

Reversible Electrophysiological Abnormalities in Hypokalemic Periodic Paralysis

GARIKAPATI RAJSHEKHER
SUDHIR KUMAR
SUBHASHINI PRABHAKAR

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 HPP have not been commonly reported. We report a 9-year-old girl with HPP, 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: *Electrophysiology, Hypokalemic periodic paralysis, Motor conduction abnormalities.*

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 minutes after maximal muscle contraction, progressive reduction in amplitude 20-

40 minutes after rest(1) and rarely reduced sensory nerve action potential (SNAP) amplitude(2). Sensory nerve conduction abnormalities that reversed with hypokalemia correction and improvement in muscle weakness have been reported(2), however, similar findings in motor nerve conduction studies have been uncommonly reported. We present a patient with reversible motor conduction abnormalities.

CASE REPORT

A nine-year old girl with distal renal tubular acidosis due to nephrocalcinosis presented with flaccid areflexic quadriparesis of four hours duration. There was no sensory loss, cranial nerve involvement, bladder disturbance or higher function disturbance. She was evaluated for electrolyte abnormalities (suspecting a possible diagnosis of HPP) and while the reports were still pending, she underwent nerve conduction (NC) study. The NC study showed significant reduction in CMAP amplitudes of all motor nerves. However, the distal latencies, conduction velocities and F-wave latencies were within normal limits. Sensory nerve action potential (SNAP) amplitudes 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. With administration of the routine dose of potassium, the patient showed dramatic clinical improvement. The serum potassium report was now available and was 2 mmol/L. Based on this, the original diagnosis of HPP with quadriparesis was confirmed. Nerve conduction (NC) studies were repeated 2 hours after the initial study and again 24 hours later, by which time the patient's motor power was back to normal. Repeat NC studies were found to be normal.

DISCUSSION

Reversible electrophysiological abnormalities of sensory nerve function have been reported earlier(2). A prospective study in ten patients with

From the Department of Neurological Sciences, Apollo Hospitals, Hyderabad, India.

*Correspondence to: Dr Sudhir Kumar, Consultant Neurologist, Apollo Hospitals, Jubilee Hills, Hyderabad 500 033, Andhra Pradesh, India.
E-mail: drsudhirkumar@yahoo.com.*

*Manuscript received: May 24, 2007;
Initial review completed: July 31, 2007;
Revision accepted: August 24, 2007.*

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 did 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, *et al.*(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 K⁺ replacement.

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

To our knowledge, this is the first report of motor nerve CMAP amplitude changes in hypokalemic weakness, with serial studies showing normalization

of the abnormalities with correction of serum potassium. 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 patient. 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 quadriplegia, 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.

Contributors: GR, SK, SP were involved in diagnosis and management of the case, GR prepared the initial draft of the paper, SK/SP made significant additions/revisions and all approved of the final version of the paper.

Funding: None.

Competing interests: None stated.

REFERENCES

1. Graves TD, Hanna MG. Neurological channelopathies. *Postgrad Med J* 2005; 81: 20-32.
2. Inshasi JS, Jose VP, van der Merwe CA, Gledhill RF. Dysfunction of sensory nerves during attacks of hypokalemic periodic paralysis. *Neuromuscul Disord* 1999; 9: 227-231.
3. Cruz-Martinez A, Arpa J. Muscle fiber conduction velocity in hypokalemic periodic paralysis. *Acta Neurol Scand* 1997; 96: 229-235.