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Hirayama Disease with Proximal Upper Extremity Atrophy mimicking Brachial Plexopathy : A Case Report

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Introduction

Hirayama disease is characterized by progressive muscular weakness and wasting of unilateral forearms and hands in young people. The hypothesis is based on chronic ischemic damage to the lower cervical cord caused by crushing against the vertebral bodies, during neck flexion movements. The patient of this case was army, and he complained insidious onset of weakness and atrophy in the upper arm. Because he occurred these symptoms following hard military work, we initially suspected the possibility of traumatic brachial plexopathy. Through electrodiagnostic study, brachial plexus, cervical spine MRI, we reported this rare case of Hirayama disease with bilateral and proximal arm muscle involvement.

Case Report

A 20-year-old military man visited our outpatient clinic with eight months history of slowly progressive atrophy that started in the right proximal upper extremity following heavy military training (Fig. 1). Physical examination revealed wasting and weakness of triceps brachii, extensor digitorum muscles but no evidence of sensory change. With initial impression of traumatic brachial plexopathy following heavy training, we studied brachial plexus MRI and electrodiagnostic study. To rule out the possibility of Guillain Barre syndrome, the CSF study and serum anti GM1 antibody study have done, the result was normal. Brachial Plexus MRI revealed no definite abnormal finding. Upper limb MRI revealed decreased volume and increased signal intensity at triceps, extensor digitorum, exstensor digiti minimi, extensor carpi ulnaris muscles, suggesting denervation change. Nerve conduction study revealed delayed F wave latency of both median and ulnar nerve. In a needle EMG, denervation potentials and polyphasic motor unit action potentials were observed in the bilateral triceps brachii, flexor carpi radialis, extensor digitorum communis, abductor pollicis brevis, and abductor digit quinti muscles. The above findings revealed segmental anterior horn cell disease (Table 1). We performed cervical spine MRI to assess the relationship of posterior dura mater with the spinal cord through extention and flexion of the neck. During neck flexion, the posterior dura showed anterior displacement. An associated anterior displacement and mild flattening of spinal cord from C5-6 to C6-7 level but no abnormal intramedullary high signal intensity were found (Fig. 2). So, we diagnosed his case as Hirayama disease involving bilateral upper extremities through electrodiagnostic and cervical MRI findings. We recommended upper extremity muscle strenghthening, and avoidance of neck flexion.

Conclusion

We experienced a rare case of the bilateral and proximal weakness of upper extremity in Hirayama disease. Because this case is quite different from other ones, we report it. Although the pathogenesis is not clear, early recognition of this condition is important because avoidance of neck flexion can retard the progression of Hirayama disease.

Fig. 1. Findings of muscular atrophy of the right arm. (a) triceps brachii muscle, (b) extensor digitorum muscle

Table 1. Nerve Conduction Study and Needle Electromyographic Findings of Both Upper Extremities in This Case.*: Abnormal data MC: musculocutaneous nerve, LABCN: lateral antebrachial cutaneous nerve, CV: conduction velocity, Amplitude (mV) in motor conduction study, Amplitude (uV) in sensory conduction study Spont. activity: spontaneous activity, MUPs: motor unit potentials, Fibs: fibrillation potentials, PSW: positive sharp waves, Poly: polyphasia, Amp: amplitude, Dur: duration, R: reduced recruitment, F: full recruitment

Nerve Conduction Study									
Nerve	Right side				Left side				
	Latency (ms)	Amplitude (mV/uV)	CV (m/s)	F-M lat (ms)	Latency (ms)	Amplitude (mV/uV)	CV (m/s)	F-M lat (ms)	
Motor									
Median	3.5/8.5	10.2/1.0	54.0	24.8*	2.9/7.4	8.7/8.6	53.8	24.4*	
Ulnar	2.7/8.4	9.9/9.1	51.1	26.8*					
Radial	3.5/6.3	3.9/2.7	49.3						
Axillary	4.3	12.6			4.4	9.9			
MC	4.9	8.4			4.4	7.1			
Sensory									
Median	3.1	70.1	45.2		3.1	70.8	45.5		
Ulnar	3.0	59.4	47.0						
Radial	2.9	36.2	47.6						
LABCN	2.6	27.5	54.7		2.5	32.2	55.6		

Needle Electromyographic Findings							
Muscle		Spont. activity		MUPs			Recruitment
		Fibs.	PSW	Poly	Amp	Dur	
Rt	Triceps brachii	2+	2+	+	large	long	R
	Abductor digit minimi	2+	2+	+	large	long	R
	Flexor carpi radialis	1+	1+	+	large	long	R
	Extensor digitorum communi	1+	1+	+	large	long	R
	Abductor pollicis brevis	-	-	+	-	-	R
	Deltoid	-	-	-	-	-	F
	Biceps Brachii	-	-	-	-	-	F
	Extensor carpi radialis	-	-	-	-	-	F
	Abductor pollicis brevis	-	-	-	-	-	F
Lt	Triceps brachii	2+	2+	+	large	long	R
	Abductor digit minimi	1+	1+	+	large	long	R
	Extensor digitorum communi	1+	1+	+	large	long	R
	Flexor carpi radialis	1+	1+	+	large	long	R

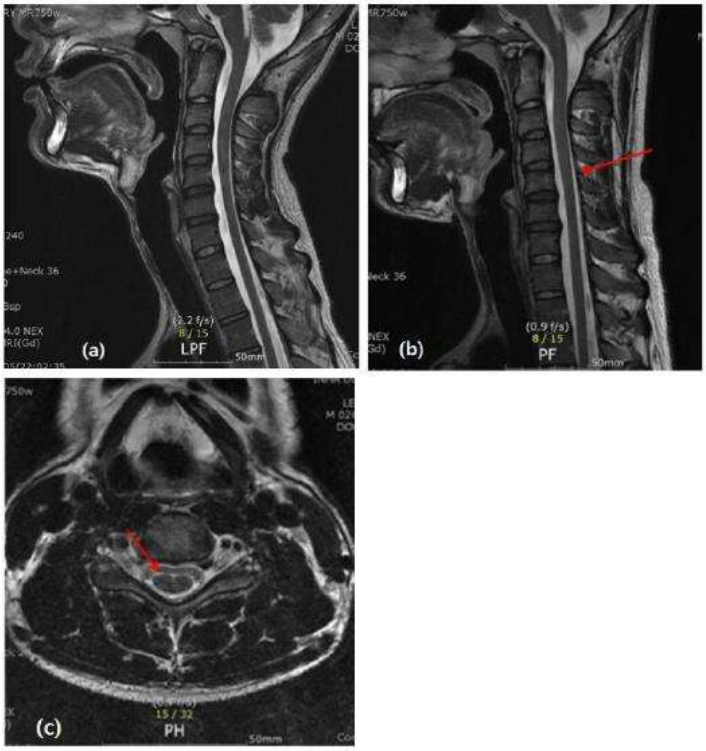

Fig. 2. Cervical spinal cord MRI in neck flexion position showed minimal posterior dural detachment at C4, 5, 6 levels. (a) extension position on T2-weighted image, (b) flexion position on T2-weighted image, (c) Axial T2-weighted image shows mild flattening of spinal cord from C6-7 level on flexion position