

CASE DESCRIPTION

Patient is a 26-year-old police officer with no past medical history who presented to the outpatient clinic for persistent right upper extremity weakness and pain, particularly in his 1st and 2nd digit. The weakness started approximately 7 months ago without history of trauma or inciting event but did recall similar symptoms in high school that self-resolved. The weakness had progressively worsened over the past several months and was starting to affect his duties at work such as difficulty firing his gun. The weakness was associated with numbness and tingling in the entire right hand. Nerve conduction study showed delay of the bilateral radial and median sensory responses, absent left ulnar sensory response, and reduced amplitude of the right sural sensory response. There was also slowing of the right peroneal, tibial, and left ulnar motor velocities. Patient was ultimately referred to neurology and was diagnosed with hereditary neuropathy with pressure palsy (HNPP).

Motor Nerve Study

Median Nerve Rec Site: APB STIM SITE Wrist	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)		C.V. (m/s)	
	L	R	L	R	L	R	L	R	L	R	L	R
	4.3	4.8	5.3	6.0	6.7	8.4	18.1	28.5	0	0	55.6	53.8
	8.8	9.6	5.7	6.1	6.2	8.2	17.6	27.7	250	260		
TEMPERATURE: Left side:35.5 C Right side:35 C												
Ulnar Nerve Rec Site: ADM STIM SITE Wrist	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)		C.V. (m/s)	
	L	R	L	R	L	R	L	R	L	R	L	R
	3.7	3.5	6.7	6.8	6.4	7.7	21.2	25.0	0	0	48.0	49.8
	9.1	8.9	7.8	7.1	6.0	6.8	21.3	25.6	260	270	44.2	48.6
	12.3	12.0	7.9	8.1	5.7	6.5	18.8	24.9	140	150		
Right Peroneal/Fib Nerve Rec Site: EDB STIM SITE Ankle	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)		C.V. (m/s)	
	L	R	L	R	L	R	L	R	L	R	L	R
	4.8		5.9		6.4		17.1		0		34.9	
	14.0		7.3		4.7		14.8		320		49.4	
	15.4		5.8		4.6		14.1		70			
Right Tibial Nerve Rec Site: AH STIM SITE Ankle	Lat (ms)		Dur (ms)		Amp (mV)		Area (mVms)		Dist (mm)		C.V. (m/s)	
	L	R	L	R	L	R	L	R	L	R	L	R
	4.2		8.8		7.1		20.2		0		34.8	
	17.7		8.9		6.6		24.5		470			

Sensory Nerve Study

Right Sural Nerve Rec Site: Ankle STIM SITE mid calf	Lat (ms)	Pk Lat (ms)	Amp (uV)	Dist (mm)	C.V. (m/s)
	3.4	4.3	5.3	0	

Sensory Nerve Study

Med/Uln/Rad Nerve Stim Site: Wrist REC SITE	Lat (ms)		Pk Lat (ms)		Amp (uV)		Dist (mm)	
	L	R	L	R	L	R	L	R
R Thumb	3.3	3.3	4.3	4.1	11.0	5.0	0	0
M Thumb	2.8	3.1	3.7	4.1	20.0	20.3	0	0
Index	3.2	4.0	4.3	5.0	10.3	13.0	0	0
5th dig	NR	4.2		3.5		3.9	0	0

EMG Study

Name	Normal	Ins Act	Fibs	PSW	Fascics	Polyph	MU Amp	MU Dur	Config	Pattern	Recruit
R. Tibialis An	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Peroneus Ln	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Gastroc.Med	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Vastus Lat.	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Biceps Ln.H	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Deltoid	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Deltoid	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Biceps Brac	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Biceps Brac	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Pronator Te	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Pronator Te	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Triceps	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Triceps	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. FDP U	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. FDP U	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. FDP M	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. FDP M	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Pronat. Qua	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Ext.Ind.Pro	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Dors.Int.1	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Dors.Int.1	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Abd.Pol.Br.	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Abd.Pol.Br.	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. Cervical Pa	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
L. Cervical Pa	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm
R. LS paraspin	Normal	norm	none	none	none	none	norm	norm	norm	norm	norm

Results

Delay of the left and right radial, and median sensory responses. Absent left ulnar sensory response. Normal ulnar right sensory response. Reduced amplitude of the right sural sensory response. Slowing of the right peroneal/fibular and tibial motor velocities. Slowing of the left ulnar motor velocities about the elbow. Normal right ulnar motor response. Delay of the right median motor distal latency. Normal left median motor study. Normal F-waves as listed. Normal EMG of the selected upper extremity of C5-T1 muscles bilaterally and cervical paraspinals. Normal EMG of the selected right lower L2-S2 muscles and paraspinals.

DISCUSSION

Prevalence of HNPP is extremely rare with studies reporting as low as 2-16 persons per 100,000 with the majority of cases resulting from a deletion of the peripheral myelin protein 22 gene (PMP22). Patients can typically present with sensory and motor deficits in nerves more prone to compression such as the peroneal nerve at the fibular head. While genetic testing is confirmatory, electrodiagnostics is a useful tool and will typically show evidence of generalized primarily demyelinating motor and sensory peripheral polyneuropathy. Treatment consists of preventative and symptom reducing measures.

CONCLUSION

This case highlights the importance of using a comprehensive clinical and electrodiagnostic approach in unclear cases of neuropathy such as HNPP.

REFERENCES

- Chance PF. Inherited focal, episodic neuropathies: hereditary neuropathy with liability to pressure palsies and hereditary neuralgic amyotrophy. *Neuromolecular Med.* 2006;8(1-2):159-174. doi:10.1385/NMM:8:1:159
- Heng H, Tang S, Goyal S, Wraige E, Lim, M. Beneficial use of steroids in hereditary neuropathy with liability to pressure palsy. *Developmental Medicine & Child Neurology.* 2011;54: 183-186. doi:10.1111/j.1469-8749.2011.04131.x
- Paprocka J, Kajor M, Jamroz E, Jespłowicz J, Seeman P, Marszał E. Case report
Hereditary neuropathy with liability to pressure palsy. *Folia Neuropathologica.* 2006;44(4):290-294.
- Topkian R, Wimmer S, Pischinger B, Pichler R. Hereditary neuropathy with liability to pressure palsies presenting with sciatic neuropathy. *BMJ Case Rep.* 2014;2014:bcr2014206883. Published 2014 Oct 17. doi:10.1136/bcr-2014-206883