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The Electrophysiologic and Ultrasonographic Change after Carpal Tunnel Release in a Patient with Hereditary Neuropathy with Liability to Pressure Palsy

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Abstract:

Hereditary neuropathy with liability to pressure palsy (HNPP) often manifests via peripheral nerve entrapment including median nerve compression at the carpal tunnel. However, there are few reports on surgical interventions for focal compression of the median nerve at the wrist in patients with HNPP. We report a case of a patient with HNPP who improved clinically, electrophysiologically, and ultrasonographically after carpal tunnel release (CTR). A 56-year-old woman presented with an 18-month history of pain in both thumbs. Nerve conduction study (NCS) revealed bilateral median neuropathy at the wrist. Ultrasonography demonstrated a markedly increased cross-sectional area (CSA) of both median nerves. Gene analysis revealed a deletion of the *PMP22* gene. She received bilateral CTR. Follow-up NCS at one year demonstrated the improvement of motor and sensory conduction; follow-up ultrasonography revealed significantly reduced CSA. Our case suggests that surgical decompression can be applicable to well-selected patients with HNPP.

Key Words:

Carpal tunnel syndrome, hereditary neuropathy with liability to pressure palsy, ultrasonography

Key Messages:

There are few reports regarding therapeutic surgical interventions for focal compression of the median nerve at the wrist in patients with HNPP. We report the case of an HNPP patient who improved clinically, electrophysiologically, and ultrasonographically after CTR. Surgical decompression needs to be considered as a treatment option to treat HNPP patients with CTS.

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Hereditary neuropathy with liability to pressure palsy (HNPP) is an autosomal dominant inherited neuropathy with recurrent motor and sensory nerve palsy, often triggered by minor compression.^[1] Carpal tunnel syndrome (CTS) is the most common entrapment neuropathy in the general population, characterized by sensory and/or motor nerve manifestations owing to median nerve compression beneath the flexor retinaculum of the wrist.^[1] In HNPP, the median nerve is also commonly affected by the wrist. In patients with CTS without inherited neuropathies, surgical intervention such as carpal tunnel release (CTR), is indicated in the case of severe weakness. However, there are few reports regarding therapeutic surgical interventions for focal

compression of the median nerve at the wrist in patients with HNPP.^[2]

Here, we report the case of one HNPP patient who improved clinically, electrophysiologically, and ultrasonographically after CTR.

Case Report

A 56-year-old bi-handed woman presented with an 18-month history of pain in both thumbs with a tingling sensation. On inspection, there was marked atrophic change in the left thenar muscles. On physical examination, Tinel's sign was positive and the sensation of both median nerve territories was altered on light touch and pinprick. Nerve conduction

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study (NCS) revealed bilateral severe median neuropathy at the wrist and suspicious left ulnar neuropathy [Table 1]. The distal latency of the right median nerve on motor NCS was significantly delayed. This finding led to an ultrasonographic (US) examination, which showed a markedly increased cross-sectional area (CSA) (right-29.6 mm², left-36.0 mm² at maximal swollen point) [Figure 1].

The multiple ligation probe amplification method for the *PMP22* gene (17p11.2-12) revealed a deletion that was diagnostic of HNPP. Considering that the electrophysiologic or ultrasonographic findings of the median nerve were more prominent than those of other nerves, we decided to perform bilateral CTR. The patient's symptoms such as pain and tingling sensation significantly improved immediately after surgery. Follow-up NCS at one year demonstrated improvement of both motor and sensory conduction with significant restoration in the distal latency of right median motor nerve [Figure 2 and Table 1]. In addition, follow-up US revealed significantly reduced CSA (right-20.5 mm², left-28.5 mm² at maximal swollen point) [Figure 1].

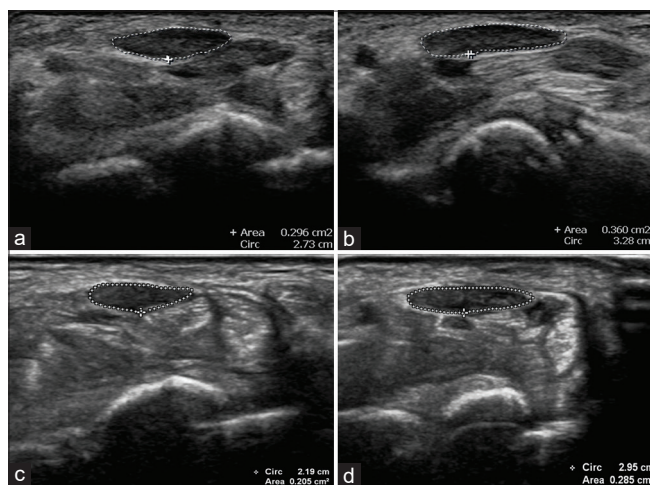


Figure 1: Ultrasonography (US) of the median nerves at the wrist. (a and b) The preoperative US showed a markedly increased cross-sectional area (CSA) of the median nerve at the wrist (right-29.6 mm², left-36.0 mm² at a maximal swollen point). (c and d) Follow-up US at 1 year after surgical decompression revealed a significantly reduced CSA (right-20.5 mm², left-28.5 mm² at a maximal swollen point)

Discussion

There are few reports regarding therapeutic surgical interventions for focal compression of the median nerve at the wrist in patients with HNPP. We report the case of an HNPP patient who improved clinically, electrophysiologically, and ultrasonographically after CTR. There were two important findings: first, surgical decompression might be an effective intervention in well-selected patients with HNPP and second, the role of the US during the diagnostic process of HNPP.

HNPP often manifests via peripheral nerve entrapments including median nerve compression at the carpal tunnel.^[3] Conventional treatment options for HNPP, such as restriction of repetitive movements, wrist splint or elbow pad, have been used to prevent focal compression or stretch injury of vulnerable peripheral nerves but data on its effectiveness are lacking.^[3] There are few reports of indications or effects

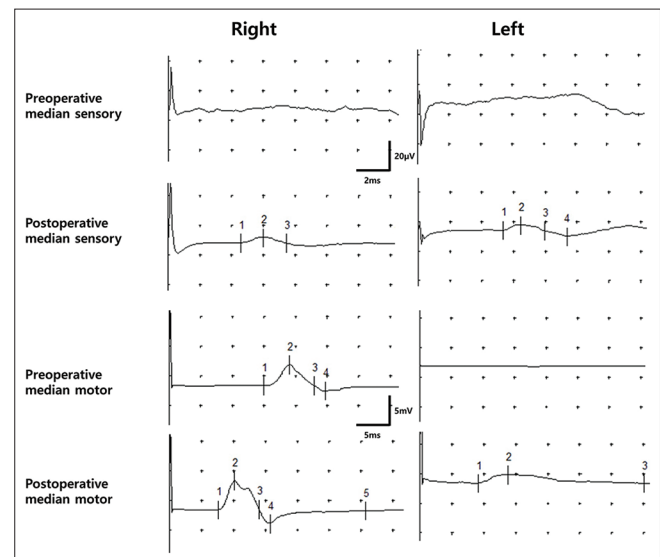


Figure 2: Pre and postoperative nerve conduction study of the median nerve. The sensory nerve action potential (SNAP) was antidromically recorded over the index finger with stimulation at wrist 14 cm away from the active electrode, and the compound motor action potential (CMAP) was recorded over thenar muscles with stimulation at the wrist 8 cm away from the active electrode. Note that the SNAP reappeared and the CMAP of the right median nerve underwent a significant recovery in distal motor latency on the postoperative study at 1 year

Table 1: Pre and postoperative electrophysiologic studies

Nerve	Recording Site	Preoperative NCS			Postoperative NCS		
		Latency (msec)	Amplitude (µV and mV)	Velocity (m/sec)	Latency (msec)	Amplitude (µV and mV)	Velocity (m/sec)
Sensory							
Rt. median	Digit-2	NR	NR		4.75	4.3	
Lt. median	Digit-2	NR	NR		5.35	3.5	
Rt. ulnar	Digit-5	2.85	10.7		3.20	9.0	
Lt. ulnar	Digit-5	3.80	7.6		3.25	8.0	
Motor							
Rt. median	APB	15.00	3.7	38.0	7.70	5.0	36.4
Lt. median	APB	NR	NR	NR	9.20	0.7	33.9
Rt. ulnar	ADM	3.55	9.0	49.4	3.80	8.5	49.3
Lt. ulnar	ADM	4.45	7.5	49.4	4.50	7.7	49.3

NCS=Nerve conduction study; NR=No response; APB=Abductor pollicis brevis; ADM=Abductor digiti minimi

of surgical decompression in patients with HNPP.^[2] Our case suggests that the demyelinating process combined with axonal loss may be reversible through surgical decompression. However, questions regarding when surgical decompression may be applied or to whom it may be applied still persist. Although many patients with HNPP have electrophysiologic evidence of median entrapment neuropathy, it is common to have no clinical symptoms. In the absence of symptoms, the effectiveness of preventive surgical decompression is unknown. If there are obvious symptoms and there is electrophysiological evidence of entrapment neuropathy, surgical decompression may be applicable to patients with HNPP as it is applied to patients with severe non-HNPP CTS.

The usefulness of the US in the diagnosis of HNPP is known to be limited.^[4-6] Many patients with HNPP have normal CSA even at vulnerable sites, and there is no cut-off value in the CSA that can distinguish HNPP and non-HNPP CTS. A recent meta-analysis reported that the cut-off value of 9 mm² of median nerve CSA yields the best diagnostic accuracy in CTS. Of the 22 CSA measurements of median nerve at the wrist in patients with HNPP, only three measurements exceeded 20 mm² of CSA.^[1,4] Considering previous ultrasonographic findings of CTS and HNPP patients, our case had extraordinary CSA of the median nerve at the wrist (right-29.6 mm², left-36.0 mm²). In our case, there was no electrophysiologic evidence of entrapment neuropathy except for both median nerves. Extraordinary CSA of median nerve on the US was a diagnostic clue to HNPP. Preoperative increase in the median nerve CSA would be a useful surgical indication because a previous US study suggested that increased CSA reflected partially reversible compression component in CTS.^[7] Further research is necessary to elucidate whether CSA may be related to responses after surgical decompression in HNPP.

Although the US alone has limited diagnostic value, it may be a diagnostic clue to HNPP if neurological examination and electrophysiological study is insufficient to conclude the diagnosis of HNPP. Furthermore, US may be a useful tool in monitoring CSAs after therapeutic interventions.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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