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Original Article

Radiology Section

A Dynamic MR Study of the rare Hirayama's Disease: Does Flexion Acquisition Preclude the Need of IV Contrast Study?

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ABSTRACT

Introduction: Hirayama disease (HD), is a self limiting, non hereditary, uncommon spinal cord disease affecting the forearms and hands of one or both upper limbs, due to involvement of lower cervical cord. The definitive diagnosis of Hirayama is by MRI in correlation with clinical findings and electromyography.

Aim: The aim of the study was to adequately demonstrate all contributory findings to the diagnosis of HD by an MRI with neutral and flexion acquisitions. Another objective was to define and establish a standard protocol for adequate flexion technique. Lastly, to demonstrate that if adequate flexion is achieved, contrast administration becomes unnecessary.

Materials and Methods: Twenty patients of clinically suspected HD presented for an MRI examination were studied. A 1.5T Philips Multiva MRI system was used. MRI was done in both neutral and flexion acquisitions and the results analysed.

Results: The younger patients in their late teens and early twenties showed a progressive disease, while older patients between the ages of 27 to 34 showed a near static disease.

There was a 100% positive predictive value in flexion MRI in demonstrating all the contributory findings. Neutral MRI showed loss of attachment of posterior dura in 45% cases. Cord thinning and flattening were present in all of our patients in both neutral and flexion acquisitions.

Conclusion: The distinctive conclusion from this study was defining the degree and technique of flexion required to demonstrate the falling forward of the dura. By achieving this described degree of flexion, the use of intravenous Gadolinium administration is deemed unnecessary and redundant.

Keywords: Cervical cord thinning, Cord flattening, Laminodural space, Loss of attachment of dura, Prominent epidural flow voids

INTRODUCTION

Hirayama disease (HD), is a self limiting, non hereditary, uncommon spinal cord disease affecting the forearms and hands of one or both upper limbs, due to involvement of lower cervical cord. Studies have shown that it develops in the late adolescence and early twenties with a definite male preponderance.

It was in Japan way back in 1959 that Hirayama et al. reported this disease as "juvenile muscular atrophy of unilateral upper extremity" [1].

Reviewing the literature, it is seen that this disease has been labeled differently by different researchers as "juvenile muscular atrophy of the distal upper extremity, juvenile asymmetric segmental spinal muscular atrophy, and benign focal amyotrophy or monomelic amyotrophy" [2,3].

The typical clinical features comprise a subtle onset and slow but steady progression of muscular atrophy, which could involve one or both upper limbs, with weakness of the forearms and hands. There is a predominant motor involvement in the lower motor neuron type. Sensory symptoms or upper motor neuron symptoms such as hyperreflexia and hypertonia are very uncommon presentations [4].

The symptoms and findings in HD are often confused with those of a closely mimicking Motor Neuron Disease (MND) [4]. The difference is, HD though initially progresses it undergoes a spontaneous arrest after a few years, MND on the other hand is a progressive disorder. This rare disease is more reported from Asian countries.

Clinically, there is asymmetric muscle weakness and muscle wasting in the C8-T1 distribution [5]. Typically, young males develop progressive loss of grip in the involved upper limb that later stabilises [6]. It takes several years for the progress to stop and the disease to stabilise.

The exact developmental methodology is yet to be established. Several hypotheses have been put forth, but the popular theory postulates that insufficient growth of the dura relative to the spinal column during late childhood and early adolescence leads to forward displacement of the dura in flexion, the dura may compress the posterior spinal cord, possibly leading to ischemia of the anterior horn cells at C8 and T1.

MATERIALS AND METHODS

This cross-sectional observational study was performed after obtaining consent from the Institutional review board and clearance from the Institutional ethics committee.

Twenty patients of clinically suspected Hirayama's disease presented for an MRI examination at the Department of Radiodiagnosis at Saveetha Medical College over a period of two years between April 2016 to May 2018, were included.

An informed written consent was obtained from all the patients before performing the study. All patients presenting with neck pain, loss of grip in one or both upper limbs, numbness in either or both upper limbs, with wasting of the small muscles of hand were included in this study.

Though there was no age restriction in our study, we saw that only young male patients between the age of 18 to 34 years presented with the required symptoms of neck pain, loss of grip in one or both upper limbs, numbness in either or both upper limbs, with wasting of the small muscles of hand.

Patients with known contraindications to MRI such as incompatible hardware such as pacemakers and Claustrophobia were excluded from the study.

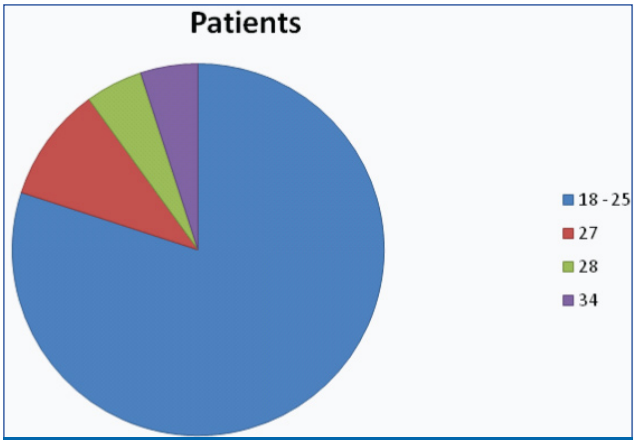
MRI was done using a 1.5 Tesla, Philips Multiva System. MRI cervical spine in neutral position was done using a T2 sagittal, T1 sagittal and T2 axial sequences. The patient was then asked to flex the neck. We provided adequate support to the back of neck, adequate flexion was when the patient's chin was in contact with the patient's anterior chest wall. To achieve this, the head coil was removed. Otherwise such extreme flexion cannot be obtained, which can then lead to false negatives. Though there is compromise in image resolution if the coil is removed, this degree of flexion is required to demonstrate the falling forward of the dura, by which means we avoided the use of unnecessary intravenous Gadolinium administration in all 20 of our patients.

Out of our 20 patients, eight of them had a unilateral hemicord involvement and the other 12 had a bilateral cord involvement, with some asymmetrical increased severity.

RESULTS

The results were tabulated based on patient demographics including age and height of the patients, the MRI findings in neutral acquisitions and finally the MRI findings in flexions acquisitions. These were then statistically analysed.

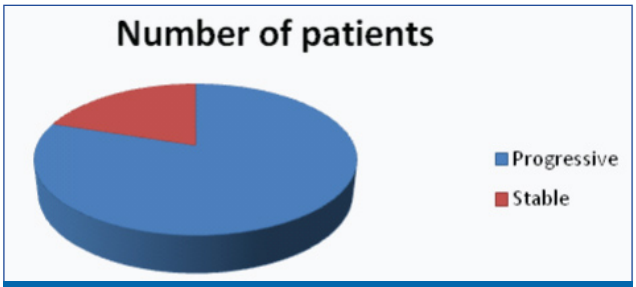
[Table/Fig-1] shows that 16 out of our 20 patients presenting to us with suspected HD were between 18 to 25. Two patients were 27 years old, one was 28 and one patient was above 34.



[Table/Fig-1]: 80% of patients were between the ages of 18 to 25. Only 4 patients were older between 27 to 34.

Height wise distribution of patients: all patients in our study group were tall and thin with long necks. The patients ranged between 175cms to 186cms in height.

Disease status of patient [Table/Fig-2]: 16 of our patients had a mild but steady progression of symptoms. Four of them had a stable disease, that is though there is no resolution of already existing numbness and weakness, there is no further progression in any of the symptoms. It is interesting to note that all these four patients with stable disease were in the older age group, which is 27 to 34.



[Table/Fig-2]: 16 patients (80%) had a progressive disease, with 4 patients showing arrest of progression.

Neutral acquisition: The MRI findings in routine neutral cervical spine positions were tabulated under the headings of lordosis state, loosening of posterior dura, cord thinning and flattening, and cord T2 hyperintensities [Table/Fig-3].

Flexion acquisition [Table/Fig-4]: The MRI findings in flexion acquisitions of the study patients were tabulated as falling forward of dura, prominence of posterior epidural flow voids and posterior cord indentation by the dura, with loss of CSF space.

It is significant to note that there was a 100% positive predictive value in flexion acquisitions in demonstrating all the necessary findings to establish the diagnosis of HD.

Finding in neutral cervical spine MRI	Number of patients	Percentage of patients
Loss of lordosis or reversed cervical lordosis	20	100%
Mild loosening of posterior dura in neutral position itself	9	45%
Cord thinning and posterior cord flattening at lower cervical levels (C5 to C7)	20	100%
Cord T2 hyperintensities (Gliosis)	18 (unilateral 8, bilateral 10)	90%

[Table/Fig-3]: MRI findings in neutral position.

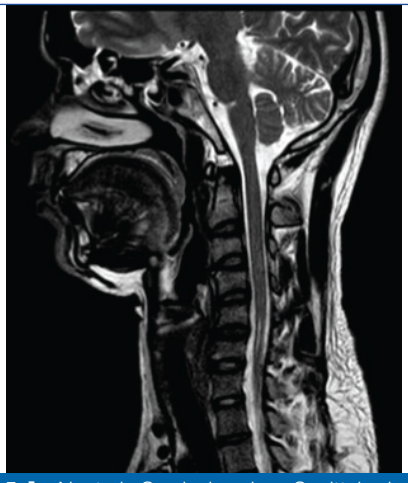
Finding in flexion cervical spine MRI	Number of patients	Percentage of patients
Significant falling forward of dura	20	100%
Prominence of epidural T2 flow voids	20	100%
Dural indentation of posterior cord, with loss of anterior and posterior CSF space	20	100%

[Table/Fig-4]: MRI findings in flexion acquisitions.

Representative Cases

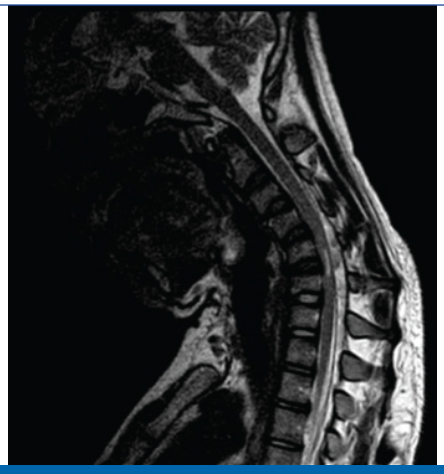
Bilateral cord involvement: A 27 year old male patient came with a complaint of progressive weakness and wasting involving the bilateral hand and forearms. The symptoms however had stabilized over the past six months. The hand weakness limited several activities of his daily living. All other neurological history was negative, including the sensory examination.

Imaging Findings: On neutral position, there is mild reversal of cervical lordosis, with thinning of cord with linear cord T2 hyperintense signal involving the spinal cord (mid to bilateral paramedian, more on the left), from C5 inferior end plate to C6 inferior endplate [Table/Fig-5a].



[Table/Fig-5a]: Neutral Cervical spine Sagittal showing cord thinning, flattening and increased T2 signal at C5 to C7 level.

On flexion acquisitions, there is significant falling forward of the posterior dura from C3 mid body to the upper dorsal level (D3). There is prominence of the posterior epidural flow voids. The anteriorly displaced dura along with the epidural flow voids is seen indenting the posterior cord, more on the left, with loss of anterior and posterior CSF space [Table/Fig-5b].

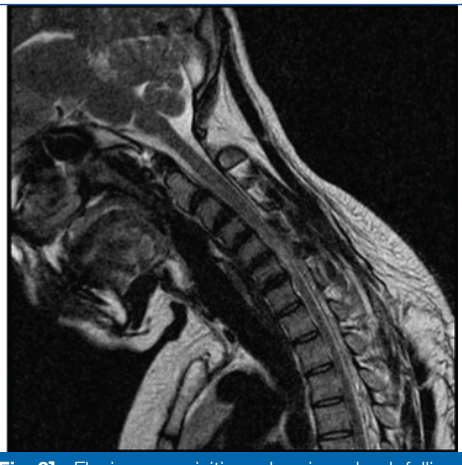


[Table/Fig-5b]: Flexion Cervical spine Sagittal showing falling forward of the dura, prominence of epidural flow voids and posterior cord indentation by the dura.

Unilateral involvement: A 21-year-old Indian man presented with a 3 years' history of slowly progressive weakness and muscle wasting in the right hand.

On neutral acquisitions, there is thinning of right cord with linear cord T2 hyperintense signal seen involving the right side spinal cord from C5 superior end plate to C7 superior endplate.

On flexion acquisitions, there is significant falling forward of the posterior dura from C3 mid body to the upper dorsal level (D4). There is prominence of the posterior epidural flow voids, and loss of visualization of anterior and posterior subarachnoid spaces [Table/Fig-6].



[Table/Fig-6]: Flexion acquisition showing dural falling forward, prominent posterior epidural flow voids and posterior right cord indentation

DISCUSSION

HD is an extremely rare form of cervical myelopathy [7]. It is a self-limiting, unilateral or asymmetrically bilateral, indolently progressive weakness and wasting of the forearms and hands. Hirayama almost exclusively occurs in young males. The anterior displacement of the posterior dura of the lower cervical spinal canal during extremes of neck flexion has been said to cause posterior cord indentation by the falling dura, leading on to cervical cord atrophy, consequent gliosis of anterior horn cells and asymmetric flattening [8-10].

What is characteristic about this disease is though there is a slow sluggish onset of symptoms, there is definite progression for a few years (averaging around 2 to 3 years). The disease then comes to a self limiting stop and the symptoms stabilize. It is important to note that there is no regression of the already existing symptoms and imaging findings. But there is no further progression. Hence it follows a relatively gentle course thereafter.

The diagnosis of Hirayama is essentially by MRI. Electromyography (EMG) can play an adjunct role in the diagnosis by demonstrating denervation findings in the involved group of muscles. [11,12].

Previously, MRI used to be performed with intravenous Gadolinium administration. In the present study, it is demonstrated that if a proper flexion technique is applied, all necessary findings required to establish the diagnosis of HD definitively, are well demonstrated in all patients. This is clinically significant since the MRI technique is less time consuming and describing the degree of flexion helps in achieving a uniform standard technique.

Majority of the patients were in their late teens and early twenties. This correlated well with prior studies. What was striking is the fact that the patients between the ages of 18 to 25 had a progressive disease at the time of the examination. The patients who were older were in the stabilized state at the time of the examination. This again re-affirms the hypothesis that the disease is initially progressive for a varying number of years and then there is arrest of progression [13].

All the patients in our study were male patients. This is again consistent with the previous studies [13].

An additional significance was all the patients were in the taller range with long necks. Though the importance of this fact is yet to be conclusively established, a long neck is more prone to increased degrees of flexion and this during puberty could have contributed to the disproportionate growth of the dura to the spine. All the study patients being tall is a contributory factor to this hypothesis. Furthermore, all the patients in neutral cervical spine MRI acquisitions had a loss of lordosis; a few even had a reversed lordosis [14,15]. Again more propensity to neck flexion in this group of study patients needs to be thought of.

Cord thinning and flattening in the lower cervical canal in neutral acquisitions was a constant finding in all the patients. Increased T2 signal in the thinned cord was present in 90% of the patients. When this finding is present with the appropriate clinical symptoms, the possibility of HD needs to be immediately considered and the patient taken up for flexion studies, even in the absence of a clinical diagnosis.

A considerable (45%) number of patients showed a loose dura (loss of attachment) posteriorly even in the neutral acquisitions. This will help in raising the suspicion in the absence of a clinical suspicion and flexion studies should be performed once the suspicion is raised.

What we established with the present study was that a properly done flexion study forms the basis of the diagnosis and has a 100% positive predictive value in establishing all the contributory findings (significant falling forward of the dura with increased lamino-dural space, prominence of epidural T2 flow voids and Dural indentation of posterior cord, with loss of anterior and posterior CSF space) [15]. Six of the patients had previously undergone an MRI elsewhere which had been reported as cervical spondylosis and disc bulges. This was due to the lack of flexion acquisition or inadequate flexion. The degree of flexion to demonstrate falling forward of the dura and consequent posterior epidural flow voids cannot be obtained if we leave the head coil in place. This could be the common error, since most MRI technicians would hesitate to do the study without an anterior coil, due to significant loss of SNR. This compromise however is necessary to achieve good flexion.

We defined adequate flexion as the closest approximation of the patient's chin to the chest with adequate support to the back of the patient's neck. This was the technique which helped us achieve a 100% positive predictive value in flexion MRI findings. To our knowledge, this distinct protocol for flexion MRI has not been previously described in literature.

The administration of intravenous Gadolinium in suspected Hirayama's is commonly done to demonstrate the engorged epidural venous plexus enhancement in the flexed position. There is no other contributory role for Gadolinium described in any literature. The argument against this step could be that since the engorged venous plexus is well demonstrated as prominent flow voids in a high resolution adequately performed T2 sagittal flexion acquisition of the cervical spine, there is no necessity for the Gadolinium administration. Once more, the caveat here is adequate flexion has to be achieved. Otherwise false negatives would be a common occurrence.

LIMITATIONS

A comparison with Gadolinium enhanced scans was not done in this study. Such studies in the future may further help in affirming that contrast MRI does not have a role in MR diagnosis of Hirayama. Other authors are encouraged to use this degree of flexion in their patients to validate these results further.

CONCLUSION

With the present study we effectively demonstrated the MRI findings in neutral and flexion acquisitions in making the diagnosis of HD.

It was re-affirmed that the predominant occurrence of the disease is in young males and it also showed a possible relationship with increased patient height.

The unique hallmark of this study was setting a defining standard for the degree of flexion required to demonstrate the falling forward of the dura. By achieving this described degree of flexion, the use of intravenous Gadolinium administration is deemed unnecessary and redundant, resulting in a significantly reduced scan time, increase in patient comfort and safety without compromise in the accuracy of diagnosis.

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