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CLINICAL PROFILE AND ELECTRODIAGNOSTIC,IMAGING FEATURES OF HIRAYAMA DISEASE IN TERTIARY CARE HOSPITAL

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ABSTRACT **Background & Objectives:** Hirayama disease(HD) is a benign condition usually present as weakness and wasting confined to one or both distal parts of upper limb in young adolescence age group .The diagnosis of Hirayama disease is well established by certain salient features in MRI cervical spine neutral and dynamic flexion studies. **Objectives:** To study CLINICAL Charcterstics, Electrodiagnostic, Imaging Findings Of Hirayama Disease **Methods:** It was an observational study conducted in department of Neurology Thanjavur Medical college over a period of 1 year from August 2022 to August 2023.All patients evaluated by detailed history and examination , Nerve conduction study of both upper limbs for analysis of Median and Ulnar both motor and sensory nerve conduction parameters and Needle EMG of Both upper limbs were done in C5 to T1 Myotomes. and MRI cervical spine neutral and dynamic flexion studies. **Results:** All 15 patients (100%) were males in the age group ranging from 18 to 30.Among 15 patients , 9(60%) patients had Unilateral Involvement and 6(40%) patients had Bilateral involvement.In MRI cervical spine and dynamic flexion studies, Atrophy or thinning of the lower cervical spinal cord on sagittal view noted in 12(80%) patients with 3 (20%)patients did not have cord atrophy ,T2 hyperintensity within the spinal cord at anterior horn region was noted in 6(40%) out of 15 patients ,12(80%) patients had abnormal curving of the cervical vertebral column, Detachment of the posterior dural sac from the overlying lamina with anterior displacement was seen in 12(80%) patients out of 15 patients , Intramedullary signal hyperintensity was seen in 6 (40%) patients and 10 (67%) among 15 patients had posterior Epidural flow voids. **Conclusion:** HD is a benign focal atrophy involving distal upper limb either unilateral or bilateral ,predominantly seen in young males.Treatment options for Hd ,avoid frequent neck flexion and cervical collar and surgery is reserved for those patient who still progressive despite conservative treatment.

KEYWORDS : Hirayama disease, MRI neutral /dynamic flexion, cervical collar.

INTRODUCTION:

Hirayama disease(HD) is also known as juvenile muscular atrophy of hand and forearm was first described by Keizo Hirayama in 1959[1] .It is also called Benign focal atrophy[2] because most of cases confined to one or both upper limb with initial 2-3 years progression, unlike other form of motor neuron disease most of HD patients won't progress beyond 5 years. Hirayama disease is most common in Males and between 15 -25 years of age[3]. Toma et al[4],found that different growth rates between males and females during puberty is related to male preponderance .There is disproportional growth between vertebral column and spinal cord contents is the cause of tight dural sac and anterior displacement of posterior dural wall .In HD when neck is flexed ,tight dural sac cannont compensate for the increased length of posterior wall leads to anterior shifting of posterior dural wall and consequent spinal cord compression .On repeated neck movements, spinal cord compression cause microcirculatory disturbances[5] in anterior spinal artery which resulted in necrosis of anterior horn cells which lead to development of weakness and wasting of distal hand muscles in HD. Hirayama disease is thought to be unilateral disease initially, but can involve contralateral side in 20% patients .Hirayama disease predominantly affecting lower cervical cord segment (C7 –T1) anterior horn cells,with sparing of brachioradialis which is also known as oblique amyotrophy.In hand muscle involvement more severe involvement in ulnar nerve distribution such as Abductor digiti minimi than median nerve innervated muscles such as abductor pollicis brevis ,this phenomenon known as reverse split hand sign In Nerve conduction study of ulnar an median nerve showed similar to clinical involvement as reduced CMAP amplitude or absent CMAP in Ulnar nerve than median nerve. Transient acute worsening of weakness on exposure to cold known as cold paresis is one of feature seen in HD is due to sodium channel dysfunction[6][7] .MRI cervical spine at neutral position showing loss of normal cervical lordosis ,loss of posterior attachment of dura at subjacent lamina,localised lower cervical cord atrophy,asymmetric flattening of cord anteroposteriorly, intramedullary T2 hyperintensity on axial section it resembles as snake eye .On flexion MRI radiological features include forward displacement of posterior dura,epidural flow voids with contrast enhancement and high intensity cresent shaped mass due to congesition of venous plexus. There is lack of awareness of HD ,this study was aimed to review clinical ,electro physiological,MRI cervical spine Neutral and flexion features of HD for early diagnosis and to prevent disability.

AIM

To study Clinical Charcterstics, electrodiagnostic, Imaging Findings Of Hirayama Disease

OBJECTIVES

1) To study the clinical profile of Hirayama disease.
2) To study the changes in Nerve conduction study parameters, of median and ulnar and Needle EMG of both upper limbs in C5-T1 myotome muscles.
3) To study the MRI changes in patients with Hirayama disease in neck neutral position and neck flexion position.

MATERIAL AND METHODS

It was an observational study conducted in department of Neurology Thanjavur Medical college over a period of 1 year from August 2022 to August 2023.Institution ethics committee approval and consent from the patients were obtained .

Selection Of Subjects

Inclusion Criteria:

- Age 15-40 years
- Weakness and wasting confined to one or both upper limbs

Exclusion Criteria:

- Lower limb weakness and wasting
- Sensory disturbances
- Sphincter disturbances
- Cranial nerve involvement

Stastitical Methods:

All the data were entered in to MS Excel and presented as bar charts.

Study Design

For the patients admitted with weakness and wasting confined to one or both upper limb suspected to have HD was evaluated by detailed history and clinical examination ,electro diagnostic studies NCS was done in both upper limbs for analysis of Median and Ulnar both motor

and sensory nerve conduction parameters and Needle EMG of Both upper limbs were done in C5 to T1 Myotomes for analysis of denervation. All patients underwent MRI cervical spine and dynamic flexion studies in neck straight - Sagittal SE T1W, TSE T2, Gradient Echo T2. Neck in flexed position - Sagittal SE T1W, TSE T2, Gradient Echo T2 sequences was done and following features were evaluated , Atrophy or thinning or the spinal cord on sagittal view ,Flattening of the cervical cord in antero posterior diameter,T2 hyperintensity within the spinal cord at anterior horn region, Abnormal curving of the cervical vertebral column, Detachment of the posterior dural sac from the overlying lamina,Crescent shaped space behind the dural in cervical region,Intramedullary signal hyperintensity.

RESULTS

In this study ,total 15 patients were studied. All 15 patients (100%) were males in the age group ranging from 18 to 30.Of 15 patients ,6 (40 %) patients in 18-20 years,7 (47%) patients were in 21 -25 years ,2 (13%)patients were in 26-30 age group .Among 15 patients we studied 9(60%) patients had Unilateral Involvement and 6(40%) patients had Bilateral involvement. Among 9 patients with Unilateral involvement Left upper limb was involved in 6 (67%) patients and Right upper limb was involved in 3 (33%)patients. Among 6 patients with bilateral involvement in 4 (67%)patients Left upper limb was involved first followed by Right upper limb , other 2 (33%) patients right upper limb was involved first followed by left upper limb. Regarding duration of disease , ranging from 3 months to 5 years .3(20%) patients with less than 1 year duration of weakness ,10 (67%) patients with 1-2 years of duration and 2(13%) patients with more than 2 years of duration .History of worsening of weakness on cold exposure was seen in 3 (20%) out of 15 patients. History of frequent neck manipulation was seen in 5(33%) patients. On clinical examination, All 15 (100%)patients had hand muscle wasting, in that reverse split hand sign(wasting of ADM>APB and 1st DI) was seen in 5 (33%) patients ,other 10(67%) patients had uniform wasting of all hand muscles.In addition to hand muscle wasting and weakness ,8(53%) patients had proximal involvement .On Myotome pattern ,All 15(100%) patients had C8 and T1 involvement,6(40%) patients had additional involvement of C7 myotome muscles,5(33%) patients with C6 myotome and 4(27%) patients with C5 myotome involvement. Oblique atrophy known as Sparing of brachioradialis were seen in 12 (80%)out of 15 patients. Fasiculatory tremulousness of distal upper limbs was seen in all 15(100%) patients, in addition Fasiculation noted in proximal muscles in 7(87%) patients out of 8 patients with proximal involvement. Brisk DTRs was noted in 5(33%) patients. In Nerve conduction parameters of Ulnar and median nerve shows ,among 15 patients -10 (67%)patients had reduced Ulnar CMAP and in 5 (33%)patients ulnar CMAP was not obtained ,5(33%) patients had reduced median CMAP ,with normal both median and ulnar SNAP in all 15 patients .

Needle EMG of C5 -T1 Myotome showed chronic denervation pattern in C8 and T1 myotome in all 15(100%) patients, 6 (40%)patients with C7 myotome muscles,5(33%) patients with C6 myotome and 4(27%) patients with C5 myotome muscle showed chronic denervation. In MRI cervical spine and dynamic flexion studies , Atrophy or thinning of the lower cervical spinal cord on sagittal view noted in 12(80%) patients with 3 (20%)patients did not have cord atrophy,T2 hyperintensity within the spinal cord at anterior horn region was noted in 6(40%) out of 15 patients ,12(80%) patients had abnormal curving of the cervical vertebral column, Detachment of the posterior dural sac from the overlying lamina with anterior displacement was seen in 12(80%) patients out of 15 patients, Intramedullary signal hyperintensity was seen in 6 (40%) patients and 10 (67%) among 15 patients had posterior Epidural flow voids.

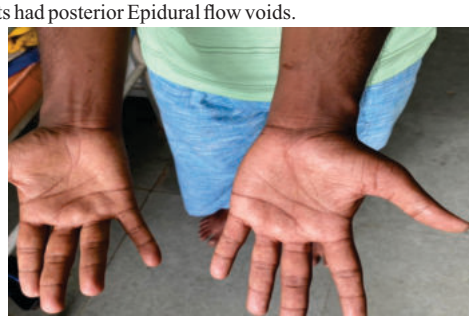

Figure 1.Reverse split hand sign Hypothenar muscles wasting >Thenar muscles wasting

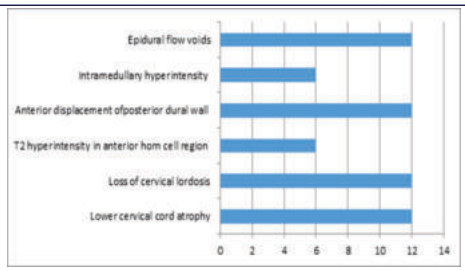

Table 1.MRI Cervical spine Findings

DISCUSSION

Hirayama disease is most common in Males and most common in the age group of 2nd to 3 rd decade .In our study all patients (100%) were males, and age group also in 2 nd and 3 rd decade with ranging from 18 -30 years with mean age of 20.4 years. Our results were similar with previous studies . In study done by WANG Xin-ning, et al[8],studied 73 patients of Hirayama disease, among 73 patients 67 patients were males in the age group of 14-38 years with mean age 21.2 years .

Male predominance and 2nd to 3 rd decade onset is due to differential pubertal spurt between males and females ,occurs in early to mid teenage leading on to disproportionate growth of the vertebral column and the contents of the spinal canal during puberty and disproportionate dural shortening accentuated during growth spurts leads to the development of microcirculatory disturbance in lower cervical spinal cord during neck flexion.

Hirayama disease was initially thought to be as unilateral and distal upper limb predominantly C8 and T1 myotome ,but later studies have demonstrated bilateral involvement and proximal arm involvement .In our study 9 (60%)patients had unilateral and 6(40%) patients had bilateral involvement .In addition to distal hand wasting and weakness proximal upper limb involvement seen in 8(53%) patients .

There is characteristic sparing of brachioradialis muscle in hirayama disease ,otherwise called ad oblique amyotrophy .In our study 12(80%) out of 15 patients had sparing of brachioradialis muscle .

Hirayama disease usually confined to upper limb with predominant LMN signs ,but some patients may develop lower limb involvement and UMN signs. In our study 5(33%) patients had Brisk DTRs in lower limb .

Strenuous physical activity is a risk factor for HD. In our study 5(33%) patients used to strenuous exercise in gym prior to disease onset. Hashimoto *et. al.*, [9] have reported that strenuous exercise of the arms was a frequent association with their HD patients.

Cold paresis is one of the feature in HD especially during early stage of illness, On exposure to cold transient acute worsening of weakness in background of slowly progressive weakness. This phenomenon is due to increased persistent sodium channel function and reduced transient sodium channel and potassium channel function in motor axon neurons leads to axonal dysfunction. In our study 3 (20%) out of 15 patients noticed this phenomenon.

In Nerve conduction study , amplitude of compound muscle action potentials is reduced in the atrophied muscles .In our study 10 (67%) patients had reduced Ulnar CMAP and in 5(33%) patients ulnar CMAP was not obtained and 5(33%) patients had reduced median CMAP. Predominant Ulnar involvement in NCS is due to ulnar nerve innervated muscles are severely affected such as abductor digiti minimi and interossei than median nerve innervated muscles abductor pollicis brevis which is called as reverse split hand sign .

Needle EMG of C5 -T1 Myotome showed chronic denervation pattern in C8 and T1 myotome in all 15(100%) patients, in addition 6(40%) patients with C7 myotome muscles,5(33%) patients with C6 myotome and 4(27%) patients with C5 myotome muscle showed chronic denervation.

In MRI cervical spine neutral and dynamic flexion studies , Atrophy or thinning of the lower cervical spinal cord on sagittal view noted in 12 (80%)patients with 3 (20%)patients did not have cord atrophy , T2 hyperintensity within the spinal cord at anterior horn region was noted

in 6 (40%)out of 15 patients , 12(80%) patients had abnormal curving of the cervical vertebral column, Detachment of the posterior dural sac from the overlying lamina with anterior displacement was seen in 12(80%) patients out of 15 patients, Intramedullary signal hyperintensity was seen in 6(40%) patients and 10 (67%) among 15 patients had posterior epidural flow voids.

The above mentioned radiological features were similar with other studies done by Pradhan S *et.al*.[10], Hassan KM *et.al*.[11], Kikuchi S *et.al*.[12], Mukai E *et.al*.[13], Biondi A *et.al*.[14], Ammendola A *et.al*.[15].

CONCLUSION

HD is a benign focal atrophy involving distal upper limb either unilateral or bilateral ,predominantly seen in young males.This study despite small number of patients described typical salient features of HD similar to previous studies. MRI cervical spine dynamic study to confirms the diagnosis of HD. Treatment of HD includes,reduce neck flexion ,cervical collar[16][17], surgical treatment[18] reserved for who still progress with conservative management.

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