Case Studies

A Case of Apoplexy Attack-Like Neuropathy due to Hereditary Neuropathy with Liability to Pressure Palsies in a Patient Diagnosed with Chronic Cerebral Infarction

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Hereditary neuropathy with liability to pressure palsies is an inherited disease associated with the loss of a copy of the *PMP22* gene. The condition leads to mononeuropathy due to compression and easy strangulation during daily life activities, resulting in sudden muscle weakness and sensory disturbance, and displaying symptoms similar to cerebrovascular diseases. We report the case of an 80-year-old man with left paralysis due to chronic cerebral infarction. His medical history indicated remarkable recovery from about 4 months after the onset of left hemiplegia with predominant involvement of the fingers. Despite subsequent recurrent monoplegia of the upper or lower limbs, brain magnetic resonance imaging consistently revealed only previous cerebral infarction in the right corona radiata without new lesions. Medical examination showed reduced deep tendon reflexes in his extremities on both the healthy and hemiplegic sides. Nerve conduction studies showed delayed conduction at the bilateral carpal and cubital tunnels and near the right caput fibulae. Genetic analysis revealed loss of a copy of the *PMP22* gene. Thus, he was diagnosed with a cerebral infarction complicated by hereditary neuropathy with liability to pressure palsies. Stroke patients develop sudden muscle weakness and sensory disturbance. However, if such patients have no hyperactive deep tendon reflexes and show atypical recovery of paralysis that does not correspond to findings of imaging modalities, nerve conduction studies and genetic analysis may be necessary, considering the complication of hereditary neuropathy with liability to pressure palsies. **Key Words:** HNPP—*PMP22* gene—TIA—cerebral infarction.

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Received June 1, 2015; revision received December 24, 2015; accepted March 17, 2016.

Grant support: This work was supported by Grants-in-Aid from the Research Committee Charcot-Marie-Tooth Disease of the Ministry of Health, Labor and Welfare of Japan (15ek0109058h002) and JSPS KAKENHI Grant Number 26750211.

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1052-3057/$ - see front matter

http://dx.doi.org/10.1016/j.jstrokecerebrovasdis.2016.03.031

Case Report

An 80-year-old man with no remarkable family history visited our hospital for further rehabilitation of chronic cerebral infarction-induced left paralysis. He first experienced sudden left hemiplegia and was diagnosed with branch atheromatous disease of the right corona radiata by brain magnetic resonance imaging (MRI) 13 years previously. Although he only had slight movement of his left fingers at 4 months post onset, he recovered remarkably thereafter, regaining functionality in his hand and becoming ambulatory without aid. Subsequently, he experienced several events of sudden-onset monoplegia without a specific trigger. Although cerebrovascular diseases, including transient ischemic attack (TIA), were suspected, no new lesion was detected on brain MRI. He was diagnosed with TIA or radiculopathy each time, and all symptoms almost completely improved within several hours to 1 year.

At the first visit to our hospital, mild left hemiplegia and left drop foot, which had suddenly occurred a few weeks prior, were observed, along with reduced deep tendon reflexes in his extremities and a slightly reduced superficial sensation in the left anterior surface of the leg. The patient could independently perform all activities of daily living. A nerve conduction study revealed bilateral carpal tunnel syndrome, cubital tunnel syndrome, and right peroneal nerve paralysis (Table 1). Needle electromyogram showed neurogenic changes with active denervation findings localized in the left tibialis anterior muscle. Loss of a *PMP22* gene copy was found based on fluorescence in situ hybridization. Consequently, despite a history of stroke, the patient's left drop foot was determined to be caused by entrapment neuropathies associated with hereditary neuropathy with liability to pressure palsies (HNPP).

Discussion

HNPP is an autosomal-inherited disease associated with loss of a *PMP22* gene copy, in which slight compression can cause sudden muscle weakness and sensory disturbance of monoplegia.[1] Cerebrovascular diseases are characterized by acute unilateral muscle weakness and sensory disturbances. Therefore, it is necessary to distinguish between the sudden-onset monoplegia of HNPP and cerebrovascular diseases for appropriate treatment and rehabilitation.

Our patient had no family history or previous diagnosis of HNPP until the age of 80 years; however, HNPP symptoms are often unnoticed.[1,2] Recovery from hemiplegia after stroke is generally poor when more than 3 months have passed after the onset.[3] Our patient followed an atypical course; his finger paralysis improved rapidly at 4 months post stroke. The delayed improvement of the paralysis could possibly be because the patient was in a position associated with nerve compression during the acute phase of stroke.

Table 1. *Nerve conduction results*

	Motor			Sensory		
	MCV (m/s)	DML (ms)	Amp (mV)	Lat (ms)	SCV (m/s)	Amp (μV)
Rt med	46.3	10.65	3.1		No evoked response	
Lt med	55.4	6.10	6.8	4.15	33.7	8.0
Rt uln forearm	50.0	4.60	4.3		No evoked response	
elbow	29.2					
Lt uln forearm	54.3	4.25	5.3		No evoked response	
elbow	26.4					
Rt tib	42.0	4.15	0.1			
Lt tib	40.2	5.65	0.6			
Rt per lower leg	31.7	7.70	0.1			
fibular head	18.6					
Lt per		No evoked response				
Rt sural					No evoked response	
Lt sural					No evoked response	

Abbreviations: Amp, amplitude; DML, distal latency; elbow, ulnar nerve across the elbow; fibular head, peroneal nerve across the fibular head; Lat, latency; Lt, left; MCV, motor conduction velocity; med, median nerve; per, peroneal nerve; per lower leg, peroneal nerve in the lower leg; Rt, right; SCV, sensory conduction velocity; tib, tibial nerve; uln, ulnar nerve; uln forearm, ulnar nerve in the forearm.

Normal range: med: Motor: MCV > 48 m/s, DML < 4.2 ms, Amp > 3.5 mV; Sensory: SCV > 44 m/s, Lat < 3.5 ms, Amp > 19 μV. uln: Motor: MCV > 49 m/s, DML < 3.4 ms, Amp > 2.8 mV; Sensory: SCV > 44 m/s, Lat < 3.1 ms, Amp > 18 μV. tib: Motor: MCV > 41 m/s, DML < 6.0 ms, Amp > 2.9 mV. per: Motor; MCV > 40 m/s, DML < 5.5 ms, Amp > 1.0 mV. sural: Sensory: SCV > 39 m/s, Lat < 3.1 ms, Amp > 6.0 μV.

This study was approved by the local ethics committee for clinical research. The patient provided informed written consent.

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