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Hypokalemic Periodic Paralysis: A case Report and Review of the Literature

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Abstract:

Hypokalemic periodic paralysis (hypoPP) is a disorder that causes occasional episodes of muscle weakness and sometimes a lower than normal level of potassium in the blood during the attack. HypoPP is one of a group of genetic disorders that includes hyperkalemic periodic paralysis and thyrotoxic periodic paralysis among others.

A 45 year old male presented with quadriplegia for 1 day of sudden onset following heavy exercise one day before the attack, Laboratory evaluation revealed a markedly low potassium level of 1.7 meq/L, normal thyroid function test , and normal magnesium level .

The patient's paralysis resolved during replacement of his low potassium within 48 hours of ICU admission and he was discharged with no neurologic deficits.

He has a family history of similar condition (1st and 2nd degree relatives).

The case report:

A 45 year-old African male with significant past medical history and family history of similar condition presented to the A/E department with sudden onset quadriplegia and shortness of breath.

The patient awoke up at early morning with muscles cramps and inability to move his upper or lower extremities. The weakness was bilateral and involved both the proximal muscles of the shoulders and hips as well as the distal muscles ofextremities. He had respiratory and swallowing difficulty and was able to move his neck and facial muscles.He denied history of trauma .This attack was preceded by history of strenuous exercise one day before. The patient denied any history of vomiting,diarrhea,fever , changing in diet .No history of trauma, He did not take any medications and denied use of alcohol or drugs.His daughter, cousin and his aunt had been exposed to similar attack and diagnosed as familiar HypoPP , based on clinical background and low potassium level .

On physical exam, he was slightly over weight, but otherwise normal in overall appearance. The patient's heart rate was 100 beat per minute , and blood pressure was

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110/80, RR 27 breath per minute ,he was not pale , jaundiced or cyanosed.There were no deformities or edema of the extremities and distal pulses were present and equal bilaterally.No jugular venous distension, goitre or lymphadenopathy.Cardiac exam revealed normal 1st and 2nd heart sounds and no murmurs. Examination of the lungs and abdomen were unremarkable.

Neurologic examination revealed hypotonia and hyporeflexia of all extremities which involved the proximal and distal muscles; power grade 2, sensation were intact , coordination difficult to assess .Back examination was normal .

Routine chemistry, liver enzymes , renal functions and complete blood count were normal except for a potassium level of 1.7meq/l, TFT normal. Electrocardiogram revealed sinus tachycardia with 1st degree atrio-ventricular block, and prominent U wave .

The patient was diagnosed as a case of familial hypokalemic periodic paralysis ,and was started on oral potassium supplement and potassium sparing diuretics .He was discharged home in good condition after 2days admission in ICU and 4 days in general ward .

Discussion:

Limb paralysis is a common presentation in both the emergency and outpatient settings. The most common causes of paralysis in ER are : hemiplegia ,cord compression ,Potts disease,GBS .Other causes are rare,e.g.HypoPP so we reported it. Diagnosis of these disorders requires obtaining a complete history with special consideration of timing, duration, and distribution of symptoms.

There are several types of Periodic Paralysis associated with metabolic and electrolyte abnormalities. Of these, hypokalemic periodic paralysis (HypoPP) is the most common with a prevalence of 1 in 100,000 [(1)] .The clinical features of the syndrome vary somewhat depending on the underlying etiology but the most striking feature is the sudden onset of weakness ranging in severity from mild , transient weakness to severe disability resulting in life threatening respiratory failure.Attacks may be provoked by strenuous exercise, stress such as a viral illness or fatigue, or certain medications such as beta-agonists, insulin or steroids, or carbonated drinks. A perturbation of sodium and calcium ion channels result in low potassium levels and muscle dysfunction [(2)] . As this is primarily a problem with muscle contraction rather than nerve conduction, tendon reflexes may be decreased or absent but sensation is generally intact.Although the serum potassium level is often alarmingly low, other electrolytes are usually normal. Indeed, total body potassium is actually normal with the change in the serum level reflecting a shift of potassium into cells [(3)].Electrocardiographic changes are common, but unlike patients who are truly potassium depleted , the changes do not correlate well with the measured serum level [(4)]. Identification of the problem between paralytic episodes is difficult as the patient may have normal strength and potassium levels. Electromyography reveals abnormalities in some patients but is often normal, especially between episodes when no clinically detectable weakness is present.EMG

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during weakness typically reveals myopathic changes with reduced amplitude of compound muscle action potential.HypoPP occurs in several settings and the diagnosis may require an extensive search for the underlying etiology since the treatment varies according to the cause. HyoPP may occur sporadically in the form of familial hypokalemic psaralysis (FHP), a poorly understood disorder which may occur spontaneously or as the result of autosomal dominant inheritance [(1)].This form of Periodic Paralysis (autosomal dominant channellopathy)is due to disordered cellular potassium regulation either due to sodium or calcium channel abnormalities [(2,5)].Mutations of the CACNA1S and SCN4Agenes have been identified and cause abnormalities in sodium channels resulting in abnormal potassium ion flux. Acute paralytic episodes are treated with potassium replacement and close monitoring of the cardiac rhythm and serum potassium levels. Spironolactone and acetazolamide have been used for prophylaxis with some success although long-term potassium supplementation may be necessary [(2)].

Conclusion:

Hypokalemic periodic paralysis is an autosomal dominant disorder which runs through families with occasional episodes of muscle weakness. Oral and intravenous potassium supplementation are usually given and progressive improvement in weakness is suspected. Missing diagnosis of HypoPP may lead to fatal outcomes like respiratory failure and cardiac arrhythmias.

Consent:

Consent was obtained from the patient for publication of this case report .

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