

High carbohydrate diet induced hypokalemic periodic paralysis: A case report.

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ABSTRACT

Hypokalemic periodic paralysis (HypoKPP) is a rare disorder characterised by the sudden onset of muscle weakness associated with low serum potassium levels. A 54-year-old man was admitted to the hospital with an abrupt onset of weakness in his right upper and lower extremities which slowly progressed to involve his left upper and lower extremities. He reported having consumed a carbohydrate-rich meal the day before the episode. Laboratory tests indicated severe hypokalemia with a serum potassium level of 2.3 mEq/L. He was treated with intravenous potassium chloride and his paralysis resolved after correction of hypokalemia. The patient was discharged on potassium supplementation and dietary guidelines were advised.

Keywords: carbohydrate-rich meal, hypokalemic periodic paralysis, potassium supplementation.

INTRODUCTION

Hypokalemic periodic paralysis is a rare genetic neuromuscular disorder characterized by transient episodes of flaccid muscle weakness. It is the most common type of periodic palsy. The HypoKPP is typically inherited or familial. The familial form is caused by mutation in either sodium or calcium ion channels in the skeletal muscle. Additionally, acquired cases of HypoKPP are found and linked to hyperthyroidism.¹ Here we present a case of HypoKPP who experienced sudden onset of muscle paralysis.

CASE REPORT

A 54-year-old male presented in the emergency department with sudden onset of weakness in his right upper and lower extremities for one day. The weakness slowly progressed to involve his left upper and lower extremities. He gave history of consuming carbohydrate rich meal a day before the episode. He had no history of shortness of breath, slurring of speech, blurred vision, or palpitation. He also reported similar episodes in the past about six years ago for which he was hospitalized. At that time, the patient received potassium supplementation and his weakness had resolved completely. He had a history of hypertension which was being treated with amlodipine. He had no significant surgical history. His family history was unremarkable.

On physical examination, his heart rate was 50 beats per min and blood pressure was 170/70mm of Hg. Cardiovascular examination revealed sinus bradycardia and no murmurs. Respiratory and abdominal examinations were unremarkable. Neurological examination revealed bilateral proximal and distal muscle weakness with motor power of 2/5 and decreased muscle tone. The sensation was intact but deep tendon reflexes were diminished in all extremities. Cranial nerve examination was intact.

Laboratory tests revealed severe hypokalemia with serum potassium level of 2.3 mEq/L. The renal function test, liver enzymes and complete blood count were within normal range. The serum magnesium level was 2.2 mg/dL (1.6 -2.4 mg/dL). The ECG revealed sinus bradycardia with prolonged QTc (495 ms). However, the cardiac enzymes were normal. MRI brain revealed old thalamic infarct and areas of microhemorrhages in left thalamus, left parietal subcortical white matter, right inferior cerebellar likely due to microangiopathy. The TSH level was normal (1.3 IU/ml).

His medical history and laboratory tests led to the final diagnosis of hypokalemic periodic paralysis. He was treated with IV potassium chloride. Twenty milliequivalents per liter of potassium chloride in 100ml of half normal saline was infused at a rate not more than 20 mEq/hr. A total of 120 mEq was administered over 24 hrs. The serum potassium levels were monitored 6 hourly. The next day acetazolamide 250 mg per oral twice a day was prescribed to the patient.

His MRI Spine showed discovertebral degenerative changes in cervical spine with mild spinal canal stenosis in C3-C4, C4-C5 and C5-C6 levels. Mild degenerative changes in the dorsal spine without spinal canal compromise. Degenerative changes in lumbar spine with spinal canal stenosis at D12-L1, L3-L4 levels.

The patient was discharged on oral potassium supplementation and antihypertensive. Dietary guidelines were advised. The patient was scheduled for a follow-up after 15 days, and then every four months thereafter. During his recent visit, he reported feeling well. He did not have any complaints of weakness in his arms or legs. He was reminded to avoid diets that are rich in carbohydrates.

DISCUSSION

Hypokalemic periodic paralysis (HypoKPP) is a form of periodic paralysis characterized by episodes of severe muscle weakness which are associated with low serum potassium levels.¹ It is usually triggered by strenuous activity or carbohydrate-rich meals. These factors cause an increase in epinephrine or insulin levels in the blood, which causes a shift of potassium into the cells.² Additional triggering factors include cold, stress, anesthesia, prolonged immobility, glucocorticoids, and alcohol.³

There are two types of HypoKPP. The hereditary type is a channelopathy caused by mutation in sodium or calcium ion channels in the skeletal muscle.¹ The calcium channel mutation is associated with a deletion in the gene for DHP-receptor α 1-subunit, CACNA1S. Muscle weakness is caused by an altered excitation-contraction coupling.⁴ The other type is caused by a mutation in the sodium channel gene, SCN4A. Acquired cases of HypoKPP are associated with hyperthyroidism.¹ The mechanism involves the activation of the sodium-potassium ATPase pump (Na/K - ATPase) by thyroid hormones, which causes an intracellular shift of potassium.⁵ Familial HypoKPP is an autosomal dominant disorder. The age of onset is in the first decade of life. The frequency of attacks tends to increase until 50 years, then it decreases in frequency.⁶ Thyrotoxic HypoKPP is more common among Asians with male predominance.⁵ The onset is usually after 20 years of age.¹

The attacks are episodic and manifest as flaccid muscle weakness occurring bilaterally, involving all four limbs but usually lower limbs are affected before the upper limbs. The proximal muscles are markedly involved compared to the distal muscles.³ The bulbar, ocular, and respiratory muscles are often unaffected but in severe attacks, involvement of respiratory muscle can prove fatal.^{5,6} Attacks develop over minutes to hours and may last several minutes to hours. They may resolve spontaneously.⁵ Neurologic examination reveals generalised muscle weakness with hyporeflexia or areflexia. Between the attacks, the neurological examination is usually normal. Myotonia is uncommon in HypoKPP.

The extent of hypokalemia varies during an episode. In one series, the mean blood potassium level was 2.4 mEq/L. The serum potassium levels remain normal during the attacks. Cardiac arrhythmias are uncommon during the attacks, but they can be induced by severe hypokalemia. Electrocardiogram abnormalities have been reported with Andersen-Tawil syndrome.⁵ It is a fatal condition that is characterized by periodic paralysis, cardiac abnormalities, and skeletal anomalies. Cardiac manifestations include prolonged QTc interval, premature ventricular contractions, prominent U waves, and ventricular tachycardia.^{7,8}

In most cases, the muscle strength is normal during the attacks and the frequency of attacks decreases in 3rd to 5th decades of life. However, few patients develop chronic interictal muscle weakness (permanent muscle weakness) manifesting as episodic paralytic attacks. Potassium therapy decreases the duration of paralytic attacks, but it doesn't have any effect on interictal weakness. Acetazolamide, a carbonic anhydrase inhibitor, reduces the frequency and severity of episodic attacks.^{3,9} Acetazolamide improves muscle strength probably by stabilising the muscle membrane.

The suspicion of HypoKPP arises when the patient presents with sudden onset of muscle weakness following strenuous exercise or carbohydrate rich meal or other above mentioned triggering factors and low potassium level as a laboratory finding. The diagnosis is supported by the history of previous similar attacks or positive family history. When there is established family history, no further testing is required to confirm the diagnosis of an acute attack, but other investigations are needed to rule out alternative diagnosis. To rule out hyperthyroidism thyroid function tests (T3, T4 and TSH level) are performed.^{1,8} Hypokalemia in thyrotoxic periodic paralysis is also associated with hypophosphatemia. Tachycardia is a useful clinical finding in this patients.¹⁰

Other diagnostic tests include genetic testing, provocative testing, and electromyography. Genetic testing is used to identify the underlying mutation.¹ Provocative testing includes administration of insulin or potassium and glucose. EMG may show reduced amplitude of compound motor action potential (CMAP) during the acute attack. The low potassium levels lead to hyperpolarization of muscle fibres. This results in decreased excitability of muscle fibres on stimulation of peripheral nerves and thus, decreased CMAP of the tested nerves. Between the attacks, EMG techniques can be used to confirm the diagnosis of periodic paralysis using exercise tests. It has high sensitivity following a recent muscle weakness and no treatment. It also has the advantage of diagnosing primary and secondary periodic paralysis. A muscle biopsy can show vacuolar changes which are non-specific and it is usually not performed for the diagnosis of periodic paralysis.⁷

The recommended treatment is administration of oral potassium chloride at a starting dose of 1mEq/kg. If there

is no improvement, then 0.3 mEq/kg is given every 30mins. The total dose of oral potassium should not exceed 200 mEq in 24hrs.³ The ECG, muscle strength and serum potassium levels should be monitored for 24hrs of starting treatment. Intravenous potassium is not preferred. It is only indicated in patients who develop arrhythmias or if the patient develops swallowing difficulties or airway compromise due to respiratory muscle paralysis. It is usually administered with mannitol and not with dextrose or saline, as they may both precipitate and trigger the attack.

Both non-pharmacological and pharmacological intervention can be used for prevention of further attacks. Non-pharmacological methods include avoiding carbohydrate rich diet and vigorous exercise. Pharmacological intervention includes chronic potassium supplementation, carbonic anhydrase inhibitors and potassium sparing diuretics. Acetazolamide and dichlorphenamide are carbonic anhydrase inhibitors being used for the empiric treatment of HypoKPP. They act by promoting potassium loss in urine and non-anion gap metabolic acidosis by increasing bicarbonate loss in urine. In addition, they may also be used to permanent muscle weakness by decreasing intracellular sodium accumulation.⁸

To conclude, early detection of HypoKPP with high index of suspicion is essential when a patient presents with sudden muscle weakness or paralysis to avoid life-threatening complications. Prompt treatment to correct low potassium levels can lead to a quick and complete resolution of symptoms.

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