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Sural sparing reversible sensorimotor abnormalities in hypokalemia: A close mimic of GBS

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Abstract

Nerve conduction studies (NCS) in hypokalemic paralysis usually show decreased compound muscle action potential (CMAP) which improves with correction of hypokalemia and normalizes in between the attacks. Inactivity of Na+ K+ ATPase pump due to hypokalemia results in decreased excitability of muscle fibers resulting in reduced CMAP on NCS. Sensory symptoms, as well as abnormalities in sensory NCS is a rare phenomenon. Here we describe a 40-year-old male who presented with acute onset rapidly progressive ascending quadriparesis with type 2 respiratory failure due to hypokalemia following hepatorenal dysfunction. NCS done before hypokalemia correction, suggested sural sparing with generalized reduced CMAPs, nonrecordable median and ulnar nerves sensory nerve action potential (SNAP) and abnormal F waves. After hypokalemia correction there was improvement in CMAPs as well as SNAPs. Thus, hypokalemia can present as an axonal sensorimotor involvement with sural sparing on NCS study similar to Guillain-Barré syndrome subtype.

Keywords: Hypokalemia, sural sparing, sensory NCS, reversibility.

INTRODUCTION

Rapidly progressing ascending flaccid quadriplegia can be a presenting symptom of Guillain-Barré syndrome (GBS) and GBS mimics like hypokalemia, acute spinal cord injury, acute CIDP, porphyria. Sural nerve-sparing and reduced or absent SNAP is a discriminating feature of AIDP subtype of GBS from GBS mimics.[1] Sural sparing is seen in axonal as well as demyelinating forms of GBS.[2] Inactivity of Na+ K+ ATPase pump due to hypokalemia results in decreased excitability of muscle fibers, resulting in a reduced CMAP in NCS though sensory conductions are usually unaffected. We present a patient with abnormal sensory nerve conductions and rapidly progressive ascending flaccid quadriplegia due to hypokalemia mimicking GBS.

CASE REPORT

A 40-year-old male presented to the emergency with complaint of multiple episodes of vomiting and fever for 2 days, yellowish discoloration of urine with acute onset rapidly progressing (reaching nadir within 6 hours) ascending quadriplegia without any positive or negative sensory symptoms and decreased sensorium with respiratory failure requiring mechanical ventilation. At admission, Glasgow Coma Scale (GCS) was E2V1M2 with pCO2 of 99% with pH of 7.10, serum sodium was 138 meq/l, serum potassium was 2.84 meq/l. Electrocardiogram was within normal limits. On examination, he was unconscious, not following verbal commands without apparent cranial nerve involvement. All extremities were hypotonic with power 0/5, with left lower limb short and wasted due to previous polio. All deep tendon jerks and plantar reflexes were bilaterally absent. The sensory system, cerebellum, and gait could not be assessed due to poor GCS. Two provisional diagnoses for quadriplegia were kept as 1) hypokalemic paralysis 2) GBS.

Nerve conduction study was done at baseline (Table 1) and was suggestive of reduced CMAPs in bilateral median, ulnar and tibial nerves. Prolonged distal latency was observed in bilateral tibial nerves while peroneal nerves were unrecordable. Nerve conduction velocities were normal in the recorded nerves. Bilateral median and ulnar nerves SNAPs were unrecordable with normal sural SNAPs. After correcting hypokalemia, the patient recovered completely with power of 5/5 in extremities and normal

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Table 1: Comparison of nerve conduction study before and after treatment of hypokalemia

	Nerve Conduction Studies Prior To Potassium Correction				Nerve Conduction Studies Two Days After Potassium Correction			
Motor Nerves	***Latency (ms)***	***Amplitude (mv)***	***Velocity (m/s)***	***F Latency***	***Latency (ms)***	***Amplitude (mv)***	***Velocity (ms)***	***F Latency***
R Median	1.9	2.2	39.9	17.3	3.0	1.0	87.3	22.4
L Median	1.9	1.9	69.9	22.0	4.2	3.9	54.2	22
R Ulnar	1.9	2.0	79.9	59.9	1.2	9.3	75.3	24.1
L Ulnar	2.1	1.0	65.8	20.4	2.4	5.8	72.6	25
R Peroneal	NR	NR	NR	NR	3.4	4.5	61.4	33.4
L Peroneal	NR	NR	NR	NR	3.0	7.5	69.8	25.5
R Tibial	5.4	4.9	82.7	NR	2.6	21.2	56.5	41.5
L Tibial	4.1	2.4	72.7	29	2.2	16.1	58.6	38.8
Sensory Nerves	***Latency (ms)***	***Amplitude (μV)***	***Velocity (m/s)***		***Latency (ms)***	***Amplitude (μV)***	***Velocity (m/s)***	
R Ulnar	NR	NR	NR		1.1	11.8	45.5	
L Ulnar	NR	NR	NR		1.5	9.6	80	
R Median	NR	NR	NR		1.8	17.6	68	
L Median	NR	NR	NR		1.6	11.5	70.3	
R Sural	1.5	21.7	89.7		2.4	33.5	55.3	
L Sural	1.7	24.6	76.5		2.0	5.0	65	

muscle stretch reflexes in the above mentioned muscles. Respiratory function also improved and he was extubated after 12 hours. Post correction NCS also suggested of improvement in CMAPs and SNAPs in previously involved nerves.

Additionally, he was evaluated for pyrexia (with detected thrombocytopenia and hepatorenal dysfunction). Tests for tropical fever including dengue serology, Typhi-dot IgM, Pan malaria smear, Scrub serology, Lyme serology, Leptospira serology - were negative. SARS COVID-19 rt-PCR was negative though COVID IgG / IgM antibody was positive suggesting recent infection. Hepatitis A, B, C, E and HIV 1/2 were non-reactive. Serology for CCHF was negative. Empirically injection ceftriaxone and oral doxycycline were started in view of tropical fever syndrome and the hepatorenal dysfunction improved completely.

DISCUSSION

In hypokalemic paralysis, electrophysiological studies usually show decrease in the CMAP amplitude that improves with correction of hypokalemia and persists in between the attacks.[3] At the resting state of a neuron, the electrical potential of its cytoplasm is more negative than the electrical potential of extracellular fluid by approximately 70 mV. This difference in the resting membrane potential (RMP) is dependent on the markedly different intracellular and extracellular concentrations of Na+, K+, and chloride ions. The large differences between external and internal concentrations of Na+ and K+ play a vital role in the production of action potentials. One of the major factors for the variance between intracellular and extracellular ion concentrations is the presence of an active transport mechanism, the Na+ K+ ATPase pump. These ion transport mechanisms are deranged by potassium deficiency due to its effect on enzyme systems.[4-6] Inactivity of Na+ K+ATPase pump due to hypokalemia results in decreased excitability of muscle fibers, resulting in reduction of CMAPs in NCS.[7]

Sural-sparing NCS is seen in axonal as well as demyelinating forms of GBS. The presence of abnormal sural SNAP with normal upper limb SNAPs in the initial NCS of a patient with suspected GBS should prompt the electro-diagnostician to question the diagnosis regarding the GBS subtype.[2] As recently demonstrated, the pathology of acute motor axonal neuropathy (AMAN) is not confined to length-dependent wallerian-like degeneration, where the sural nerve is most affected, but also includes reversible conduction failure (RCF).[8] These two pathologies are likely to coexist in the same patient at different

periods of the illness. The immunopathology at the node and paranode leads to loss of sodium channels that can lead to RCF and if severe, wallerian-like degeneration. Sural sparing in myelinopathy like acute inflammatory demyelinating polyneuropathy (AIDP) as well as the axonopathy as AMAN can be explained by the proposed hypothesis that the immunological injury in GBS is maximum at areas with a disrupted blood-nerve barrier as is also present sub-clinically in common entrapment syndromes, namely carpal tunnel syndrome and ulnar neuropathy at the elbow. The sural nerve, not commonly affected by entrapment, is hence spared. The second hypothesis is that the distal end of any nerve is most affected in GBS. As such the sural nerve is recorded near the lateral malleolus, some distance proximal to its terminal end.[2]

As in the present case, Rai *et al*. and Inshasi *et al*. also observed non recordable SNAP during hypokalemia in NCS.[7,9] Inshasi *et al*. proposed inactivity of Na+ K+ ATPase pump at the dorsal root ganglia because of the paucity of blood-nerve barrier as the mechanism for the absent SNAPs.[9] Rai *et al*. proposed that inactivation of pump over entire nerve fiber explains the distal absent SNAPs.[7] However, selective nerve involvement is questionable as sural nerves NCS were bilaterally normal. Similar to Rai *et al*., our patient had no sensory symptoms on admission though NCS was abnormal- a pattern seen in GBS and rarely in hypokalemia. Thus electrophysiological methods are more sensitive than clinical assessment for sensory evaluation and probably detects the abnormalities earlier. Also, single NCS is not helpful in differentiating hypokalemia-induced acute flaccid quadriplegia from AMSAN variant of GBS.

In conclusion, the present case highlights that sural sparing with sensory nerve involvement alongside reduced CMAPs can be a NCS feature of hypokalemic paralysis. This can be misdiagnosed on a single NCS as an AMSAN variant of GBS. Therefore, serial NCS should be done to differentiate hypokalemia with respiratory failure from AMSAN variant of GBS.

DISCLOSURE

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REFERENCES

1. Derksen A, Ritter C, Athar P, *et al*. Sural sparing pattern discriminates Guillain-Barré syndrome from its mimics. *Muscle Nerve* 2014;50(5):780-4. DOI: 10.1002/mus.24226
2. Umapathi T, Li Z, Verma K, *et al*. Sural-sparing is seen in axonal as well as demyelinating forms of Guillain-Barré syndrome. *Clin Neurophysiol* 2015;126(12):2376-80. DOI: 10.1016/j.clinph.2015.01.016
3. Rajshekher G, Kumar S, Prabhakar S. Reversible electrophysiological abnormalities in hypokalemic periodic paralysis. *Ind Pediatr* 2008;45:54-5. DOI: 10.4103/0972-2327.128566
4. Knochel JP. Neuromuscular manifestations of electrolyte disorders. *Am J Med* 1982;72:521-35. DOI: 10.1016/0002-9343(82)90522-8
5. Daut J. Inhibition of the sodium pump in guinea-pig ventricular muscle by dihydroouabain. Effects of external potassium and sodium. *J Physiol* 1983;339:642-62. DOI: 10.1113/jphysiol.1983.sp014740
6. Cruz-Martínez A, Arpa J. Muscle fiber conduction velocity in situ in hypokalemic periodic paralyses. *Acta Neurol Scand* 1997;96:229-35. DOI: 10.1111/j.1600-0404.1997.tb00274.x
7. Rai NK, Singh R. Reversible sensory motor conduction abnormalities in hypokalemic paralysis. *J Clin Neuromus Dis* 2013;15(2):44-6. DOI: 10.1097/CND.0000000000000013
8. Uncini A, Ippoliti L, Shahrizaila N, *et al*. Optimizing the electrodiagnostic accuracy in Guillain-Barré syndrome subtypes: criteria sets and sparse linear discriminant analysis. *Clin Neurophysiol* 2017;128(7):1176-83. DOI: 10.1016/j.clinph.2017.03.048
9. Inshasi JS, Jose VP, van der Merwe CA, *et al*. Dysfunction of sensory nerves during attacks of hypokalemic periodic paralysis. *Neuromusc Dis* 1999;9:227-31. DOI: 10.1016/s0960-8966(98)00129-1