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Hirayama-Like Disease In The Thoracic Spine : A Case Report

Moon-Jung Kim M.D.[1], Kyung-Lim Joa M.D.[1], Myeong-Ok Kim M.D.[1], Han-Young Jung M.D.[1], Ro-Woon Lee M.D.[2]
Chang-Hwan Kim M.D.[1]
[1]Department of Physical & Rehabilitation Medicine, College of Medicine, Inha University, Incheon, Republic of Korea.
[2]Department of Radiology, College of Medicine, Inha University, Incheon, Republic of Korea.

Introduction

Hirayama disease is a flexion disorder of spine that involves the lower cervical segment. The pathophysiology is unknown but would be due to dynamic compression of spinal cord by forward displacement of the dorsal dura during neck flexion. It usually develops asymmetric motor weakness of forearms in male patients. It should be differentiated from other motor neuron diseases. We reported a young man who had features similar to Hirayama disease in the thoracic spine level at spinal cord MRI and electrophysiologic studies.

Case

An eighteen-year-old young man without any past illness visited our clinic with 4 weeks history of slowly progressive left side upper arm weakness. Physical examination revealed weakness of left elbow and wrist extensor muscles, without sensory change. Deep tendon reflex and pathologic reflex examinations revealed no abnormal findings.

He visited other clinic a week ago and already did electrodiagnostic study. At that time, the motor, sensory nerve conduction studies (NCS) showed normal responses, including F waves. Needle electromyography (EMG) study revealed abnormal spontaneous activities (ASA) at bilateral upper extremity muscles (extensor digitorum communis (EDC), triceps brachii (TB) and left upper extremity muscles (flexor carpi ulnaris (FCU), abductor pollicis brevis (APB). Polyphasic motor unit action potentials (MUAPs) were observed in the left upper extremity muscles (TB, EDC, FCU, APB).

We conducted the follow-up study a month after the previous test. NCS revealed normal motor and sensory nerve responses, including F waves in upper and lower extremities. Needle EMG study showed ASA at right flexor carpi radialis muscle. Neurogenic polyphasic (longer duration and increased amplitude) MUAPs were observed in the left upper extremity muscles (flexor carpi radialis (FCR), FCU, extensor carpi radialis, 1st dorsal interosseous, triceps brachii) and right upper extremity muscles (FCR, 1st dorsal interosseous, EDC). The above findings revealed segmental anterior horn cell disease. (Table 1, 2)

We performed cervical spine flexion MRI sequences. During neck flexion, forward displacement of the dura mater was seen along the T1-2 to T7,8 levels and spinal cord moved forward, but no abnormal intramedullary high signal intensities were found. So, we diagnosed this case as Hirayama-like disease involving thoracic spine level. (Figure 1)

We applied neck brace to prevent the neck flexion stress. After wearing a neck brace for 3 months, the strength in the left elbow and wrist extensors was improved.

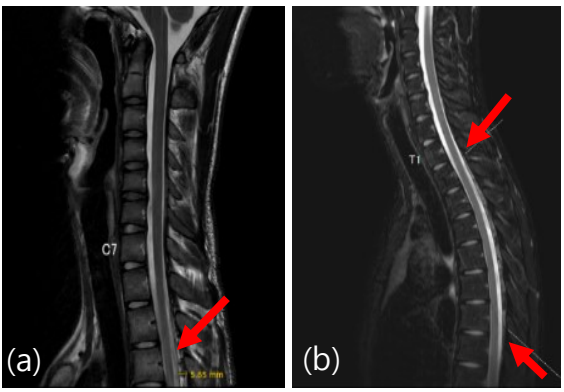

Figure 1. Cervical spinal cord MRI in neck flexed position showed posterior ligamentum flavum was seen along and spinal cord move to forward from T1-2, T7,8 levels. (a) Flexion position on T2-weighted image, (b) Sagittal T2-weighted image shows forward movement of spinal cord from T1-2, T7,8 levels.

Table 1. Nerve conduction studies of both upper and right lower extremities.

Nerve conduction study								
Nerve	Right side				Left side			
	Latency(ms)	Amplitude(mv)	CV(m/s)	F-M latency(ms)	Latency(ms)	Amplitude(mv)	CV(ms)	F-M latency(ms)
Motor								
Median(APB)	2.71/6.04	11.5/10.4	64.5	21.3	2.55/5.99	10.0/9.3	62.5	19.9
Ulnar(ADM)	2.50/5.21	11.3/10.4	59.1	22.7	2.55/5.10	9.5/9.5	66.6	21.0
Radial(EIP)	1.61/4.43	6.1/5.8	56.9		1.77/4.53	5.6/5.5	58.0	
Axillary	2.19	8.9			2.19	8.9		
Peroneal(EDB)	4.38/10.78	6.8/6.6	50.0	43.0				
Sural(AH)	3.75/11.46	13.5/11.9	51.9	43.5				
Sensory								
	Right side			Left side				
	Latency(ms)	Amplitude(uv)	CV(m/s)	Latency(ms)	Amplitude(uv)	CV(ms)		
Median	2.86	49.3	48.9	2.86	55.2	48.9		
Ulnar	3.02	35.1	46.3	2.97	41.7	47.2		
Radial	2.40	45.6	50.1	2.40	38.5	50.1		
Superficial peroneal	3.33	18.9	42.0					
Sural	3.33	18.9	42.0					

*: Abnormal data, APB: Abductor pollicis brevis, ADM: Abductor digit minimi, EIP: Extensor indicis proprius, EDB: Extensor digitorum brevis, AH: Abductor hallucis, CV: Conduction velocity, Amplitude (mV) in motor conduction study, Amplitude (uV) in sensory conduction study.

Table 2. Needle electromyographic findings of both upper and left lower extremities.

Needle Electromyographic Findings									
	Spontaneous					MUAP			Recruitment
Muscle	IA	Fib	PSW	Fasc	CRD	Amp	Dur.	Poly	Pattern
Both Masseter	None	None	None	None	None	N	N	N	Full recruitment
L. Trapezius (upper)	None	None	None	None	None	N	N	N	Full recruitment
Both Deltoid	None	None	None	None	None	N	N	N	Full recruitment
L. Triceps brachii	None	None	None	None	None	N	N	Normal & Poly	Slightly reduced
L. Extensor carpi radialis longus	Increased	None	None	None	None	N	N	Normal & Poly	Full recruitment
L. Flexor carpi radialis	Increased	None	None	None	None	N	N	Normal & Poly	Slightly reduced
L. Flexor carpi ulnaris	None	None	None	None	None	N	N	Poly	Slightly reduced
L. First dorsal interosseous	Increased	None	None	None	None	N	N	Poly	Slightly reduced
R. Triceps brachii	Increased	None	None	None	None	N	N	N	Full recruitment
R. Extensor digitorum communis	Increased	None	None	None	None	N	N	Poly	Reduced
R. Flexor carpi radialis	Increased	2+	2+	None	None	Large	N	Poly	Slightly reduced
R. First dorsal interosseous	None	None	None	None	None	N	N	Normal & Poly	Full recruitment
L. Tibialis anterior	None	None	None	None	None	N	N	N	Full recruitment
L. Peroneus longus	None	None	None	None	None	N	N	N	Full recruitment
L. Gastrocnemius (Medial head)	None	None	None	None	None	N	N	N	Full recruitment
Both C6,7 paraspinals	None	None	None	None	None				
Both T4,5 paraspinal	None	None	None	None	None				
Both Lumbar paraspinals (low)	None	None	None	None	None				

MUAP: Motor unit action potential, IA: insertional activity, Fibs: Fibrillation potential, PSW: Positive sharp waves, Fasc: Fasciculation, CRD: Complex repetitive discharge, Amp: amplitude, Dur: duration, Poly: polyphasic MUAP, N: Normal

Conclusion

We reported a rare case who had bilateral proximal weakness of upper extremities and imaging features similar to Hirayama disease in the thoracic spine level at spinal cord MRI. This case is quite different from classic Hirayama disease. The pathophysiology of this type disorder is unclear, but early recognition is important to differentiate from other motor neuron diseases, to avoid neck flexion stress and to prevent disease progression.