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Progressive Muscular Atrophy with Past History of Hypokalemic Periodic Paralysis : Case Report

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Introduction

Progressive muscular atrophy (PMA) is a very rare disease and one of the neuromuscular diseases with lower motor neuron(LMN) symptoms. LMN degeneration occurs, resulting in muscle weakness, atrophy, and fasciculation. Hypokalemic periodic paralysis (HypoPP) is an autosomal dominantly inherited channelopathy, which is temporarily paralyzed with a decrease in potassium level in the blood. The life expectancy is normal, but in some cases, proximal myopathy may occur at a late age. However, PMA in HypoPP patients is not well known. We report a case in which a patient with a history of HypoPP is identified as PMA.

Case report

A 64 - year - old patient visited the outpatient clinic due to weakness of the ankle. The patient had a history of periodic paralysis that improved after potassium injection in his childhood. In MMT, the first left toe and ankle dorsiflexor muscle was grade 1, and the patient complains of tingling pain in the left whole lower extremity. In both knee and ankle, DTR was normotonous and UMN sign was not observed. Atrophy was detected in the left vastus medialis, tibialis anterior, and extensor digitorum brevis muscles. In the electromyography test, Left lower limb CMAP amplitude was abnormal compared to the right. H-reflex was normal on both side. The reinnervation pattern was confirmed in the left lower limb muscles(Table 1). Lt. Lumbosacral plexopathy with sciatic neuropathy was diagnosed and sciatic neuropathy was considered to be improving. Two years later, the patient returned to the outpatient clinic due to weakness on the contralateral lower extremity. In MMT, bilateral ankle and 1st toe dorsiflexor muscle strength were identified as grade 1. Muscle strength of bilateral toe, left plantarflexor, and left knee and hip were grade 2. Other leg strengths were identified as grade 3. The bilateral DTR was decreased in both sides. Both lower limb CMAP amplitudes were abnormal. The denervation pattern and reinnervation pattern were observed in needle EMG performed in both lower extremities muscles(Table 2). The patient was admitted with suspicion of motor neuron disease, and there were no lesions that caused weakness in brain and spine MRI(Figure 1). Genetic testing excluded other genetic diseases. Tongue fasciculation was confirmed and UMN sign did not appear. The patient was diagnosed with PMA.

Discussion

Myopathy after HypoPP is relatively well known, and because of the specific findings in electromyography, rapid diagnosis is possible, but the relationship between PMA and HypoPP is not well known. It is difficult to diagnose PMA patients whose initial symptoms is foot drop because they show similar findings to polyradiculopathy in electromyography study. Therefore, if a patient with a history of HypoPP develops weakness after middle age, serial electromyography study must be performed and careful observation should be made to see if a new abnormality has occurred in the area that was initially normal.

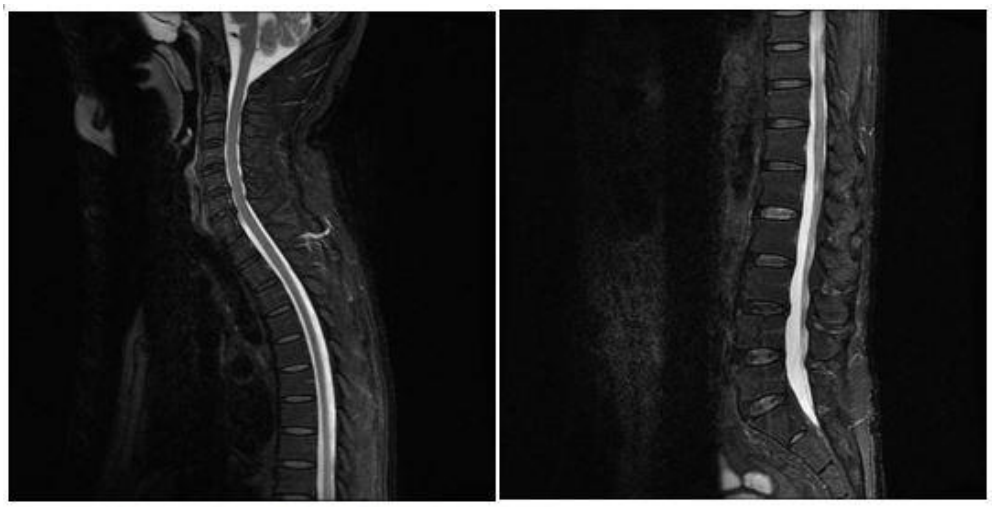

Figure 1. Sagittal T2-weighted images of the spine

Table 1. Electromyography in both lower extremities.

NERVE CONDUCTION STUDY				
Nerve Stimulation (Record)	Amplitude (uV) Distal / Proximal	Conduction Velocity (ms)	Distance (cm)	Latencies Distal / Proximal
Motor				
Rt. Tibial (AH)	7600/4100	42	39	4.3/13.5
Rt. Peroneal (EDB)	1400/1200	44	37	4.7/13.2
Rt. Peroneal (TA)	5000/3700	54	7	4.4/5.7
Rt. Femoral (VM)	4400			3.6
Lt. Tibial (AH)	4600/3100	41	39	4.5/14.1
Lt Peroneal (EDB)	NR			
Lt. Peroneal (TA)	700/400	50	8	3.9/5.5
Lt. Femoral (VM)	3200			3.6
Sensory				
Rt. Sural (Ankle)	10	47	11	2.4/3.2
Lt. Sural (Ankle)	11	50	11	2.2/3.0

Needle EMG

Muscles		Insertion activity	Spontaneous activity			Motor Unit Potential		
			Fib	PW	Fasc	Recruitment	Dur/Amp	Phases
Rt	L4/5 paraspinal muscle	Normal	0	0	0			
	L5/S1 paraspinal muscle	Normal	0	0	0			
Lt	Gluteus maximus	Normal	0	0	0	Complete	Normal	-
	Adductor magnus	Normal	0	0	0	Reduced	Long	Inc
	Biceps femoris(short head)	Normal	0	0	0	Reduced	Normal	-
	Tensor fasciae latae	Normal	0	0	0	Complete	Normal	-
	Iliopsoas	Normal	0	0	0	Complete	Normal	-
	Vastus medialis	Normal	0	0	0	Reduced	Long	Inc
	Peroneus longus	Normal	0	0	0	Reduced	Long	Inc
	Tibilalis anterior	Normal	0	0	0	Complete	Normal	-
	Gastrocnemius	Normal	0	0	0	Reduced	Normal	-
	Extensor digitorum brevis	Normal	0	0	0	Not detectable		
	L4/5 paraspinal muscle	Normal	0	0	0			
	L5/S1 paraspinal muscle	Normal	0	0	0			

AH: Abductor hallucis, EDB: Extensor digitorum brevis, TA: Tibialis anterior, VM: Vastus medialis
NR: No response

Table 2. Electromyography in both lower extremities.

NERVE CONDUCTION STUDY				
Nerve Stimulation (Record)	Amplitude (uV) Distal / Proximal	Conduction Velocity (ms)	Distance (cm)	Latencies Distal / Proximal
Motor				
Rt. Median	6400/5700	56	25	3.7/8.2
Rt. Ulnar	12000/10100	59	27	2.67.2
Lt. Median	4700/4500	52	26	4.4/9.4
Lt. Ulnar	8700/6100	57	25	2.6/7.0
Rt. Tibial (AH)	2000/300	36	41	5.8/17.2
Rt. Peroneal (EDB)	300/300	40	35	9.5/18.3
Rt. Peroneal (TA)	600/600	60	6	5.9/6.9
Rt. Femoral (VM)	NR			3.6
Lt. Tibial (AH)	1000/900	40	38	5.8/15.3
Lt Peroneal (EDB)	NR			
Lt. Peroneal (TA)	NR			
Lt. Femoral (VM)	NR			
Sensory				
Rt. Median	20	51	14.5	2.8/4.1
Rt. Ulnar	18	52	12	2.3/3.1
Lt. Median	31	50	14	2.8/4.1
Lt Ulnar	19	52	12	2.3/2.9
Rt. Sural (Ankle)	14	41	9	2.2/3.1
Lt. Sural (Ankle)	11	41	9	2.2/3.1

Needle EMG

Muscles		Insertion activity	Spontaneous activity			Motor Unit Potential		
			Fib	PW	Fasc	Recruitment	Dur/Amp	Phases
Rt	Biceps brachii	Normal	0	0	0	Reduced	Long	Inc
	Flexor carpi radialis	Normal	0	0	0	Complete	Normal	-
	Abductor pollicis brevis	Normal	0	0	0	Complete	Normal	-
	Gluteus maximus	Normal	0	0	0	Complete	Normal	-
	Iliopsoas	Normal	+	+	0	Complete	Normal	-
	Tensor fasciae latae	Normal	+	+	0	Reduced	Long	Inc
	Vastus medialis	Normal	0	0	0	Reduced	Long	Inc
	Gastrocnemius	Normal	0	0	0	Reduced	Long	Inc
	Tibialis anterior	Normal	0	0	0	Poor volition		
	Semimembranosus	Normal	++	++	0	Reduced	Long	Inc
	L4/L5/S1 paraspinal muscle	Normal	0	0	0			
Lt	Gluteus maximus	Normal	0	0	0	Complete	Normal	-
	Iliopsoas	Normal	+	+	0	Reduced	Normal	-
	Tensor fasciae latae	Normal	0	0	0	Single MUAP		
	Vastus medialis	Normal	0	0	0	Markedly Reduced	Normal	-
	Gastrocnemius	Normal	0	0	0	Reduced	Long	Inc
	Tibialis anterior	Normal	0	0	0	No volition		
	Semimembranosus	Normal	++	++	0	Reduced	Long	Inc
	L4/L5/S1 paraspinal muscle	Normal	0	0	0			

AH: Abductor hallucis, EDB: Extensor digitorum brevis, TA: Tibialis anterior, VM: Vastus medialis

NR: No response