Reversible electrophysiological abnormalities in hypokalemic paralysis: Case report of two cases

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Abstract

Compound muscle action potential (CMAP) amplitude declines during a paralytic attack in patients with hypokalemic periodic paralysis (HPP). However, serial motor nerve conduction studies in hypokalemic paralysis have not been commonly reported. We report two cases with hypokalemic paralysis, who had severely reduced CMAPs in all motor nerves at presentation during the episode of quadriparesis. However, the amplitude of CMAPs increased and reached normal levels, as the serum potassium concentration and motor power returned to normal state.

Key Words

Compound muscle action potential, electrophysiology, hypokalemic periodic paralysis, motor conduction abnormalities

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Introduction

Electrophysiological abnormalities reported in hypokalemic periodic paralysis (HPP) include reduced compound muscle action potential (CMAP) amplitude during a paralytic attack, increased CMAP amplitude 5 min after maximal muscle contraction, progressive reduction in amplitude 20-40 min after rest[1] and rarely reduced sensory nerve action potential (SNAP) amplitudes.[2]

Sensory nerve conduction (NC) abnormalities that reversed with hypokalemia correction and improvement in muscle weakness have been reported;[2] however, similar findings in motor NC studies have been uncommonly reported. We report case reports of two patients with reversible motor conduction abnormalities.

Case Reports

Case 1

A 20-year-old girl with no significant past medical illness presented with flaccid are flexic quadriparesis of 2 day duration. She had bulbar and respiratory involvement in form of dysphagia and her single breath count was nine. She also had cranial nerve involvement in form of bilateral facial weakness, neck flexor weakness, and dysphagia. The power in all muscle groups was 1/5. There was no sensory loss, bladder disturbance, or higher function disturbance. She was evaluated for electrolyte abnormalities (suspecting a possible diagnosis of hypokalemic paralysis). The electrocardiography (ECG) showed U waves. Arterial blood gas analysis showed normal pH, normal partial pressures of oxygen and carbon dioxide, and normal bicarbonate levels. Simultaneously, she underwent NC study. The NC study showed significant reduction in CMAP amplitudes of all motor nerves. The distal latencies and conduction velocities were normal in the motor nerves. However, the F-waves were non recordable in all the tested motor nerves. SNAP were also normal. Based on the electrophysiological findings, an alternate diagnosis of acute motor axonal neuropathy (AMAN) form of Guillain-Barre syndrome (GBS) was also considered. In the meantime serum potassium report was available and was 1.9 mmol/L. With administration of potassium (160 meq intravenous over 24 h followed by 60 meq per day in three divided doses orally in form of potassium chloride) patient showed dramatic clinical improvement. Based on this, the original diagnosis of hypokalemic paralysis with quadriparesis with cranial nerve and respiratory involvement was confirmed. NC studies were repeated 24 h after the initial study, by which time patient's motor power was back to normal and serum potassium was 2.9 mmol/L. Repeat NC was found to be normal. The secondary causes of hypokalemia including thyrotoxicosis, renal tubular acidosis, gastrointestinal loss were ruled out (FT3-3.20 pg/mL, FT4-1.10 ng/dL, TSH – 3.80 uIU/mL, arterial pH – 7.45). The short and long duration exercise test to document channelopathy

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as cause of hypokalemia was planned, but patient did not give consent for the same. As no definite secondary cause of hypokalemia could be found, the patient was not put on any prophylactic therapy. The patient was advised against heavy manual labour and avoiding heavy carbohydrate meals.

Case 2

A 52-year-old male with no past medical illness presented with flaccid areflexic quadriparesis of 1 day duration. There was no sensory loss, cranial nerve involvement, bladder disturbance, or higher function disturbance. The power in upper limb muscle groups was 2/5 and in the lower limb muscle groups was 3/5. The ECG done in the emergency ward showed U waves. He was evaluated for electrolyte abnormalities (suspecting a possible diagnosis of hypokalemic paralysis) and while the reports were still pending, he underwent NC study. The NC study showed significant reduction in CMAP amplitudes of all motor nerves. However, distal latencies and conduction velocities of motor nerves were normal. F-wave latencies were non recordable in all tested motor nerves. SNAP amplitudes were also normal. Based on the electrophysiological findings, an alternate diagnosis of AMAN form of GBS was also considered. The serum potassium report was 2.0 mmol/L. With administration potassium (160 meq intravenous over 24 h followed by 60 meq per day in three divided doses orally in form of potassium chloride), the patient showed dramatic clinical improvement. Based on this, the original diagnosis of hypokalemic paralysis with quadriparesis was confirmed. NC study was repeated 24 h after the initial study, by which time the patient's motor power was back to normal and serum potassium was 3.9 mmol/L. Repeat NC study was found to be normal. The secondary causes of hypokalemia including thyrotoxicosis, renal tubular acidosis, gastrointestinal loss were ruled out (FT3-3.09 pg/mL, FT4-1.03 ng/dL, TSH – 2.90 uIU/mL, arterial pH – 7.4) The short and long duration exercise test to document channelopathy as cause of hypokalemia was planned, but patient did not give consent for the same. As no definite secondary cause of hypokalemia could be found the patient was not put on any prophylactic therapy. The patient was advised against heavy manual labor and avoiding heavy carbohydrate meals.

The NC study report before and after correction of hypokalemia and recovery of muscle power depicting the changes in the CMAP of the motor nerves.

As there was no asymmetry between nerve conduction findings of left and right side, only right-sided nerves are described for sake of convenience [Table 1].

Discussion

Reversible electrophysiological abnormalities of sensory nerve function have been reported earlier.[2] A prospective study in 10 patients with HPP revealed a pattern of reduced sensory action potentials during paralytic attacks, which normalized with correction of serum potassium. The mechanism could be related to the dorsal root ganglia having an incomplete blood-nerve barrier, and neuronal inexcitability was postulated to occur consequent upon possible inactivation of the sodium-potassium pump by the low concentration of extracellular potassium. These authors could not find any abnormalities in the motor nerve function.

In another study on muscle fiber conduction velocity (MFCV) in patients with hypokalemic weakness of various etiologies, Cruz-Martinez and Arpa[3] found inexcitability of most muscle fibers during an acute attack, with associated slowing of MFCV. They also found increased threshold in the axons, consistent with hyperpolarization. Activity-dependent conduction block was induced by voluntary contraction and excitability abnormalities resolved with potassium replacement.

In our patients, the predominant finding on motor conduction studies was the severe reduction in amplitudes of the CMAPs and nonrecordable F-wave latencies. The sensory conduction studies were normal. The most dramatic finding in our study is the progressive improvement in the CMAP amplitudes and normalization of F-wave latencies with the administration of potassium and improvement of motor power, reaching normal levels by 24 h with normalization of motor power.

To our knowledge, there has been only a single case report before this, of motor nerve CMAP amplitude changes in hypokalemic weakness, with serial study showing normalization of the abnormalities with correction of serum potassium.[4] But in that case report, the patient did not have bulbar or respiratory involvement and there were no abnormality in the F-wave latency. Based on published reports, we believe that hypokalemia-induced inactivity of the sodium-K-ATPase leading to inexcitability of muscle fibers underlies the abnormalities detected in our patients. Hyperpolarization of muscle fibers occurs with decreased extracellular potassium concentration, although in severely reduced potassium levels, myofibers paradoxically depolarize to a stable potential of −60 mV from their baseline potential of −85 mV.[5] This results in failure of excitation of muscle fibers by supramaximal stimulation of peripheral nerves, thus resulting in decreased

Table 1: Nerve conduction studies

Patient nerve (right)	First								Second							
	CMAP amplitude (mV)		Conduction velocity (m/s)		Distal latency (ms)		F wave latency (ms)		CMAP amplitude (mV)		Conduction velocity (m/s)		Distal latency (ms)		F-wave latency (ms)	
	B	A	B	A	B	A	B	A	B	A	B	A	B	A	B	A
Median	1.8	11.4	59	59	2.6	2.6	NR	23.1	1.8	11.0	50	56	4.2	4.3	NR	29.9
Ulnar	0.6	6.8	64	64	2.4	2.6	NR	22.5	2.1	11.4	49	50	2.6	3.2	NR	28.6
Tibial	2.4	12.8	49	46	3.4	4.1	NR	39.8	2.0	10.6	43	46	4.0	3.9	NR	54.3
Peroneal	1.0	3.9	55	53	4.2	4.4	NR	40.4	0.3	6.9	48	52	3.4	3.5	NR	47.8

B=Before treatment, A=After treatment, mV=Milli-volt, ms=Milli-second, m/s=Meter/second, NR=Nonrecordable, CMAP=Compound muscle action potential

CMAP of the tested nerves. Also during acquired hypokalemic paralysis, there is axonal hyperpolarization, which reverses after serum potassium becomes normal.[6] This could explain the decreased CMAP of peripheral nerves and its recovery after correction of hypokalemia. Such abnormalities are not more often detected in patients presenting with hypokalemic weakness probably because nerve conduction studies are not routinely performed in these patients. This report highlights the fact that NC studies can be misleading in patients presenting with flaccid quadriparesis, especially when there is no reason to suspect hypokalemia. One should suspect a diagnosis of hypokalemic weakness in a patient presenting with NC features of AMAN variant of GBS.

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