RECURRENT HYPOKALEMIC PERIODIC PARALYSIS –THREE RARE SECONDARY CAUSES

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ABSTRACT

Periodic paralysis (PP) is a rare neuromuscular disorder related to a defect in muscle ion channels, characterized by episodes of painless muscle weakness, which may be precipitated by heavy exercise, fasting or high-carbohydrate meals. Hypokalemic PP is still quite rare with an estimated prevalence of 1 in 100,000. Here we present a case of 41 years old female patient having hypokalemic periodic paralysis with secondary causes including renal tubular acidosis (RTA), Sjogren’s syndrome and hypothyroidism.

KEY WORDS: Hypokalemic Periodic Paralysis, Renal tubular acidosis, Sjogren’s syndrome, Rheumatoid Arthritis.

INTRODUCTION

Hypokalemic periodic paralysis (HypoKPP) is a rare neuromuscular disorder characterized by transient episodes of flaccid muscle weakness and especially- respiratory failure and death. Hypokalemic PP is the most common of the periodic paralyses, it may be familial (primary) or secondary (acquired).¹¹ Hypokalemic periodic paralysis as well as thyrotoxicosis episodes occur in association with low potassium blood levels, concentrations less than 2 mEq/L suggest a secondary cause of hypokalemic paralysis such as Renal Tubular Acidosis.²²

Most cases of PP are hereditary, usually with an autosomal dominant inheritance pattern. The defect is mediated through mutation in the voltage-gated calcium or sodium ion channels,
which further results in abnormal sarcolemma excitation.\textsuperscript{[3]} Recurrent hypokalemic paralysis is an extremely rare presentation of hypothyroidism.\textsuperscript{[4]}

**CASE REPORT**

A 41yrs old female with hypothyroidism for past 4 years and rheumatoid arthritis presented with severe myalgia since 1 week and sudden onset of weakness of both limbs of 1 day duration. Vitals were stable, patient was conscious and oriented, power-2/5(both upper and lower limbs) and plantar-B/L equivocal. No history of neurological and renal disorders. She was on regular medications Tab.levothyroxine 75mcgOD and Tab.Hydroxychloroquine 200mg OD. She had a history of similar episode 4 months back.

Investigations revealed hypokalemia (K+ levels were 1.7-3.1mEq/L), metabolic acidosis (urine pH -7.0) and Anion gap of (17.8 mmol/L) all together suggestive of probable distal renal tubular acidosis. On investigation she also had elevated erythrocyte sedimentation rate of 120 mm/hr. Antinuclear antibody profile showed strong positivity (ANA-1.8) for Sjogren’s syndrome with RA.

She was treated with intravenous potassium replacements as infusion under observation followed by oral correction, oral sodium bicarbonate 500mg BD,Tab.Hydroxychloroquine 200mg OD and Tab.Prednisolone 5mg BD. Paralysis symptoms improved within 2 days and patient was better within 6-7 days of therapy. She is on regular follow up with rheumatologist.

**Table: 1 Lab Investigation reports of the patient.**

<table>
<thead>
<tr>
<th>Lab Investigations</th>
<th>Patient Value (Normal Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium</td>
<td>146mmol/L (135-145mmol/L)</td>
</tr>
<tr>
<td>Bicarbonate</td>
<td>11.2(10-29mmol/L)</td>
</tr>
<tr>
<td>Potassium(Admission)</td>
<td>1.7mmol/L(3.5-4.5mmol/L)</td>
</tr>
<tr>
<td>Potassium(Discharge)</td>
<td>4.5mmol/L(3.5-4.5mmol/L)</td>
</tr>
<tr>
<td>Creatinine Phosphokinase</td>
<td>176 (25-170 IU/L)</td>
</tr>
<tr>
<td>ESR</td>
<td>120 (0-20mm/hr)</td>
</tr>
<tr>
<td>Anion Gap</td>
<td>17.8 (7-16mmol/L)</td>
</tr>
<tr>
<td>ANA</td>
<td>1.8(1.4-1.6)</td>
</tr>
<tr>
<td>Urine Potassium</td>
<td>23.6 (25-125mEq/L)</td>
</tr>
<tr>
<td>Urine pH</td>
<td>7(4.6-8)</td>
</tr>
</tbody>
</table>

**DISCUSSION**

Hypokalemic PP may be familial (primary) with autosomal dominant inheritance or may be acquired (secondary) in patients with thyrotoxicosis. Disturbances of potassium equilibrium
can produce a wide range of disorders including myopathy, marked muscle wasting, diminution of muscle tone, power, and reflexes.\cite{4} Clinical presentation is often incomplete, especially in women. Primary hypokalemic periodic paralysis is believed to be due to an increase in muscle membrane potassium permeability.\cite{5} Hypokalemia is the consequence of a rapid and massive shift of potassium from the extracellular to the intracellular compartment, mainly into the muscles. This is believed to be related to increased sodium-potassium-adenosine triphosphatase (Na/K-ATPase) pump activity.\cite{6} Whether by potassium movement into cells or by potassium loss, the resulting hypokalemia reduces the resting membrane potential and blocks the action potential.\cite{1}

These attacks vary in frequency and duration. Intervals of weeks to months are common, but some patients experience several attacks per week. Attacks typically last for several hours, but it can range from minutes to days. It may be triggered by rest after vigorous exercise, stressor a high-carbohydrate meal, often after a delay of several hours. These events are often associated with an increased release of epinephrine or insulin both of which cause movement of potassium into cells and low potassium blood levels. Certain medications like beta-agonists and corticosteroids have also been incriminated as triggers.\cite{5}

ECG may show signs of hypokalemia, such as ST depression, decrease of the amplitude of the T-wave and an increase of the amplitude of the U-waves; but arrhythmias such as atrial fibrillation, supraventricular paroxysmal tachycardia, or ventricular fibrillation are not common.\cite{1}

Distal Renal Tubular Acidosis requires the administration of alkaline salts. In addition to potassium correction, Sodium bicarbonate should be administered to satisfy the alkali requirements and compensate the bicarbonate loss.\cite{1} Prophylaxis against recurrent periodic attacks has been successful with a wide variety of treatment modalities including spironolactone and acetazolamide.\cite{5}

**CONCLUSION**

It is an extremely rare and unusual presentation of hypothyroidism, renal tubular acidosis and Sjogren’s syndrome along with periodic hypokalemic paralysis, which shows a prevalence of 1 in 100,000. Each of the above conditions along with hypokalemic periodic paralysis are rare but completely treatable if diagnosed properly on time. Failure to diagnose properly and treat Periodic Paralysis can be fatal, rapidly correcting the potassium can resolve the
symptoms completely. Here in this case, all the secondary conditions along with periodic paralysis were treated accordingly and prevented further attacks.

ACKNOWLEDGMENT
The authors would like to thank the managements of Lourdes hospital and St. Joseph’s College of Pharmacy for their support and encouragement.

AUTHORS’ CONTRIBUTIONS
Dr. Sunu Kurian treating doctor provided the clinical details of the case and reviewed the manuscript, Ms. Anjaly P Nair involved in the preparation, reviewing and editing of the manuscript and Dr. Siby Joseph and Ms. Lakshmi R were involved in the organizing, reviewing and editing of the manuscript.

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