

Neuromuscular Disorder: Immune Mediated Neuropathy



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Immune mediated neuropathy

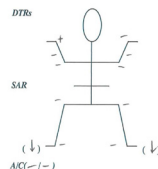
- Guillain-Barré syndrome
 - Acute inflammatory demyelinating polyneuropathy
 - AMAN
 - Reversible conduction failure
- Chronic acquired demyelinating polyneuropathy
 - Chronic inflammatory demyelinating polyneuropathy
 - MADSAM (multifocal acquired demyelinating sensory and motor neuropathy)
 - Multifocal motor neuropathy

Case I

- 여자, 86세
- 주호소: 양팔다리 힘이 빠짐
 - 입원 5일 전 양다리에 힘이 빠져 겨우 걸음
 - 입원 3일전 양손의 힘도 빠짐.
 - 입원 당일 수저질도 힘들 정도로 양팔의 힘이 빠짐. 하지근력저하하고 더욱 진행

Case I

- N/Ex
 - Motor
 - Neck – flx: GIV+; ext :GV-
 - U/Ext – Prox GIV+/GIV+
 - U/Ext – Dist GIV-/GIV-
 - L/Ext – Prox GIV-/GIV-
 - L/Ext – Distal GIV-/GIV-
 - Sensory
 - Touch – intact
 - Pinprick – Both G.toe 10/6
 - Position – intact
 - Vibration – Lt G.toe decreased



Case I

- CSF - WBC: 0, Protein: 54.8
- Nerve conduction study (motor)

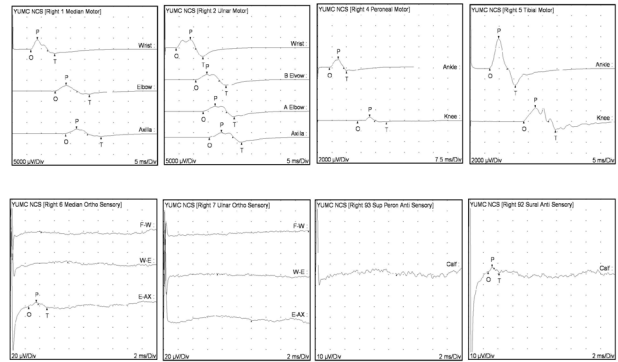
	Latency (msec)	Distance (cm)	CV (m/sec)	Amplitude (uV)
Right Median Motor				
TL	6.3	5		6500
W-E	14.8	21	24.7	4300
E-Ax	18.4	11	30.6	3700
Right Ulnar Motor				
TL	4.0	5		8100
W-E	10.8	20.5	30.0	5200
E	13.3	10	40.0	5000
Ax	15.4	7	33.3	6400
Right Peroneal Motor				
TL	6.2	5		2200
A-K	20.7	31	21.4	900
Right Posterior Tibial				
TL	6.8	5		8000
A-K	18.5	35	29.9	4200

Case I

• Nerve conduction study (sensory)

	Latency (msec)	Distance (cm)	CV (m/sec)	Amplitude (uV)
Right Median Sensory				
F-W	NR			
W-E	NR			
E-Ax	3.6	11.5	31.9	9.4
Right Ulnar Sensory				
F-W	NR			
W-E	NR			
E-Ax	NR			
Right Superficial peroneal				
TL	NR			
Right Sural				
TL	3.2	10	31.3	5

Case I



Case I

• Impression

- Acute inflammatory demyelinating polyneuropathy

• Treatment

- IVIg

Case I

Duration (Sx onset)	1st week	2nd week	5th week
INCAT score, total	5	3	3
Arm disability	2	1	1
Leg disability	3	2	2
MRC sum score	59	66	66
Hand grip power			
Right (kg)	12	14	14
Left (kg)	10	14	14
ISS	7	8	7
R-ODS	15	18	18

Case I

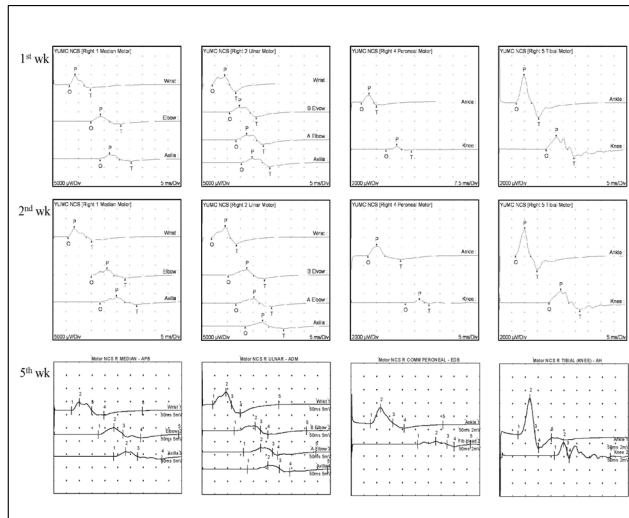
• Nerve conduction studies (Motor)

Duration (Sx onset)	2nd week		3rd week		5th week	
	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)
Right Median Motor						
TL	[6.3]	6500	[5.9]	6200	[6.8]	5000
W-E	24.7	4300	25.6	4300	20.3	4100
E-Ax	30.6	3700	25.7	4000	19.2	3000
Right Ulnar Motor						
TL	[4.0]	8100	[3.8]	8900	[4.2]	8000
W-E	30.1	5200	32.3	4800	24.9	3400
E	40.0	5000	30.0	4300	32.4	3000
Ax	33.3	4600	24.3	3500	30.0	2600
Right Peroneal Motor						
TL	[6.2]	2200	[6.7]	2900	[7.55]	3100
A-K	21.4	900	18.0	700	17.5	900
Right Posterior Tibial						
TL	[6.8]	8800	[6.5]	8500	[7.1]	7600
A-K	29.9	4200	27.5	4000	26.3	2500

Case I

• Nerve conduction studies (Motor)

Duration (Sx onset)	2nd week		3rd week		5th week	
	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)
Right Median Sensory						
F-W	NR		NR		NR	
W-E	NR		NR		NR	
E-Ax	31.9	9.4	NR		28.9	2.8
Right Ulnar Sensory						
F-W	NR		NR		NR	
W-E	NR		NR		NR	
E-Ax	NR		NR		NR	
Right Superficial peroneal						
TL	NR		NR		32.4	1.6
Right Sural						
TL	31.3	5.0	33.3	8.4	29.1	2.6

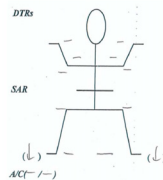


Case II

- 여자, 66세
- 주호소 : 양팔다리 힘이 빠진다.
 - 입원 8일전 cough, diarrhea, abdominal pain
 - 입원 1일전 전신에 힘이 없고 걸을 때 휘청거림
 - 입원 당일 전신에 힘없는 증상 더욱 심해짐
 - 호흡곤란

Case II

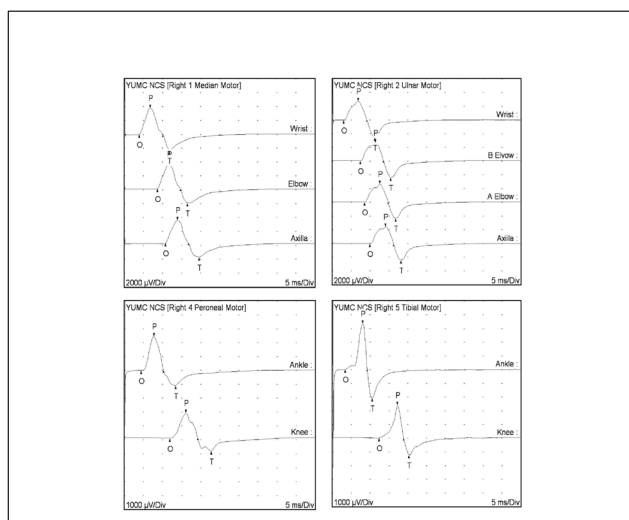
- N/Ex
 - Motor
 - U/Ext - Prox GIV/GIV+
 - U/Ext - Distal GIV-/GIV
 - L/Ext - Prox GIII/GIV-
 - L/Ext - Distal GIII/GIV-
 - Sensory - intact



Case II

- CSF - WBC: 0, Protein: 23.8
- NCS (normal sensory NCS)

	Latency (msec)	Distance (cm)	CV (m/sec)	Amplitude (uV)
Right Median Motor				
TL	3.9	5	[3.9]	6100
W-E	8.7	24.5	51	5600
E-Ax	10.8	12	57.1	5300
Right Ulnar Motor				
TL	3	5	[3]	5300
W-E	7.4	23.5	53.4	5100
E	8.5	6.5	59.1	5000
Ax	9.9	7.5	53.6	4900
Right Peroneal Motor				
TL	4.4	5	[4.4]	3600
A-K	11.9	33	44	2700
Right Posterior Tibial				
TL	3.4	5	[3.4]	5400
A-K	12.4	38	42.2	3600



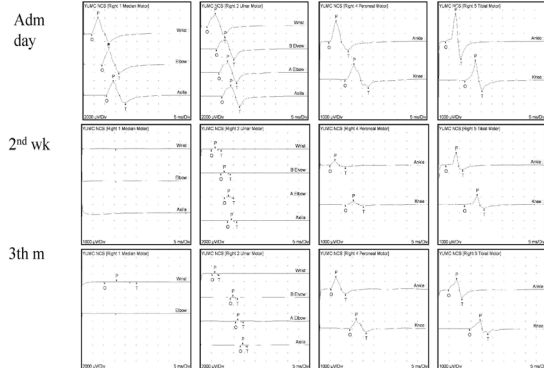
Case II

- Impression
 - Acute motor axonal neuropathy (AMAN)
- Treatment
 - IVIg
 - Mechanical ventilator

Case II

Duration (Sx onset)	Adm day	2nd week	4th week	3th month
INCAT score, total	10	10	10	10
Arm disability	5	5	5	5
Leg disability	5	5	5	5
MRC sum score	27	13	14	15
Hand grip power				
Right (Kg)	0	0	0	0
Left (Kg)	0	0	0	0
ISS	1	1	1	1
R-ODS	0	0	0	0

Case II

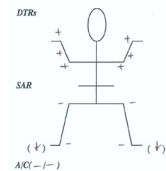


Case III

- 남자, 41세
- 주호소: 입원 2주전 양팔다리 힘이 빠짐.
 - 입원 3주전 감기
 - 입원 2주전 양쪽 발목과 발가락의 힘이 빠졌고 걷기 어려워짐. 다음날 젓가락 등 손을 쓰기도 불편해짐.
 - 입원 이틀 전부터 증상이 조금씩 회복됨.

Case III

- N/Ex
 - Motor
 - Neck - flx: GV; ext :GV
 - U/Ext - Prox GIV+/GIV+
 - U/Ext - Dist GIV+/GIV+
 - L/Ext - Prox GIV+/GIV+
 - L/Ext - Distal GIV-/GIV-
 - Sensory - Intact

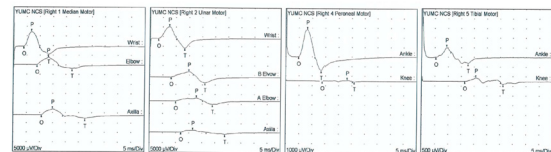


Case III

- CSF - WBC: 0, Protein: 53.5
- NCS (Normal sensory NCS)

	Latency (msec)	Distance (cm)	CV (m/sec)	Amplitude (uV)
Right Median Motor				
TL	3.8	5		11900
W-E	8.8	26.5	53	5700
E-Ax	10.6	10.5	58.3	4900
Right Ulnar Motor				
TL	3.3	5		11200
W-E	8.2	24.5	50	5700
E	9.9	8	47.1	3300
Ax	12	9.5	45.2	2100
Right Peroneal Motor				
TL	4.8	6		4300
A-K	13.8	36	40	200
Right Posterior Tibial				
TL	5.3	5		800
A-K	16.4	44.5	40.1	300

Case III



Case III

- Impression
 - Acute motor axonal neuropathy (AMAN)
 - AMAN with partial conduction block
- Decision
 - 경과 관찰
 - 일상생활이 가능한 정도의 경미한 신경학적 소실
 - 회복 중인 경과

Case III

- 증상발생 6주째 거의 정상으로 회복
 - 달리기만 약간 불편

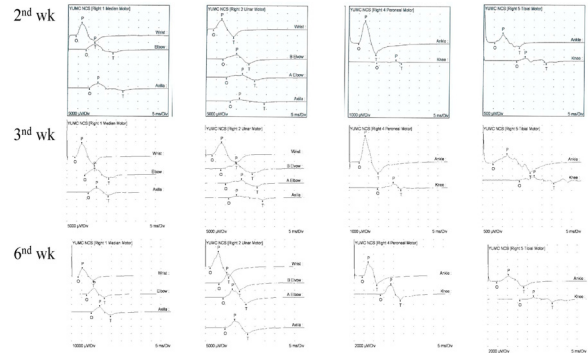
Duration (Sx onset)	2nd week	3rd week	6th week
INCAT score, total	3	2	1
Arm disability	2	1	0
Leg disability	1	1	1
MRC sum score	62	70	78
Hand grip power			
Right (kg)	16	22	40
Left (kg)	16	20	37
ISS	2	0	0
R-ODS	36	39	46
Inflammatory neuropathy cause and treatment sensory sum score (ISS); Total score of R-ODS = 48			

Case III

- Nerve conduction studies

Duration (Sx onset)	2nd week		3rd week		6th week	
	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)	CV (m/sec)	Amplitude (uV)
Right Median Motor						
TL	[3.8]	11900	[4.1]	13900	[3.2]	15700
W-E	53	5700	51	6300	60.5	12400
E-Ax	58.3	4900	52.8	5100	60.9	11200
Right Ulnar Motor						
TL	[3.3]	11200	[3.4]	12900	[2.3]	16500
W-E	50	5700	57	6600	54.4	11300
E	47.1	3300	36	4400	52.4	8900
Ax	45.2	2100	46.9	2800	55.6	8500
Right Peroneal Motor						
TL	[4.8]	4300	[5.1]	4900	[4.1]	5700
A-K	40	200	39.9	800	40.8	3900
Right Posterior Tibial						
TL	[5.3]	800	[5.2]	1200	[4.5]	2900
A-K	40.1	300	33.1	400	40.5	1300

Case III



Case IV

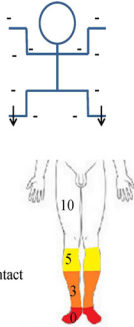
- 남자 64세
- C.C: 양팔다리 힘이 약하다. 2013년 4월부터
- Past Hx :
 - HTN/DM/pul.TBc./hepatitis (-/-/-/-)

Case IV

- 2013.4 월
 - 서서히 양 손, 발 끝 부터 저리기 시작. 감각이 둔함.
 - 점차 온몸이 저림
- 2013.7 월
 - 손목까지 감각저하 발생. 발바닥에 불이 나는 느낌.
 - 다리 힘이 없어 붙잡고 걸었고 지팡이 짚고 걸어야 했음.
 - 타 병원 내원, 허리 디스크로 판단하여 허리 수술 받았으나 호전 없었음.
- 2013.8월
 - 상기 증상에 대해 cervical OPLL 진단, 수술 받았으나 호전 없었음.
 - 감각저하가 팔과 무릎까지 진행. 근력은 더욱 약해져서 휠체어 ambulation.
- 2013.9월
 - 허벅지 까지 감각이상 증상 올라오는 양상
- 2013.10월
 - 타 병원 내원 CIDP 하에 steroid pulse, 경구 corticosteroid 2주간 사용
 - 호전되는 느낌은 특별히 없고 발바닥 통증만 약간 호전된 느낌

Case IV

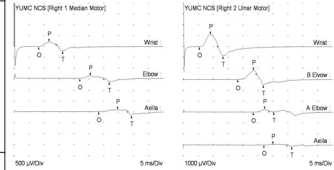
- N/E
 - Cranial nerve function - intact
 - Motor
 - UEx proximal GIV+/GIV+
wrist flx GIV/GIV ext GII/GIII
finger flx GIV/GIV spreading GII/GII
 - LEx. Proximal GIV-/GIV-
ankle df GI/GI Pf GII/GII
toe df GI/GI pf GI/GI
 - Sensory
 - Light touch : below knee 10:5, lower calf 10:3, foot 10:0
 - Pinprick or pain : below knee 10:5, lower calf 10:3, foot 10:0
 - Position : index finger : intact g.toe : impaired, both
 - Vibration : index finger : intact g. toe : impaired, both knee : intact



Case IV

- Nerve conduction study

Motor		Amp.(mV)	Vel.(m/s)
Median	Wrist	0.4	8.3*
	Elbow	0.3	15.7
	Axilla	0.3	18.8
	Wrist	2.0	5.4*
Ulnar	Below elbow	1.3	17.2
	Above elbow	0.3	23.1
	Axilla	0.2	19.4
	Wrist	NP	NP
Peroneal	Ankle	NP	NP*
	Knee	NP	NP
	Post. tibial	NP	NP*
	Knee	NP	NP
Sensory/Mixed		Amp.(μV)	Vel.(m/s)
Median	F-W	NP	NP
	W-E	NP	NP
	E-Ax	NP	NP
	F-W	NP	NP
Ulnar	W-E	NP	NP
	E-Ax	NP	NP
	F-W	NP	NP
	W-E	NP	NP
Sural	Calf	NP	NP
	Sup. Peroneal	NP	NP
	Calf	NP	NP
	Sup. Peroneal	NP	NP
F-wave		Latency(ms)	
Ulnar/Tibial		45.24	NP

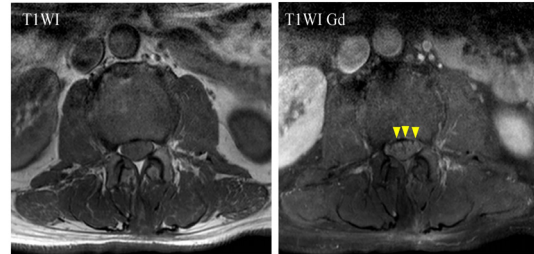


Case IV

- Lab
 - CSF study
 - WBC: 3; Protein 278.1 mg/dL
 - Protein electrophoresis & Immunofixation
 - 특이 소견 없음.
 - ANA – 1:40 negative
 - Anti ganglioside antibody: negative
 - GM1, GM2, GD1a, GD1b, GD3, GT1a, GT1b, GQ1b

Case IV

- L spine MRI



Gadolinium enhancement and hypertrophy of the cauda equina

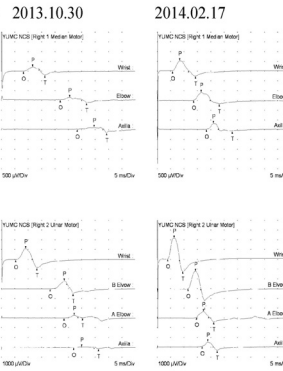
Case IV

- Dx
 - CIDP
- Tx
 - Prednisolone 30 mg/day
 - IVIg 400 mg/kg for 5 days (2013/11/1 시작)
 - 이후 1개월마다 IVIG 400mg/kg

Case IV

- 치료 경과
 - IVIg 시작하고 3일째 (2013/11/3)
 - “왼쪽 다리 불편했던 것이 좋아졌어요”
 - 이후 점차 근력이 회복
 - IVIg 시작하고 5일째 (2013/11/5)
 - 다리 근력 회복 중이며, 혼자서 앉아있을 정도로 회복됨.
 - 2014년 2월
 - 붙잡고 설 수 있을 정도로 호전.

Date	2013.10.30		2014.02.17	
Motor	Amp(mV)	Vel(m/s)	Amp(mV)	Vel(m/s)
Median				
Wrist	0.4	8.3*	0.8	5.8*
Elbow	0.3	15.7	0.6	29.1
Axilla	0.3	18.8	0.4	25.5
Ulnar				
Wrist	2.0	5.4*	3.4	4.0*
Below elbow	1.3	17.2	3.0	30.3
Above elbow	0.3	23.1	1.1	35.5
Axilla	0.2	19.4	0.9	33.3
Peroneal				
Ankle	NP	NP*	NP	NP*
Knee	NP	NP	NP	NP
Post. tibial				
Ankle	NP	NP*	NP	NP*
Knee	NP	NP	NP	NP
Sensory/Mixed	Amp(μV)	Vel(m/s)	Amp(μV)	Vel(m/s)
Median				
F-W	NP	NP	NP	NP
W-E	NP	NP	NP	NP
E-Ax	NP	NP	NP	NP
Ulnar				
F-W	NP	NP	NP	NP
W-E	NP	NP	NP	NP
E-Ax	NP	NP	NP	NP
Sural	NP	NP	NP	NP
Calf	NP	NP	NP	NP
Sup. Peroneal Calf	NP	NP	NP	NP



Case IV

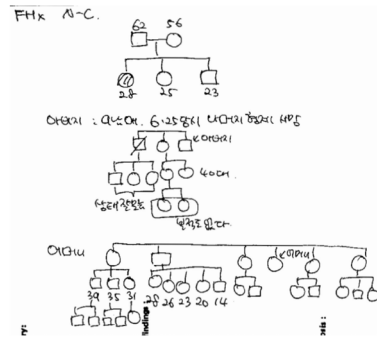
날짜	13.10.29	13.11.27	14.02.17
INCAT score total	8	7	7
Arm disability	3	3	3
Leg disability	5	4	4
Hand grip power			
Rt(kg)	0	2	2
Lt(kg)	0	2	2
ISS	6	5	5
R-ODS	8	9	9

Case V

- 여자 26세
- 주소: 왼쪽 foot drop, 왼손 힘 약하다
 - 1년전 복시 발생
 - 3개월전 왼발목이 잘 들리지 않기 시작
 - 1개월전 피아노 연주할 때 왼손이 약함
 - 점차 새로운 증상이 발생함.

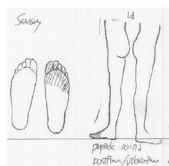
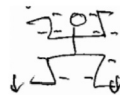
Case V

- 가족력 FHx A-C.



Case V

- Motor
 - Shoulder and: GV/GV
 - Elbow flex: GV/GV, ext: GV/GIV+
 - Wrist flex: GV/GIV+, ext: GV/GIV
 - Thumb abd: GV/GIV, finger spreading: GV/GIV
 - Hip flex: GV/GV, ext: GV/GV
 - Knee flex: GV/GV, ext: GV/GV
 - Ankle DF: GIV+/GIV-, PF: GIV+/GIV+
 - Heel gait: (↓-)
 - Toe gait: (↓/ ↓)

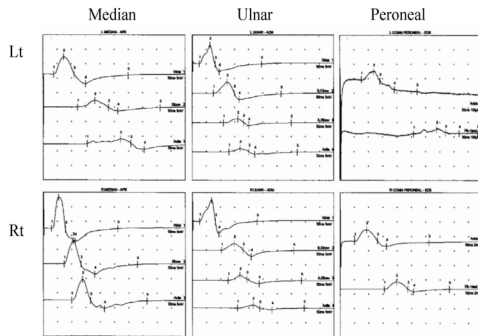


Case V

- Nerve conduction study (Motor)

	Left			Right		
	Latency (msec)	NCV (m/sec)	Amp (uV)	Latency (msec)	NCV (m/sec)	Amp (uV)
Median motor						
TL	3.7		9400	3.0		15300
W-E		26.2	3700		46.5	11900
E-Ax		36.8	3600		44.4	10200
Ulnar motor						
TL	2.55		9400	2.55		11800
W-BE		41.6	6000		25.8	4800
BE-AE		23.2	2500		29.8	3000
E-Ax		41.0	1900		27.3	1600
Peroneal motor						
F-A	7.6		100	5.15		2300
A-K		18.7	10		32.5	1400
Tibial motor						
F-A	6.6		4200	6.6		200
A-P		39.5	1100		40.0	100
H reflex	NP			NP		

Case V



Case V

• Nerve conduction study (Sensory)

	Left			Right		
	Latency (msec)	NCV (m/sec)	Amp (uV)	Latency (msec)	NCV (m/sec)	Amp (uV)
Median sensory						
F-W	3.0	40.0	12	2.95	42.4	11.1
Ulnar sensory						
F-W	2.55	41.2	18.1	2.80	35.7	7.7
Sural sensory						
	4.25	31.8	1.2	4.4	26.1	1.1

Case V

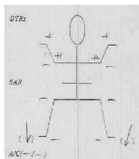
- Lab
 - Anti-GM1 Ab IbM – Negative
 - PMP22 – no deletion
 - ANA – 1:40 negative
 - Cerebrospinal fluid
 - WBC 0, protein 89 mg/dL
 - Protein electrophoresis & immunofixation
 - No monoclonal gammopathy

Case VI

- 여자, 34세
- 주호소: 양손에 힘이 없다. 왼손이 심하다.
 - 6개월전 – 아기 옷 입히면서 단추 끼우거나 지퍼 올릴 때 왼쪽 손에 힘이 떨어지는 것 발견
 - 점차 손의 힘이 더 약해진다.
 - 외부병원에서 C spine MRI 촬영 (정상)

Case VI

- Motor
 - Shoulder and: GV/GV
 - Elbow flex: GV/GV, ext: GV/GV
 - Wrist flex: **GIV/GIV**, ext: **GIV/GIV**
 - Thumb abd: **GIV/GIV**, finger spreading: **GIV/GIV**
 - Hip flex: GV/GV, ext: GV/GV
 - Knee flex: GV/GV, ext: GV/GV
 - Ankle DF: **GIV/GV**, PF: GV/GV
 - G.toe DF: **GII/GV**, PF: GV/GV
- Sensory: intact

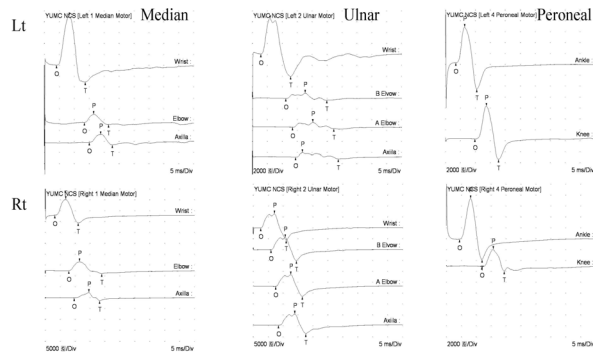


Case VI

• Nerve conduction study (Motor)

	Left			Right		
	Latency (msec)	NCV (m/sec)	Amp (uV)	Latency (msec)	NCV (m/sec)	Amp (uV)
Median motor						
TL	4.1		12100	3.2		10700
W-E		21.5	900		45.7	5500
E-Ax		62.5	900		58.3	3700
Ulnar motor						
TL	2.8		10600	2.5		12000
W-BE		26.2	1300		63.2	10600
BE-AE		45.5	1100		56.3	9900
E-Ax		50	1100		61.1	8900
Peroneal motor						
F-A	3.4		11300	4.4		11200
A-K		47.7	9700		44	4000
Tibial motor						
F-A	3.8		26500	3.8		20800
A-P		44.6	12700		44.1	9600
H reflex	NP			NP		

Case VI



Case VI

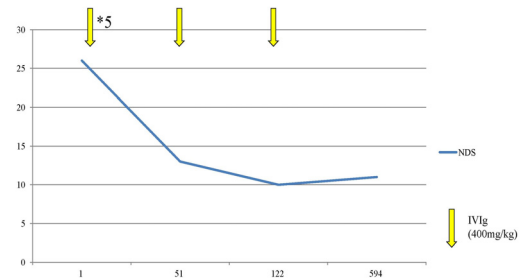
- Lab
 - Anti-GM1 Ab IbM – Positive
 - PMP22 – no deletion
 - ANA – 1:40 negative
 - Cerebrospinal fluid
 - WBC 0, protein 16.3 mg/dL
 - Protein electrophoresis & immunofixation
 - No monoclonal gammopathy

Case VI

- Impression
 - Multifocal motor neuropathy
- Decision
 - IV immunoglobulin treatment

Case VI

• Progress



Case VI

- Progress
 - IVIg 치료 중 임신 (임신 후 증상이 호전됨)
 - IVIg 치료 중단하고 경과 관찰
 - 출산 이후 팔힘이 약해진다.
 - 손끝의 저림도 호소
 - MADSAM 가능성 고려하여 corticosteroid 시도

Case VI

• Progress

