Imperial Journal of Interdisciplinary Research (IJIR)
Vol-2, Issue-5, 2016
ISSN: 2454-1362, http://www.onlinejournal.in

Hypokalemic periodic paralysis: A case report.

Hafez Mohammad Ammar Abdullah[1], Muhammad Asim Shahzad[1], Syed Shahmeer Raza [1], Amer Kamal Hussain[2] & Muhammad Daniyal Nadeem[2]

[1]Dept. of Medicine, Khyber Teaching Hospital, [2]Khyber Medical College, Peshawar

Abstract*: A rare heterogeneous group of disorders which is characterized by a sudden onset of reversible muscle paralysis. The Primary PP is due to inherited ion channel defects while secondary PP is due to other medical conditions like thyrotoxicosis. HPP is the most common primary Periodic paralysis and it usually resolves completely with K replenishment. This study presents the case of a 30 year old male who presented with an acute onset of flaccid paralysis and a low serum K level, which resolved with K repletion.*

Introduction.

A rare heterogeneous group of disorders which is characterized by a sudden onset of reversible muscle paralysis. A 30 years old male patient presented to the emergency department at 10am in the morning with an acute history of paralysis. The patient was ok the previous day but had some weakness in the lower limbs the previous night. However in the morning when the patient woke up, he was unable to move at all. The weakness was more prominent in the lower limbs and it was more severe in the proximal muscles of both the lower and upper limbs. ECG showed a normal rate and rhythm but there was flattening of the T waves. The patient was diagnosed with hypokalemic periodic paralysis, probably precipitated by an acute chest infection. Periodic paralysis (PP) is a rare but often overlooked cause of acute flaccid paralysis. It may be primary or secondary periodic paralysis. In most cases a clinical diagnosis can be made after measuring the serum K levels. During an attack, the serum conc. of K ranges from 0.9 to 3.0 mmol/L (normal range: 3.5-5.0 mmol/L). However the standard test is a specialized form of electromyography (EMG) testing called the long exercise test. The paralysis resolved completely following k replenishment . At the time of discharge, he had no neurologic findings and was vitally stable.

CASE REPORT

A 30 years old male patient presented to the emergency department at 10am in the morning with an acute history of paralysis. The patient was ok the previous day but had some weakness in the lower limbs the previous night. However in the morning when the patient woke up, he was unable to move at all. The weakness was more prominent in the lower limbs and it was more severe in the proximal muscles of both the lower and upper limbs. He had no difficulty in breathing and there was no problem in swallowing. The patient could also move his facial and neck muscles. The patient was also having a cough ,a sore throat and intermittent fever for the last 3 days. There was no history of smoking. There was no history of any recent GI infection. On systemic review there was no history of any ear nose eye symptoms. There was no history of palpitations, excess sweating, GI discomfort, dysuria, urgency or frequency. The patient had no serious past medical or surgical history and no history of any previous instance of hospital admission. The patient was not on any long-term medication. There was no history of any such previous episode in the patient or any of his family members. The patient had no recent travel history. He worked as a farmer.

Upon **physical exam,** the patient was stable vitally stable. His BP was 110/70 mmhg, pulse was72 BPM, Temp. was 99F and respiratory rate was 20. On general inspection the patient was not in any obvious discomfort. He was warm on touch. There were no obvious signs of dehydration. There was no jaundice, lymphadenopathy, thyroid enlargement or clubbing. The patient was not apparently anemic. His throat was congested and tonsils were inflamed.

on neurological exam the patient was oriented in time place and person. the power in the lower and upper limbs was 2/5.The weakness was more obvious in the proximal muscles. The tone was normal. The deep tendon reflexes were also normal. there was no sensory loss of any modality. There were no fasciculation or obvious muscle atrophy. There were no upper motor neuron signs.

On chest exam the patient had no obvious chest deformity. He had normal tympanic notes on percussion, and bilateral crackles and wheezes, more prominent in the lower lung fields. The cardiac and abdominal exams were unremarkable.

Routine **Labs** showed an Hb of 12g/dl, TLC of 10,800 and Platelet count of 17700. the Serum electrolytes at the time of admission were Na 146.9 , K 1.81, Cl 104 meq. The serum Creatinine, urea,

total bilirubin , AST and alkaline phosphatase were in the normal range. The serum ALT was 69 IU. The ECG showed a normal rate and rhythm but there was flattening of the T waves.

The patient was diagnosed with hypokalemic periodic paralysis, probably precipitated by an acute chest infection. The patient was started on IV KCl in a 1L infusion of normal saline. the patient responded quickly and the paralysis disappeared within an hour the patient was started on oral antibiotics for his throat infection. he was also given symptomatic treatment for his fever and sore throat. the repeat Serum electrolytes showed a K of 2.8. follow up investigations were performed to find any secondary cause. the thyroid function test showed T4-7.10 ng, T3-0.7ng and Tsh-0.2 IU and the patient was clinically Euthyroid.

The patient was educated about his condition. He was counseled about the precipitants to avoid. He was discharged on oral antibiotics for 7 days and oral K supplements for 3days.At the time of discharge his K was 4.7 meq.

DISCUSSION.

Periodic paralysis (PP) is a rare but often overlooked cause of acute flaccid paralysis. It may be primary or secondary periodic paralysis. PP may be hypokalemic or hyperkalemic. Hypo. PP is the most common Primary PP. It is characterized by episodes of paralysis that begin to occur in the teens or 20s. Thyrotoxic PP is the most common cause overall and occurs in approximately 1-3% of Thyrotoxic patients.

Hypokalemic Periodic Paralysis

The prevalence of hypokalemic periodic paralysis (HypoPP) is approximately 1 in 100,000 with an estimated 5000 cases in the united states.(1)The worldwide prevalence may vary and is not known.HypoPP varies greatly in severity. There are 2 main subtypes. HypoPP1 has a younger onset (10 yrs vs 16yrs), a longer duration of symptoms (20hr vs 1hr) and more commonly involves the proximal muscles. Severe forms may first present at a younger age (10yrs) while milder cases can be asymptomatic until the thirties. HypoPP can be triggered in predisposed individuals by a carbohydrate meal and after strenuous exercise. Other triggers include stress, excitement, infections, a high sodium intake, sudden temperature changes and drugs like insulin and beta blockers.

Primary HypoPP is caused by mutations in genes coding for ion channels and are inherited in an autosomal dominant fashion with reduced prevalence in females. (2) However some cases occur sporadically due to new mutations. The most common ones described include CACNA1S which encodes a voltage-gated Ca-channel Cav1.1 found in the transverse tubules of skeletal muscle cells (HypoPP type1) and SCN4A a voltage-gated Na channel Nav1.4 found at the neuromuscular junction (HypoPP type2). Another mutation KCNE3 which encodes a voltage-gated K channel has also been described, but is currently disputed. These mutations are loss-of-function, such that the channels cannot open normally however, the mechanism linking these defect to paralysis and hypokalemic remains obscure.(3)The gene defects described don't occur in 100% of the cases. It has been proposed that some are yet undescribed new mutations may underlie those cases that don't carry the traditional mutations.(4)

HypoPP is an important differential to consider when seeing a patient with sudden onset weakness or paralysis, especially if they are young and have no history or evidence of other diseases and no significant risk factors for stroke. This easily reversible condition can prove fatal if diagnosis is delayed , which makes its diagnosis even more important. Any underlying condition should be actively sought after and treated if present in all cases.

Diagnosis.

In most cases a clinical diagnosis can be made after measuring the serum K levels. During an attack, the serum conc. of K ranges from 0.9 to 3.0 mmol/L (normal range: 3.5-5.0 mmol/L).However the standard test is a specialized form of electromyography (EMG) testing called the long exercise test. This test measures the Compound Muscle Action Potential (CMAP) for 40 to 50 minutes following a few minutes of exercise. In affected patients, there is a progressive fall in the CMAP. Serum concentration of TSH and free T3 and T4 may distinguish between primary HypoPP and Thyrotoxic periodic paralysis (TPP). Urine Na and K levels may help exclude hyperaldosteronism.

Management.

The acute management is focused more on the replenishment of K and assessing the need for assisted ventilation if respiratory muscles are involved. The best route for K replacement is oral with KCL. However if the patient cannot swallow, then parenteral administration is recommended. The

recommended dose for a 60kg patient is (ie, 0.5-1 mEq/kg) is 30- 60 mEq.

Recently the FDA approved Dichlorphenamide (Keveyis) a carbonic anhydrase inhibitor for the long-term treatment of HypoPP. However the exact mechanism by which dichlorphenamide is able to prevent recurrence is unknown. Recent data suggest carbonic anhydrase inhibitors activate skeletal muscle BK channel (Ca2+ -activated K channel)(5). In long-term carbonic anhydrase therapy, oral K supplements may be required.

Most of the patients recover though they may have recurrent episodes in the future. however deaths have been reported due to HypoPP. Only a small minority may develop progressive muscle weakness that may be permanent.

CONCLUSION.

This patient described in this report presented with sudden onset paralysis and a reduced serum potassium level. All other labs including TSH, T3 and T4 levels were normal. He was also suffering from a chest infection and pharyngitis which was the most probable precipitant. It was his first episode. The paralysis resolved completely following k replenishment . At the time of discharge, he had no neurologic findings and was vitally stable.

References.

1.Fontaine B. Primary periodic paralysis and muscle sodium channel. Adv Nephrol Necker Hosp 1994; 23: 191–7. PMID 8770943

2.Ke Q, Luo B, Qi M, et al. Gender differences in penetrance and phenotype in hypokalemic periodic paralysis. Muscle Nerve 2013; 47:4 PMID 23019082

3.Jurkat-Rott K, Lehmann-Horn F . "Muscle channelopathies and critical points in functional and genetic studies". J. Clin. Invest. 115 (8): 2000–9. doi:10.1172/JCI25525. PMC 1180551. PMID 16075040.

4.Venance SL, Cannon SC, Fialho D, et al. The primary periodic paralyses: diagnosis, pathogenesis and treatment. Brain 2006; 129:8. PMID 16195244

5.Tricarico D, Barbieri M, Mele A, et al. Carbonic anhydrase inhibitors are specific openers of skeletal muscle BK channelof K+-deficient rats. FASEB J. 2004 Apr. 18(6):760-1

Fig. 1 showing flattening of T waves in ECG.