Case Report

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Hypokalemic Peroidic Paralysis: The Unusual Ones in Pediatrics

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ABSTRACT

Hypokalaemic periodic paralysis is an uncommon but potentially life threatening clinical syndrome. It should consider in any child with recurrent motor paralysis. A positive family history with a low potassium level during attacks, will confirm the diagnosis If recognised and treated appropriately, patients recover without any clinical sequelae. No extensive investigations are needed. KCL, acetazolamide, spiranolactone and life style modifications- will help them to bring to normal activity.

KEYWORDS: Recurrent paralysis; Hypokalemia; Pediatrics; Motor weakness.

INTRODUCTION

Hypokalemic periodic paralysis (HPP) is a channelopathy caused by skeletal muscle ion channel mutation.[1] The genetic disorder is inherited as an autosomal dominant trait with incomplete penetrance in females. The patients present with sudden onset of generalized or focal flaccid paralysis associated with hypokalemia which persists for several hours before it resolves spontaneously.To avoid the morbidity associated with the condition it should be diagnosed and treated with prophylactic therapy. This condition is rare in pediatrics and that makes our topic interesting.

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CASE REPORT

A 12 yrs old boy admitted with history of weakness of both upper limb and lower limb of acute onset. He developed weakness in the early morning hours following calf muscle pain. Initially weakness developed in the lower limb which gradually affected upper limb also. Weakness resolved spontaneously with in 24 hours.

No history of altered sensorium or seizure. No history suggestive of any cranial nerve involvement. No history of abnormal sensation. No history of bladder and bowel symptoms.

He had similar episode first at 3.5 yrs of age and the second episode was at 11 years. After 11 years he had multiple episodes. For the past 1 year child is having weakness 1-2 episodes per month. There is history of similar illness in his mother's uncle and mother's brother.

On examination child is active, alert, vitals stable and neurological examination within normal limit except for grade 0-1 power in all limbs. Tone and reflexes were normal with a flexor plantar response.

At the time of weakness serum potassium was

2.7. Blood routine, RFT, LFT, sodium, calcium, CPK and blood gas analysis were normal. ECG shown flattening of T wave. Thyroid function tests were normal. Nerve conduction study done during weakness and found to have bilateral symmetrical axonal neuropathy. Considering the clinical pattern, family history and biochemical abnormalities, a diagnosis of hypokalemic periodic paralysis was made.

Child was treated with oral potassium chloride followed by acetazolamide. His weakness improved after potassium correction and potassium value became 4.1. Nerve conduction study was normal after correction of hypokalemia, when there were no weakness. He is now on follow up.

DISCUSSION

Hypokalaemic paralysis (HKPP) is an uncommon but potentially life-threatening clinical syndrome. If recognised and treated appropriately, patients recover without any clinical sequelae. It is associated with transient alterations in serum potassium levels, most often hypokalemia but also occasionally hyperkalemia.[1] Periodic paralysis may be primary (genetic) or secondary due to renal or gastrointestinal potassium loss as in gastroenteritis, renal tubular acidosis, or secondary to endocrine causes.[2]

Familial HPP is caused by a mutation in either calcium or sodium ion channel genes. The most common familial form, type 1 HPP, has a mutation in the dihydropyridine sensitive, skeletal muscle calcium channel gene, CACNA1S. Other familial form, type 2 HPP, has mutations in the voltage sensitive skeletal muscle sodium channel gene, SCN4A. Disease causing mutations in the gene KCNJ2 and KCNJ18, code for inward rectifier potassium (Kir) channel, have also been identified. Acquired HPP has been associated with thyrotoxicosis.[3,4]

The final common mechanism for all mutations is the formation of anomalous gating pore current through the voltage sensor domain of ion channel that makes sarcolemmal muscle inexcitable, resulting in failure of muscle action potential followed by subsequent attacks of flaccid paralysis. The anomalous gating pore current results in non selective cation leak causing aberrant depolarization, which is sufficient to make the resting potential of the muscle fibres unstable. When serum potassium level dips below 3.0 mmol/L, the affected fibres paradoxically undergo sustained depolarization making the muscle electrically inexcitable. In contrast, normal fibres undergo hyperpolarization when serum postassium drops. Normally inward rectifying potassium (Kir) channel and membrane Na-K-ATPase maintains the normal negative resting membrane potential. However in the presence of CACNA1S and SCN4A mutations, the depolarization induced by the gating pore current, at the modest drop of serum potassium level to around 3.0 mmol/L, counterbalance the Kir current leading to sustained depolarization.[5]

Late childhood or adolescence is the typical age of onset of hypokalemic periodic paralysis. It is precipitated in some children by a heavy carbohydrate meal, alcohol ingestion, insulin, epinephrine (including that induced by emotional stress), and cold exposure.[6] Before and during the attack, patients may have oliguria and excessive thirst. Weakness typically begins with an aching sensation in proximal muscles. Patients are unable to move after awakening, but gradually recover muscle strength. Lower limbs are involved more than upper lims. Diaphragm, extraocular muscles and cardiac muscle, are usually not affected. Patients are normal between the attacks, but as the child grows the attacks become more frequent, and progressive myopathy ensues with permanent weakness even in between attacks.

In the presence of an established family history of HKPP, further diagnostic investigations are not needed to confirm the diagnosis. In the absence of family history, a low serum potassium level during a typical attack of weakness establishes the diagnosis. Thyroid function test can be done to rule out hyperthyroidism. An electrocardiogram (ECG) may be taken to look for changes consistent with hypokalemia. A low serum potassium level between attacks usually represents a secondary cause of hypokalaemia.

During attacks of weakness, electromyography (EMG) may demonstrate reduced amplitude of compound muscle action potential (CAMP) and may show electrical silence, based on the degree of muscle weakness. In the long exercise test, an attack of focal muscle weakness is induced by vigorously exercising a single muscle for 2-5 minutes, and the change in postexercise CAMP in muscle fibers is measured by EMG. The reduction of 40% or more in CAMP is considered abnormal and typical for periodic paralysis.[6]

Muscle biopsy is not essential to diagnose periodic paralysis. Muscle biopsy are often normal between attacks, but during an attack a vacuolar myopathy is demonstrated.

Aim of the treatment is to normalize the serum potassium level by administering oral potassium chloride (KCL), which is more readily absorbed compared to other oral potassium solutions. Oral KCL is administered in increasing doses, starting with 0.5 to 1 mEq/kg. If there is no response to the initial dose, then 30% of the initial dose is repeated every 30 minutes.[6] The total dose of oral potassium should not exceed 200 mEq within the 24 hours. Continuous ECG monitoring is needed. Muscle strength should be examined.

Another drug used for HPKK is acetazolamide. It abolishes attacks in the majority of cases. The mechanism of action of acetazolamide is not fully understood, but it may block the flux of potassium from blood into the muscle.[7] The metabolic acidosis that it produces may contribute to its beneficial effect. In some cases it may lower serum potassium. To achieve adequate response, supplementation of potassium and avoidance of high carbohydrate meals are also needed. Chronic acetazolamide therapy may be associated with renal calculus, a complication which should be monitored for. In some patients, attacks may not respond to acetazolamide (or may even be exacerbated by it) in which case spironolactone may be affective.

Spironolactone, in a dose of 100-200 mg/day PO in school age children may be beneficial as well. FDA recently approved dichlorphenamide. It can be used as the first choice or as a substitute for patients who do not respond to acetazolamide.

IV potassium is not preferred initially and is reserved for severe manifestations like arrhythmias due to hypokalemia or if the patient has swallowing difficulties or respiratory muscle paralysis. IV potassium is preferentially administered with mannitol, not with dextrose or saline as both carbohydrate and salt can trigger muscle paralysis and may worsen the weakness.[8]

Most important aspect is education of parents-about triggering factors and lifestyle modifications. Avoidance severe exercise, heavy carbohydrate meals, alcohol, cold exposure and stress are important.

To conclude HKPP should consider in any child with recurrent motor paralysis. A positive family history with a low potassium level during attacks, will confirm the diagnosis. No extensive investigations are needed. KCL, acetazolamide, spiranolactone and life style modifications will help them to bring to normal activity.

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