a flexion myelopathy due to tight cervical dural canal amenable to collar therapy. Annals Indian Acad Neurol. 2012;15(2):106.
4. Nascimento OJ, Freitas MR. Non-progressive juvenile spinal muscular atrophy of the distal upper limb (Hirayama's disease): a clinical variant of the benign monomelic amyotrophy. Arquivos de neuro-psiquiatria. 2000;58(3):814-9.
5. Chen CJ, Chen CM, Wu CL, Ro LS, Chen ST, Lee TH. Hirayama disease: MR diagnosis. American J Neuroradiol. 1998;19(2):365-8.
6. Kikuchi S, Tashiro K, Kitagawa M, 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. Clin Neurol. 1987;27(4):412-9.
7. Gandhi D, Goyal M, Bourque PR, Jain R. Case 68: Hirayama Disease 1. Radiology. 2004;230(3):692-6.
8. Hirayama KE, Tomonaga MA, Kitano K, Yamada T, Kojima S, Arai K. Focal cervical poliopathy causing juvenile muscular atrophy of distal upper

extremity: a pathological study. J Neurol, Neurosurg Psychiatry. 1987;50(3):285-90.
9. Lin MS, Kung WM, Chiu WT, Lyu RK, Chen CJ, Chen TY. Hirayama disease: clinical article. Journal of Neurosurgery: Spine. 2010;12(6):629-34.
10. Yoo SD, Kim HS, Yun DH, Kim DH, Chon J, Lee SA, et al. Monomelic amyotrophy (Hirayama disease) with upper motor neuron signs: a case report. Annals of rehabilitation medicine. 2015;39(1):122-7.

Cite this article as: Palla A, Vinay Jain K, Naveen Kumar L, Rangaswamy BT. Hirayama disease – a rare neurological condition of cervical cord pathology. Int J Res Orthop 2017;3:886-9.