Clinical,electrophysiological and MRI profile of Hirayama disease: A case series

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ABSTRACT

Introduction:

Hirayama disease (HD) is focal amyotrophy in young adults, and commonly involves distal upper limbs, often misdiagnosed as motor neuron disease and writer's cramp. This often leads to delay in the diagnosis and results in disease progression. In this study, we have analyzed clinical, radiological and electrophysiological profile of patients presenting with hand wasting and weakness.

Materials and Methods:

Patients presenting with insidious onset of hand wasting (January 2014 to February 2017) were evaluated clinically and electro physiologically. Cervical MRI in neutral and flexion position was done.

Results:

All 16 patients were male, were less than 25 years of age, with median age of 22 years. Duration of illness was 3 months to 7 years. All patients presented with insidious onset of progressive weakness and lower motor neuron type of wasting of hands. Nine (56.25%) patients presented with right sided, five (31.25%) with left sided and another two (12.5%) presented with bilateral asymmetric weakness and wasting. None of the patients had neck pain or radicular symptoms. Twelve (75%) had cold paresis, eight (50%) had minipolymyoclonus and five (31.25%) had fasciculations. Electromyography (EMG) showed chronic denervation in the C7T1 myotomes. In MRI, localized lower cervical cord atrophy was seen in 13 (81.25%) cases. Asymmetric cord flattening was noted in 14 (87.5%) cases. Loss of dural attachment in 13 (81.25%), anterior displacement of dorsal dura on flexion in 14 (87.5%) and epidural flow voids were seen in 14 (87.5%) cases. Enhancing epidural crescent in flexion was seen in all 12/16(75.0%) cases. Intramedullary hyper intensity was seen in 2 (12.5%) patients.

Conclusions:

Clinical features of HD corroborated well with electrophysiological diagnosis of anterior horn cell disease of lower cervical cord. While dynamic contrast MRI is characteristic, routine studies have a high predictive value for diagnosis. Prompt diagnosis is important to differentiate it from other progressive conditions and to institute early therapy.

Key words: **Electrophysiology; flexion magnetic resonance imaging; hand wasting; Hirayama disease.**

Introduction

Hirayama disease (HD) was first reported from Japan in 1959 as "juvenile muscular atrophy of unilateral upper extremity". Since then, similar patients in their teens or 20s have been described under a variety of names, not only in Japan but also in other Asian countries as well as in Europe and North America[1].

HD is characterized by insidious onset of unilateral or asymmetric atrophy of the hand and forearm with sparing of brachioradialis, giving the characteristic appearance of oblique amyotrophy involving mainly C8 and T1 myotomes and occasionally C7 myotome. It is thought to be a kind of cervical myelopathy related to flexion movements of the neck[2].

HD is frequently misdiagnosed as motor neuron disease (MND), or spinal muscular atrophy but these differ considerably. This disease entity should be more widely recognized as prognosis in this condition is better and early detection and effective and timely intervention can be instituted[1]. The present study reviews the clinical, electrophysiological and MRI features of HD.

Materials and methods

The study was conducted at a super specialized tertiary care hospital. Patient selection criteria were:

(a) weakness and wasting predominantly in C7, C8 and T1 myotomes in one upper limb or asymmetrically in both upper limbs,

(b) insidious onset in the teens or in the early 20s,

(c) progression for 1–3 years followed by arrest of disease or relatively benign course, (d) irregular coarse tremors (minipolymyoclonus) in the fingers of the affected hand(s), (e) mild transient worsening of symptoms on exposure to cold,

(f) absence of substantial sensory abnormalities, cranial nerve, pyramidal tract signs in lower limbs, sphincter or cerebellar deficits[3].

Electrophysiological study included nerve conduction study (NCS) and electromyogram (EMG). Motor and sensory nerves conduction studies were carried out in the median and ulnar nerves of both upper limbs. Needle EMG studies were done in muscles of the C5 toT1 myotomes.

MRI was performed on a 1.5 Tesla MRI (Phillips). The MRI protocol included imaging in neutral position followed by imaging in hyperflexion. The sequences performed in neutral position included Sagittal SE T1W, TSE T2, and Gradient Echo T2. The sequences performed in hyperflexion of cervical spine included Sagittal SE T1W with and without fat saturation, TSE T2, and Gradient Echo T2. Maximal possible hyperflexion of neck was achieved by asking the subject to first move the head as forward as possible and then to touch the chin to the chest. The shoulders were pushed as far caudal as possible. The position was maintained by supporting the neck and shoulders with MR compatible foam pads.

In MRI, the following features were evaluated:

(a) localized lower cervical cord atrophy,

(b) asymmetric cord flattening,

(c) loss of attachment between the posterior dural sac and subjacent lamina,

(d) anterior shifting of the posterior wall of the cervical dural canal,

(e) enhancing epidural component with flow voids and

(f) intramedullary signal hyper intensity.

All observations were done in the flexion series of dynamic T2 weighted MRI. Lower cervical cord was defined as the cord between C4 and C7. Localized cord atrophy was defined as a decrease in cord size in comparison with the normal cord above and that below the affected level on sagittal MR images and confirmed on transverse MR images[4]. Asymmetric cord flattening was evaluated on transverse MR images. Cord flattening was defined as flattening without a narrowed or obliterated adjacent subarachnoid space. An elliptic spinal cord was considered normal, a pear shaped spinal cord was considered asymmetric cord flattening and a triangular spinal cord was considered symmetric cord flattening. For evaluating the loss of attachment between the posterior dural sac and subjacent lamina, the lamina was defined as the part of vertebra between junctions of laminae medially and laterally by a tangential line along the medial aspect of the pedicle. This was divided equally into three parts. More than one third loss of attachment between the posterior dural sac and the subjacent lamina was considered significant. Anterior displacement of dural sac and appearance of epidural flow voids with enhancing epidural component posterior to the thecal sac was noted. Non compressed intramedullary high signal

intensity was considered if patent subarachnoid space was seen along with intramedullary high signal intensity were observed; whereas such high signal with compression was assumed to be a result of local pressure effects[4].

Results

Sixteen patients fulfilled the clinical criteria and were included in the study

Clinical features

All the patients were male, with mean age 22 years (range 18–31 years) at the time of clinical examination. Duration of illness at the time of presentation was three months to seven years. Four patients had symptoms for less than 1 year, seven had symptoms between 1 and 2 years and five had symptoms for more than 2 years. All patients presented with insidious onset of progressive weakness and lower motor neuron type of wasting of hands. Nine (56.25%) patients presented with right sided, five (31.25%) with left sided and another two (12.5%) presented with bilateral asymmetric weakness and wasting. None of the patients had neck pain or radicular symptoms. Twelve (75%) had cold paresis, eight (50%) had minipolymyoclonus and five (31.25%) had fasciculations. Regional reflexes were variably absent. Plantars were flexors and sensory examination was normal in all patients.

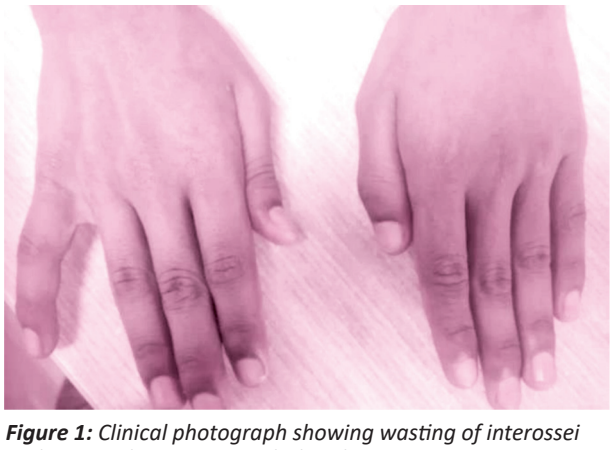

Figure 1: *Clinical photograph showing wasting of interossei with Wartenberg sign in right hand.*

Figure 2: *Clinical photograph showing wasting of ulnar side of forearm in the right side.*

Table 1: Clinical profile

S. No.	Age/ Sex	Weakness in grip	Wasting	progression	Duration (Months)	Minipolymyoclonus	Cold Sensitivity	Wartenbers sign	Side involvement
1	19/M	Y	Intrinsic + FA	Yes	12	yes	Yes	No	R
2	17/M	Y	Intrinsic	Yes	10	No	Yes	yes	L
3	31/M	Y	Intrinsic	Yes	3	No	No	No	R
4	21/M	Y	Intrinsic + FA	Yes	6	yes	Yes	No	R
5	18/M	Y	Intrinsic + FA	No	30	No	No	No	L+ R
6	22/M	Y	Intrinsic + FA	yes	7	No	Yes	yes	R
7	18/M	Y	Intrinsic + FA	Yes	12	yes	Yes	No	L
8	28/M	Y	Intrinsic + FA	No	72	No	No	yes	L+ R
9	24/M	Y	Intrinsic + FA	No	84	No	Yes	No	R
10	18/M	Y	Intrinsic + FA	No	24	yes	Yes	No	L
11	18/M	Y	Intrinsic + FA	No	18	No	Yes	No	L
12	28/M	Y	Intrinsic	Yes	24	yes	Yes	No	R
13	37/M	Y	Intrinsic + FA	No	100	Yes	No	no	L
14	18/ M	y	Intrinsic	Yes	12	yes	Yes	yes	R
15	18/M	Y	Intrinsic	Yes	24	No	Yes	No	R
16	17/m	Y	Intrinsic + FA	Yes	6	yes	yes	yes	R

Electrophysiology

NCS and EMG were done in all the patients within 1 week of the clinical examination. Median and Ulnar compound muscle action potentials (CMAPs) were reduced in all patients. However, distal latencies and F-wave latencies were within the normal range. Conduction velocities were normal in all nerves. Sensory NCS was normal in all the patients. EMG revealed features of active denervation in the form

of fibrillations in five patients and fasciculation in six patients, and features of chronic denervation in the form of neurogenic changes in C7, C8 and T1 myotomes in 13 patients and neurogenic changes in the C8, T1 in three patients. EMG of C5, C6 myotomes, namely Deltoid, Biceps brachii and Brachioradialis was normal.

Table 2: Electrophysiological profile

S. No.	Age/ Sex	NCV	EMG	
1	19/M	Normal	C7C8/T1 Myotome polyphasic MUPS	Fibrillation
2	17/M	Normal	C8/T1Myotome polyphasic MUPS	Fibrillation
3	31/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
4	21/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
5	18/M	Normal	C7/C8 Myotome polyphasic MUPS	Fibrillation
6	22/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
7	18/M	Normal	C7C8/T1 Myotome polyphasic MUPS	Fasciculation
8	28/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
9	24/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fibrillation
10	18/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
11	18/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fibrillation
12	28/M	Normal	C8/T1 Myotome polyphasic MUPS	Fasciculation
13	37/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fibrillation
14	18/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fasciculation
15	18/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fibrillation
16	17/M	Normal	C7/C8/T1 Myotome polyphasic MUPS	Fibrillation

MRI findings

Localized lower cervical cord atrophy was seen in 13/16 (81.25%) of the suspected cases of HD. Asymmetric cord flattening was noted in all 14/16 (87.5%) cases. Loss of dural attachment in 13 (81.25%), anterior displacement of dorsal dura on flexion in 14(87.5%) and epidural flow voids were seen in 14 (87.5%) cases. Enhancing epidural crescent in flexion was seen in all 12 (75.0%) cases. Intramedullary hyper intensity was seen in 2 (12.5%) cases. Salient MRI features are shown in Table 3.

Table 3: MRI profile

S. No.	localized lower cervical cord atrophy	Asymmetric cord flattening	loss of attachment between the posterior dural sac & subjacent lamina	Anterior shifting of the posterior wall of the cervical dural canal	Enhancing epidural component with flow voids	Intramedullary signal hyperintensity
1	yes	Yes	Yes	Yes	No	No
2	No	No	Yes	Yes	Yes	No
3	Yes	Yes	No	No	No	Yes
4	Yes	Yes	Yes	Yes	Yes	No
5	Yes	Yes	Yes	Yes	Yes	No
6	Yes	Yes	Yes	Yes	Yes	No
7	No	Yes	Yes	Yes	Yes	No
8	Yes	Yes	No	yes	yes	Yes
9	Yes	Yes	Yes	Yes	Yes	No
10	Yes	Yes	Yes	Yes	No	No
11	Yes	No	Yes	Yes	Yes	No
12	Yes	Yes	Yes	Yes	Yes	No
13	Yes	Yes	No	No	No	Yes
14	Yes	Yes	Yes	Yes	Yes	No
15	Yes	Yes	Yes	Yes	Yes	No
16	No	Yes	Yes	Yes	Yes	No

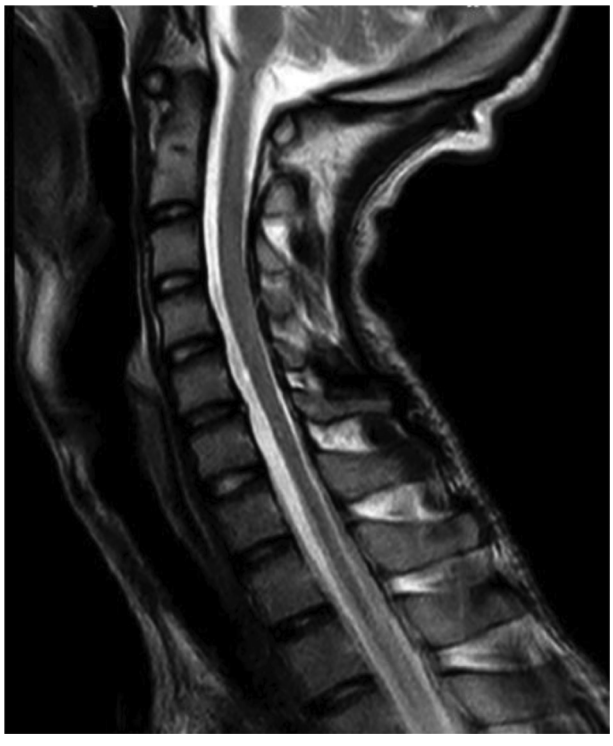

Figure 3: *T2W sagittal MRI in the extension*

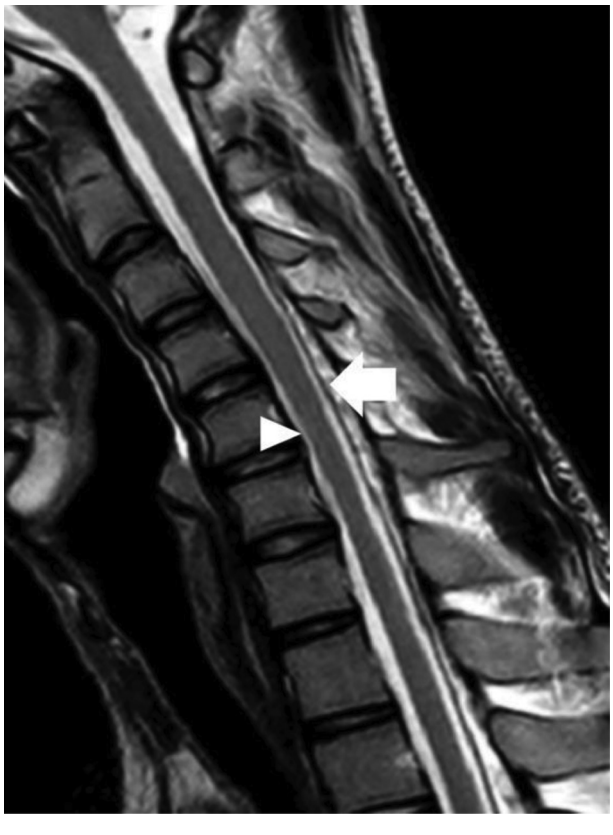

Figure 4: *T2W Sagittal MRI in flexion showing cord atrophy (white arrowhead) and anterior shift of posterior dura (white arrow)*

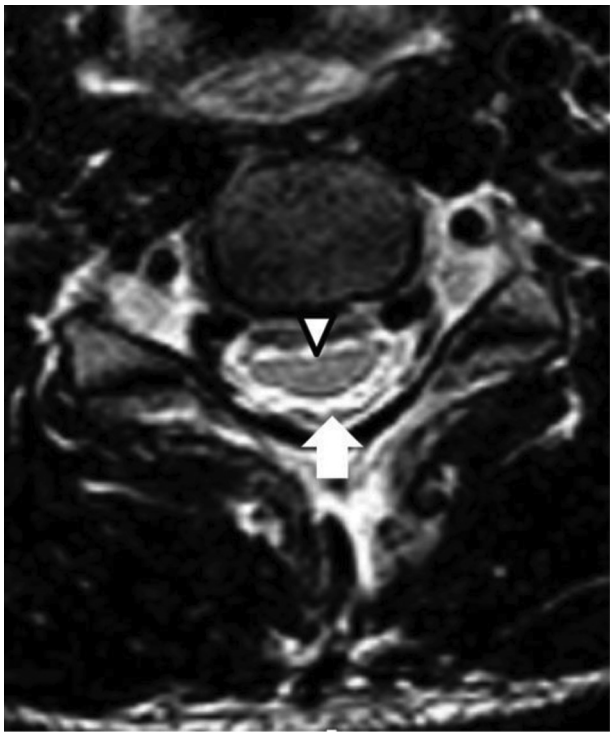

Figure 5: *T2W axial image in flexion at the level of C5 showing asymmetrical cord flattening (white arrowhead with black border) and anterior shift of dura with epidural flow void (white arrow)*

Discussion

Hirayama disease was initially recognized in Japan in 1959 and reported under the name of juvenile muscular atrophy of unilateral upper extremity[5]. Since then similar cases have been described from many countries, mostly from Asia. In a report in 1991, Chan et al. estimated 150 cases from Japan, 37 from India, and 102 from Sri Lanka[6]. There are reports from several other countries including Singapore, Taiwan, Hong Kong, Nigeria, Denmark, Holland, USA, Austria, UK, France, Canada, Poland and Germany[5,6]. From Nepal, there is a case report[7] reported in the year 2013 but this is the first reported series from Nepal.

This disorder mainly occurs in males usually between 15 to 25 years. Clinical features include insidious onset, asymmetric involvement of upper extremities and self-limiting course after few years of progression[6]. The atrophy and weakness typically involves the intrinsic muscles of hand (interossei, thenar, and hypothenar muscle groups) as well as the ulnar side of the forearm[5]. There is sparing of brachioradialis muscle giving the impression of an 'oblique atrophy'[8,9,10]. There is unilateral involvement in majority, but asymmetric and symmetric

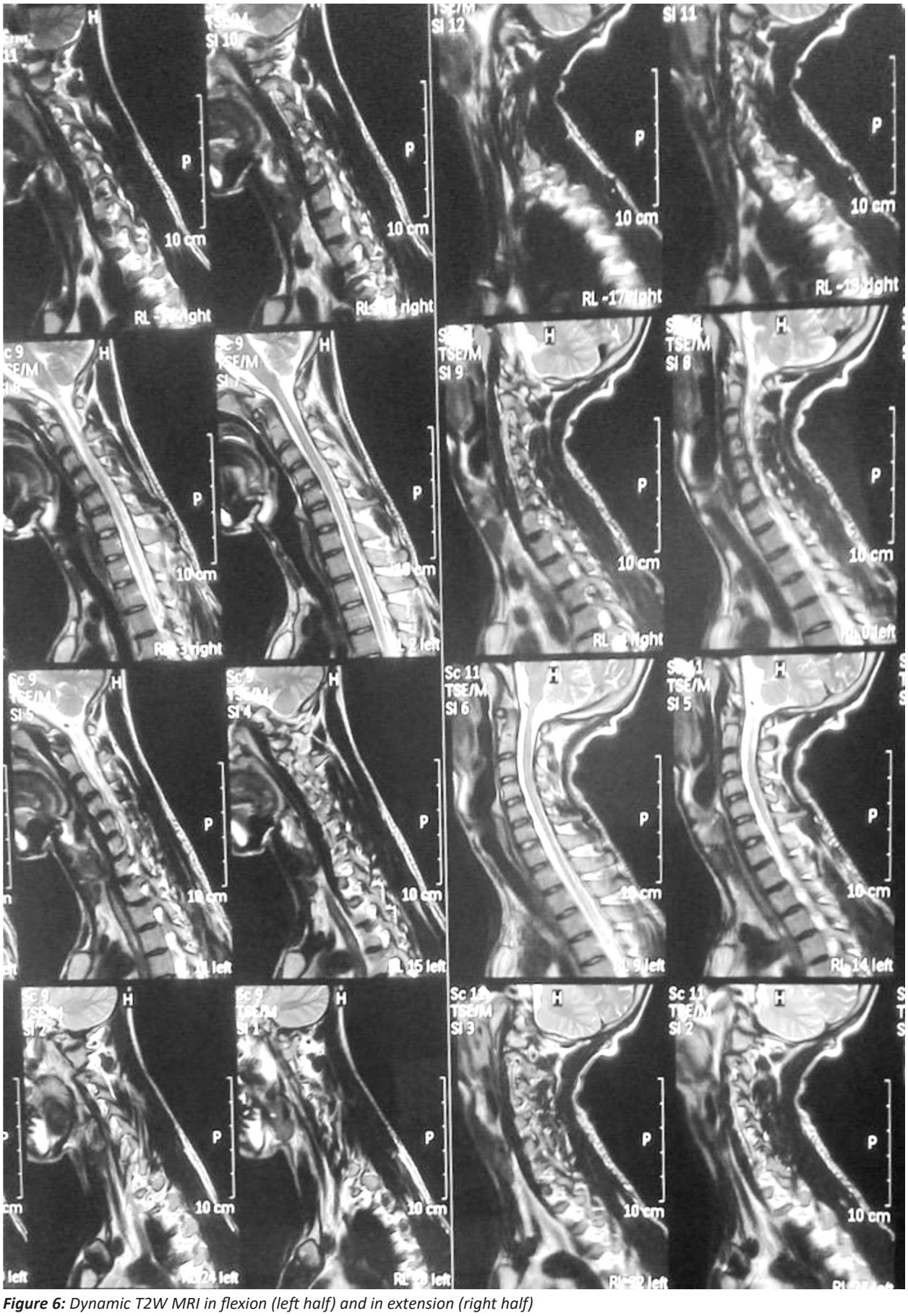

Figure 6: *Dynamic T2W MRI in flexion (left half) and in extension (right half)*

bilateral involvement is also observed[9]. In 18 cases of Kikuchi et al.[5] 71% patients had unilateral and 29% had bilateral involvement. There is no relationship between the patient's handedness and side of greater muscular atrophy[11,12]. Deep tendon reflexes in upper extremities are normal, however, hypoactive triceps reflex and mild hyperreflexia in legs have been reported[9]. Sensory symptoms and signs are conspicuously absent[11,13]. A distal irregular jerky tremor (minipolymyoclonus) is also seen in some patients[8,10,11]. Another interesting phenomenon is exacerbation of weakness in cold environment[5,10].

The nerve conduction studies frequently demonstrate low-amplitude compound muscle action potentials (CMAPs) commensurate with degree of weakness and atrophy[2,5]. The electromyographic (EMG) findings usually show loss of motor unit potentials (MUPs) which are rapidly firing, high amplitude, and polyphasic, as seen in anterior horn cell disorders[8,5,11]. These changes are localized to C7, C8 and T1 myotomes. Fibrillation potentials and sharp waves may be seen[10]. Our patients had low amplitude CMAPs with giant MUPs and active denervation.

Conventional radiographic studies of the cervical spine show no specific abnormalities. Myelography can show forward movement of posterior dural wall when neck is flexed but this examination is difficult to perform as contrast medium is not easily retained in the flexed cervical subarachnoid space[2]. MRI with flexion contrast study is the gold standard for diagnosis. The findings reported more frequently are asymmetrical or symmetrical atrophy of lower cervical cord, prominence and enhancement of posterior epidural venous plexus on flexion studies and anterior shifting of posterior dural sac on flexion. Loss of attachment between the posterior dural sac and subjacent lamina on neutral position, anterior shifting of the posterior wall of cervical dural canal, enhancing epidural crescentic mass in the lower cervical and thoracic region and prominent posterior epidural flow voids suggestive of dilated epidural venous plexus on flexion studies are reported as highly suggestive for the diagnosis of HD[3,4,14,15].

Among the imaging features discussed above, localized lower cervical cord atrophy, asymmetric cord flattening and loss of attachment have an accuracy of 80% in identification of the disease; loss of attachment has been proposed as the most valuable finding for diagnosing HD in the neutral position[15,16]. In our study, localized lower cervical cord atrophy was seen in all the cases which is comparable to the earlier studies. Pradhan et al. found focal cord atrophy in neutral neck position on MRI in 100% of their cases[14], while Hirayama has reported this finding in only 50% of his cases[3]. Asymmetric cord flattening was noted in all of our cases. Loss of dural attachment, anterior displacement of dorsal dura and epidural flow voids on flexion were seen in 90% of the cases. Enhancing epidural crescent in flexion was seen in all cases. Our findings are in greater conformity with the findings of Raval et al.[17] than that of Sonwalkar et al.[18]. Such variation is likely due to our selection criteria which is inclusive of the HD.

The pathogenesis is debated. Dynamic spinal cord compression due to neck flexion with forward displacement of posterior dura is considered as the primary mechanism[9]. When neck is flexed, the cervical cord is longitudinally stretched. In Hirayama disease, the lower cervical cord moves forward in flexion and contacts the posterior surface of the vertebrae; the lower cervical cord becomes flattened at contact point[5]. Additionally, the posterior wall of the dural tube moves forward, the posterior epidural space expands forming a crescent shaped mass in the posterior epidural space. This mass is formed by the congestion of posterior internal vertebral venous plexus[5,15]. In a report of 73 patients, Hirayama concluded that dynamic cord compression in flexion with forward displacement of posterior dura is an unequivocal finding in progressive stage[3]. The mechanism of myleopathy may involve ischemic changes or chronic trauma by repeated neck flexion affecting anterior horn cells along with spinal cord thinning, termed as flexion myelopathy[9]. The chronic circulatory disturbance resulting from repeated or sustained flexion of neck may produce necrosis of the anterior horns resulting in gliosis and localized cord atrophy at the lower cervical region[3,18,19]. Vulnerability of the anterior horn to ischemia accounts for the atrophy that follows[3]. Patients with severe cervical cord compression in flexion may also develop extensive cord injury beyond the anterior horns. The neutral-position MRI may show lower cord atrophy, asymmetric cord flattening, non-compressed intramedullary high signal intensity on T2-weighted imaging[20].

There is no definitive treatment of this condition. The primary principle of treatment is based on avoiding neck flexion. Low pillows are recommended while sleeping. A neck collar is used on regular basis for at least 3 to 4 years[5,15]. Anterior fusion of cervical vertebrae and duraplasty, with or without anterior fusion has also been performed. The indications and methods of surgical treatment remain controversial[5].

Deriving from our cases, we can state that asymmetry is one of the most characteristic findings of this disease, both clinically and radiologically. Thus, in cases of adolescent onset of distal upper limb weakness, the finding of asymmetric cord atrophy on routine non-flexion MR studies, especially at the lower cervical cord, should raise the suspicion of HD. When this finding is seen, flexion MR study should be performed to confirm the diagnosis[15,18]. Our study supports the hypothesis that HD differs from classical MND or its variants involving the distal upper limbs in young males by being a chronic ischemic myelopathy rather than a degenerative condition. Monomelicamyotrophy (MMA) has been classified under the idiopathic group of MND seen in India[22]. Young patients with unilateral upper limb atrophy have been classically labeled brachial MMA, which shows resemblance with the HD. Gourie Devi et al., in their study, from year 1977 to 1981, reported 13 patients with single upper limb atrophy[23]. The clinical description of these patients with single upper limb atrophy reported by Gourie Devi et al. is similar to the clinical profile of HD described by Hirayama et al.[24] Recently, Pradhan et al. in their series of 106 patients of HD, from 1992 to 2008, reported around 10% of all the patients to have bilaterally symmetric involvement, a severe form of classic HD, which remains undiagnosed due to a common notion that it is a unilateral or grossly asymmetric disease[25]. In our study we had 14 cases with unilateral, and only 2 cases (12.5%) with bilateral involvement.

In young males with distal upper extremity weakness, atrophy, preserved reflexes and normal sensory examination, Hirayama disease should be suspected. To ensure that this diagnosis is not missed in patients presenting with focal hand wasting, flexion contrast MRI should be done if the routine MRI otherwise looks normal and if MND has been excluded. Several conditions like syringomyelia, MND, cervical spondylotic myelopathy, spinal cord tumor and traumatic myelopathy may cause localized amyotrophy of the distal arm, and these should be excluded first by imaging modalities.[21]

Conclusions

Hirayama disease, a rare neurologic entity affecting young men in the second to third decades of life, is characterized by insidious onset and slowly progressive course followed by static phase of unilateral or asymmetric atrophy of the hand(s) and forearm(s) with sparing of the brachioradialis, characterized as oblique amyotrophy. Dynamic MRI has a high predictive value for diagnosis with characteristic features. Prompt diagnosis is important to differentiate it from other progressive condition to avoid repeated consultation and anxiety in these young patients.

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