

## Case Reports

# Hypokalaemic Periodic Paralysis Associated with Distal Renal Tubular Acidosis

Sujeet Raina\*, Abhishek Goyal+, Satinder Singh Kaushal\*\*, Dalip Gupta\*\*\*

### Abstract

A patient presented with acute flaccid quadriplegia which on evaluation was found to be due to hypokalaemia. Further investigation revealed, metabolic acidosis with normal anion gap. Alkaline urine, transtubular potassium concentration gradient more than four. A diagnosis of distal renal tubular acidosis presenting as hypokalaemic periodic paralysis was made.

### Introduction

The purpose of this case report is to emphasize that distal renal tubular acidosis may be the cause of acute flaccid quadriplegia. The usual causes of HPP include familial, hyperaldosteronism, gastrointestinal losses, barium poisoning, dRTA and thyrotoxicosis.<sup>1</sup> Distal RTA is a non-uraemic syndrome of defective urinary acidification. It is characterized by presence of hypokalaemia hyperchloraemic metabolic acidosis with normal anion gap, inability to lower urinary pH < 5.5, nephrocalcinosis and features of osteomalacia or rickets.<sup>2</sup>

### Case Report

A 26 year old female housewife was admitted with clinical finding of acute flaccid paralysis of two days duration. She had experienced similar episode two year back which recovered with symptomatic treatment. There was no history of similar episodes of weakness in any of family members. There was no history of drug intake. At the time of admission general physical examination revealed bradycardia with pulse rate of 48/minute. There was no clinical evidence of thyrotoxicosis. On examination of nervous

system higher mental functions, cranial nerves, speech was normal. Motor examination revealed normal muscle bulk with hypotonia. She had grade zero power and all the deep tendon reflexes were absent. Plantars were bilaterally flexor. There were no signs of sensory and autonomic dysfunction. Investigation revealed normal haemogram, random blood sugar and liver function tests. Serum urea and creatinine was 40 mg/dl and 0.8 mg/dl respectively. Serum electrolytes revealed Sodium (Na<sup>+</sup>) as 144 meq/l, potassium (K<sup>+</sup>) as 1.5 meq/l and chloride (Cl) as 109 meq/l. Arterial blood gas study at admission revealed pH as 7.29, bicarbonate was 11.6 mmol/l, PaCO<sub>2</sub> was 30 mm Hg and PaO<sub>2</sub> was 95 mmHg. Anion gap was normal with the value of 9. The 24 hour urine potassium was 38.9 meq and urine osmolality was 779.4. Urine was alkaline with the pH of 7.0 (fresh morning sample). Serum osmolality was 308.4. Transtubular potassium concentration gradient (TTKG) was 6.15. Thyroid profile was normal. ECG at admission revealed bradycardia, increased PR interval, diminished T wave amplitude and U waves. Ultrasound abdomen did not reveal nephrocalcinosis. The diagnosis of distal RTA was made by the findings of systemic acidosis, low bicarbonate, hypokalaemia, a normal anion gap and alkaline urine despite the acidaemia.

Patient was treated with intravenous (IV) potassium infusion. The weakness improved drastically over next 24 hours. Patient was started on Shohl's solution in a dose of 1 mmol/kg/day in divided doses. At discharge serum potassium was 3.82 meq/l, chloride 102 meq/l, pH 7.41 and bicarbonate 23.5 mmol/l. The electrocardiogram at discharge was

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\*Senior Resident; +Postgraduate Student; \*\*Professor and Head; \*\*\*Associate Professor, Department of Medicine, Indira Gandhi Medical College, Shimla – 171002 (H.P).

normal. The patient is being followed up and remains asymptomatic.

## Discussion

Acute HPP is a relatively uncommon, potentially fatal, if untreated and completely reversible clinical condition. Ascending paralysis is likely to occur at serum potassium level of 2-2.5 meq/l. Besides potassium replacement therapy the approach to a patient with HPP includes a thorough evaluation to find out the underlying cause of hypokalaemia. The usual causes of HPP includes familial periodic paralysis, thyrotoxicosis, distal RTA, hyperaldosteronism, gastrointestinal loss and barium poisoning.<sup>1</sup> Gastrointestinal losses and barium poisoning can be ruled out on the basis of history. Hyperaldosteronism is suspected in the presence of hypertension, hypernatraemia and metabolic alkalosis. Thyrotoxic periodic paralysis has signs and symptoms of hyperthyroidism and deranged thyroid function test. In general when hypokalaemia is present in combination with hyperchloraemic acidosis with normal anion gap ( $\text{Na}^+ - [\text{Cl}^- + \text{HCO}_3^-] = 8-16 \text{ mmol/l}$ ) in a patient without evidence of gastrointestinal  $\text{HCO}_3^-$  losses and who is not taking acetazolamide or ingesting exogenous acid, distal RTA must be suspected.<sup>3</sup> Distal RTA is a clinical syndrome consisting of a hypokalaemia, hyperchloraemic metabolic acidosis with normal anion gap, inability to lower urinary pH below 5.5, nephrocalcinosis, osteomalacia or rickets. The basic defect is impairment of H-K ATPase proton pump; failure of this pump leads to defect in acidification and urinary potassium loss with all the features of distal RTA.<sup>2</sup> Primary distal RTA is inherited in an autosomal dominant fashion but most cases are sporadic. Rarely distal RTA occurs in autosomal recessive forms. Secondary causes

of distal RTA are Sjogren's syndrome, use of Amphotericin B, and certain blood disorders like sickle cell anaemia.<sup>4</sup> Treatment of distal RTA requires alkali administration (equivalent to the sum of endogenous acid production and amount of accompanying bicarbonate wastage) in the form of Shohl's solution, which is a combination of sodium citrate and citric acid. The usual dose of Shohl's solution is 0.5-2.0 mmol/kg bodyweight in 4-6 divided doses per day.<sup>5</sup> It is essential to differentiate between HPP due to RTA and FPP as the emergency treatment and prophylaxis differ. Bicarbonate is contraindicated in FPP as it facilitates intracellular potassium influx and fatal hypokalaemia may result. In FPP no applicable drug affords constant prophylaxis against muscle paralysis. The best combination is of acetazolamide and potassium. While acetazolamide is strongly contraindicated in prophylaxis of distal RTA because it can increase the metabolic acidosis.<sup>6</sup>

## References

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