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SPORADIC CASES OF NON-FAMILIAL HYPOKALEMIC PERIODIC PARALYSIS IN A SECONDARY HEALTH CARE FACILITY

General Medicine

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ABSTRACT

It is a heterogeneous group of disorders, which is characterized by sudden onset of reversible muscle paralysis1. The Primary PP (periodic paralysis) is due to inherited channel defects while Secondary PP is due to other medical conditions like thyrotoxicosis. HPP is the most common type of primary periodic paralysis and it usually resolves completely with k+ replenishment. This study presents 2 such cases, a 35y/male and 57y/F who presented with acute onset of flaccid paralysis and a low serum k+ level, which resolved completely with k+ administration.

KEYWORDS

Potassium, Glucose, Paralysis, Skeletal Muscle, ECG, channel

INTRODUCTION:

Hypokalemic Periodic Paralysis (HPP)[1] The most common form of periodic paralysis is reasonably rare with an approximately prevalence 1:100000. [2]HPP is a condition in which affected individual may experience the flaccid paralytic episodes with concomitant hypokalemia. The clinical features varies depending on the underlying etiology but most striking feature is sudden onset of weakness ranging in severity from mild, transient weakness to severe disability resulting in life threatening respiratory failure. A perturbation of Na+ and Ca+ channel resulting in low potassium levels and muscle dysfunction. [3]Although the serum k+ is alarmingly low, other electrolytes are usually normal with the change in the serum level reflecting a shift of potassium into cells.

[4]ECG changes are common. Most of the cases are characterized as familial or primary HPP. Whereas sporadic cases are associated with various other conditions like Barium poisoning, Hypothyroidism, renal disorder, endocrinopathies and gastrointestinal potassium losses. Respiration, deglutition and ocular motility are usually affected in severe attacks, furthermore studies shown the HPP does not affect heart but can trigger cardiac arrhythmias very rarely patient can develop permanent weakness, but this phenomenon is usually affects/seen in young individuals. [5,6]In Sporadic HPP, the triggering factors can be surgery, anesthesia, pregnancy, insulin, alcohol, strenuous exercise, and steroids. [7,8]The guidelines for care include control of plasma potassium, avoidance of large glucose and salt loads, maintenance of body temperature acid-base balance and careful usage of neuromuscular blocking agents. [9]Approximately 1/3rd cases show new mutations, treatment consists of diet changes andavoiding triggering factors.

[10]**Carbonic anhydrase (-)**: The Medications increase the flow of k+ commonly used drugs. dichlorphenamide and Acetazolamide,

[10]**Potassium supplements**: oral k+ supplements may be given to help stop an attack that is in progress.**CASE REPORTS:**

Case-1: A 57 years female with no significant history or family history came to the casualty with the complaint of acute onset of weakness of all 4 limbs progressing to complete quadriplegia over one night, on examination she had bilateral symmetrical flaccid quadriplegia with diminished reflexes and normal sensations , patient is not a known case of diabetes, hypertension or thyroid disorder, There was a one such episode in the past but was not educated about the disease and treated at local clinic. Investigation revealed hypothalamic → k+ 1.9 with all other electrolytes within normal limits. ABG finding was normal. Serum and urine calcium were normal, her thyroid profile was also normal, CT abdomen and pelvis, CT brain showed no finding, ECG revealed T inversion in lead V4 V5 V6, ST depression in lead V4 V5 V6. She was given supportive treatment along with intravenous potassium chloride correction. Her condition improved gradually, muscle power was regained, and reflexes become normal. She was followed up for 3 months and later lost in follow up.

Case-2: A 35 year male with no significant past or family history came to the casualty with the history of weakness over both the lower limb gradually progressed to left upper limb over a period of 6 hours, who does not have any comorbid condition, but history of intake of heavy carbohydrate meal, on examination, The power of both the lower limb 3/5 each and left upper isa 3/5 with diminished reflexes and intact sensation, with no significant systemic abnormalities on investigation his serum k+ - 2.4, Na+ - 141, Cl-102 with normal thyroid profile. LFT, RFT was found to be within normal limits, with no significant ECG changes. Ultrasound abdomen and CT brain shows no finding patient was treated symptomatically and educated about disease along with potassium correction and he is under the phase of follow-ups.

DISCUSSION:

[11]HPP is usually an autosomal dominant disorder affecting the ion channels of skeletal muscles**.** [12]HPP is most common with prevalence of 1:100000 results from abnormal k+ regulation due to Ca+ or Na+ channel abnormalities. [13,14,15]Mutation 7 CACNALS gene is responsible for 70% of cases and for rest is due to SCN4A gene mutation. In this gene mutation patient, channels have reduced excitability and signals from CNS are unable to depolarize the muscle, as a result the muscle cannot contract efficiently.HPP varies greatly in severity; there are two main subtypes i.e.,[1]Hypo-PP1 has a younger onset (10yrs or 6 yrs.) a longer duration of symptom (20hr v/s 1hr) and more commonly involves proximal muscles. Severe forms present at younger age(10yr) and mild forms can be triggered in predisposed individuals by a carbohydrate meal as in CASE-2 and strenuous exercise other triggering factors include stress, excitement, infection, high sodium intake, sudden temperature fluctuation. Drugs like insulin and β-blockers. However, some cases occur sporadically due to new mutations. HypoPP is an important differential to consider when seeing a patient with sudden onset weakness or paralysis especially when they are young with no significant associated risk factors or family history. This easily reversible condition can prove fatal if diagnosis is delayed which makes its diagnosis even more important. Any underlying conditions are treated if present in all cases, in most cases clinical diagnosis made by measuring serum k+ level. However, the standard test is a specialized form of Electromyography (EMG) testing called long exercise test, which measures (CMAP) compound muscle action potential for 40-50 min following a few minutes of exercises.

In affected individuals there is progressive fall in the CMAP, serum concentration of TSH and FT3, FT4 may distinguish between primary Hypo PP of Thyrotoxic Periodic Paralysis (TPP), Urine Na+ and K+ levels may help exclude hyperaldosteronism. The Acute management involves potassium replenishment and need of assisted ventilation when respiratory muscles are involved. The best route for k+ replacement is oral, however parental administration can be recommended if the patient cannot swallow the recommended dose, for a 60kg patient the dose is 30-60mEq (i.e.,0.5-1mEq 1kg). Recently the FDA approved dichlorphenamide (Keveyis) a carbonic anhydrase inhibitor for the long-term treatment of Hypo PP. However, the exact mechanism by which dichlorphenamide prevents recurrence is unknown. Recent data suggest that Carbonic anhydrase inhibitor activates skeletal muscles BK channel (Ca+ activated K+ channel), In long term Carbonic anhydrase inhibitor therapy, oral K+ supplements may be required. Most of the patients recover though they may have

recurrent episode in the future, only a small minority may develop progressive muscle weakness that may become permanent.

CONCLUSION:

HPP is a rare genetic disease of skeletal muscle. High index of suspicion is necessary for diagnosing this otherwise rare entity. Especially when encountering a patient with an acute attack of paralysis accompanied by documented hypokalemia and no other explanation could be found. Failure to properly diagnose and treat HPP can be fatal, but rapid corrections of potassium resolve the symptoms quickly and completely. Physicians should have appropriate preparedness to deal with these diseases.

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