Case Report
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Hypokalemic quadriparesis and rhabdomyolysis as a rare presentation of distal renal tubular acidosis

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Abstract

Distal renal tubular acidosis is a syndrome of abnormal urine acidification and is characterized by hyperchloremic metabolic acidosis, hypokalemia, hypercalciurea, nephrocalcinosis and nephrolithiasis. Despite the presence of persistent hypokalemia, acute muscular paralysis is rarely encountered in males. Here, we will report an eighteen year old male patient who presented with flaccid quadriparesis and was subsequently found to have rhabdomyolysis, severe short stature, skeletal deformities and primary distal renal tubular acidosis.

Keywords: Nephrocalcinosis, Hypokalemia, Rhabdomyolysis, Short stature, Renal tubular acidosis.

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Introduction

Renal tubular acidosis, first described in 1935, is a broad term applied to various transport defects in kidney. It includes two major forms, failure to absorb bicarbonate in proximal tubule (proximal renal tubular acidosis or pRTA) or inability to secrete hydrogen ion in distal tubule (distal renal tubular acidosis, dRTA) (1). Distal renal tubular acidosis was recognized as a distinct entity by Albright et al, the clinical syndrome consisting of hypokalemia, hyperchloremic metabolic acidosis, inability to lower urine pH below 5.5, nephrocalcinosis and nephrolithiasis (2). Etiology of distal renal tubular acidosis is diverse ranging from genetic causes which can manifest as autosomal dominant or autosomal recessive (4) to autoimmune disorders like autoimmune thyroid disease, Sjogren's syndrome and chronic active hepatitis (5,6). Clinical presentation of the disease may be variable depending upon the age and underlying cause. In neonates, the disease can manifest as constipation, polyurea and polydipsia while in children prominent clinical features include impairment of growth, polyuria, hypercalciuria, nephrocalcinosis and nephrolithiasis. In adults, symptoms associated with underlying cause may predominate.

Case Report

An eighteen year old boy, born out of non-consangious marriage, first in birth order, born at term by vaginal delivery with normal perinatal history and normal motor and mental mile stones, was admitted in

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Fig.1. Severe short stature

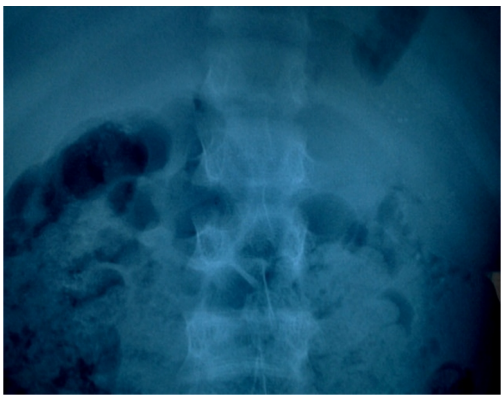
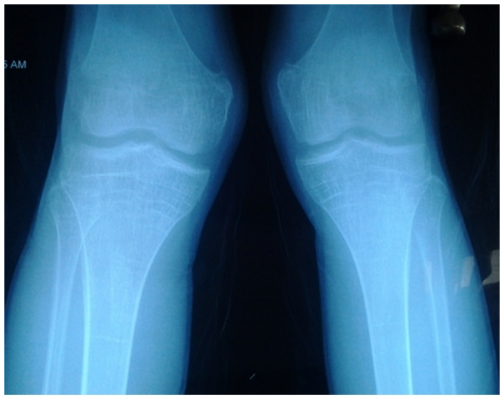


Fig.2. Fine calcific foci bilaterally in renal area (black Arrow)



accident and emergency department with history of progressively worsening crampy leg pain from one month, increased frequency of micturation and increased thirst for last three weeks. There was history of inability to move his limbs from last 3 days. There was no history of rash, headache, fever, vomiting, diarrhea, and weight loss and bladder or bowel incontinence. There was no history of any drug or herbal medicine intake. On examination, patient was conscious, cooperative and well oriented in time, place and person. General physical examination revealed pulse 92 per minute, blood pressure 110/70mmHg and respiratory rate of 16 per minute. There were bony deformities in the form of widening of bilateral wrists and bilateral knock-knees. Chest, cardiovascular and abdominal examinations were unremarkable. Nervous system examination revealed normal higher mental functions and no cranial nerve deficit. Motor examination revealed hypotonia and a power grade of 3/5 in both arms and 2/5 in both lower limbs. Deep tendon reflexes were depressed all over and bilateral planter response was flexor. Differential diagnosis in our patient included hypokalemic periodic paralysis, myositis and renal tubular acidosis. Laboratory investigation revealed random blood glucose of 112 mg/dl, serum sodium 144 mmol/liter, serum potassium 1.3 mmol/liter, pH 7.30, serum bicarbonate 20.3 mmol/liter, serum chloride 112 mmol/liter, partial pressure oxygen 83 mmHg, oxygen saturation of 93% and partial pressure carbon dioxide 34 mmHg. Haemogram, kidney function and liver function tests at presentation are depicted in (table 1). Muscle enzymes in the form of Creatine phosphokinase (CPK) were 1823 U/liter (0-195) and Lactate dehydrogenase (LDH) 886 U/liter (100-235). In view of severe hpokalemia, patient was immediately started on intravenous potassium chloride infusion at a rate of 30 meq/hour and oral potassium was started simultaneously in a dose of 20 meq 6 hourly. There was a predictable improvement in muscle weakness after initiation of potassium therapy and patient became ambulatory after 24 hours of treatment. In view of severe hypokalemia, systemic acidosis and skeletal deformities, a strong possibility of renal tubular acidosis was made. Subsequently, patient was subjected to anthropometric examination which revealed severe dispro-

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Table 1. Laboratory parameters of the patient

Parameter	Reference Value	Presentation	Patient Value In Hospital	Discharge
Creatinine	0.5-1.5 mg/dl	0.8	0.67	0.65
Calcium	8.5-10.5 mg/dl	9.6	9.9	9.3
Phosphorus	1.8-4.6 mg/dl	3.4	3.3	2.3
ALT	0-45 U/Liter	117	77	43
ALP	50-140 U/Liter	490	497	481
AST	0-45 U/Liter	102	68	57
Bilirubin	0-1.5 mg/dl	0.96	0.80	0.73
Total protein	5.5-8.5 gram/dl	7.5	7.3	7.9
Albumin	3.5-5.5 gram/dl	4.4	4.7	4.9
CPK	0-195 U/Liter	1823	1453	489
LDH	100-235 U/Liter	886	543	346
Haemoglobin	11-14.5 gram/dl	12.7	12.9	12.2
WBC	4-10 $\times 10^3/\mu L$	9.67	7.65	8.65
Platelet	100-400 $\times 10^3/\mu L$	143	156	156
PH	7.35-7.45	7.30	7.27	7.36
Serum bicarbonate	24-28 meq/Liter	20.3	16.5	24.3
Serum sodium	135-145 meq/Liter	144	139	137
Serum potassium	3.5-5.3 meq/Liter	1.3	3.9	4.4
24 hour urinary Ca	50-250 mg/day	-----	472	-----

ALT= Alanine transaminase, ALP= Alkaline phosphatase, AST = Aspartate transaminase, CPK= Creatine phosphokinase, LDH= Lactate dehydrogenase, WBC=white blood cell count, mg/dl= milligram per deciliter, U/L= units per liter, gram/dl= gram per deciliter, µL= Microliter, meq/Liter= miliequivalent/Liter.

portionate short stature (Fig.1) with height of 144 cm (<3rd centile, standard deviation score (SDS)=-4.30), upper segment (74 cm), lower segment (70 cm), upper segment to lower segment ratio (1.05), arm span (142 cm), head circumference (52 cm) and mid parental height of 162 cm. Patient´s weight was 33 kg with a body mass index (BMI) of 15.9 kg/m² (<3rd centile, SDS=-2.85). Patient had well developed secondary sexual characters in the form of pubic hair stage 5, stretched penile length (SPL 10 cm), bilateral testicular volume >15ml. Hearing tests in the form of Rinne and Weber and ophthalmic tests including Schirmer's test were unrevealing. 24 hour urinary calcium was 472 mg per day, urinary creatinine 702 mg per day, urinary phosphorus 824 mg per day and urinary protein of <50mg per day and no glycosuria. After correcting hypokalemia, there was further fall in serum pH to 7.27 with serum bicarbonate of 16.5 mmol/litre. Urinary pH at this time was 7.0.Abdominal ultrasonography (USG) revealed bilateral medullary nephrocalcinosis. Digital abdominal X- ray revealed fine calcified foci in bilateral renal area (Fig. 2). Skeletal survey revealed widening of both wrists and bilateral knock knees (Fig. 3). In view of severe hypokalemia and normal anion gap (12 mmol/litre) hyperchloremic metabolic acidosis and inability to acidify urine with associated hypercalciurea and no apparent secondary cause, a diagnosis of primary distal renal tubular acidosis was made. However exact etiological diagnosis could not be established because of unavailibity of genetic testing in our Centre.

Discussion

Renal tubular acidosis (RTA) refers to the development of metabolic acidosis because of a defect in the ability of the renal tubules to maintain acid base homeostasis. All forms of RTA are characterized by a normal anion gap (hyperchloremic) metabolic acidosis (3). Distal (type 1) RTA is characterized by an impaired capacity of hydrogen ion secretion in the collecting tubules. The major causes of distal RTA in adults are autoimmune diseases (eg, Sjögren's syndrome and rheumatoid arthritis), hypercalciuria (which is the primary defect in some families), dysproteinemic syndromes and certain genetic mutations (2, 3, 6). In comparison, hereditary distal RTA is the most common cause in children. Genetic mutations in the basolateral chloride-bicarbonate exchanger and in the apical hydrogen-ATPase have been identified (9). In children, distal RTA is almost always ob-

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served as a primary entity (4). Prominent clinical features include impairment of growth, polyuria, hypercalciuria, nephrocalcinosis and potassium depletion. Although hypokalemia is common in renal tubular acidosis (RTA), acute paralytic presentation is very rare (1, 5). There is a wide variety of conditions giving rise to potassium deficiency as well as a wide variety of effects that such a deficiency might have on many processes related to cellular metabolism (6). Although, the most common form of hypokalemic periodic muscular paralysis is reported to be the familial periodic paralysis (FPP) variety, some patients with chronic potassium depletion may exhibit episodic weakness (7, 8). The clinical presentation of hypokalemic periodic paralysis secondary to RTA is quite similar to FPP (1). The muscle weakness is episodic and generally related to limbs and limb girdle while the respiratory, cranial and bulbar musculatures are rarely involved. However in its severe form, most deaths have been reported due to respiratory failure and cardiac arrhythmias (6). While FPP presents acutely, RTA may have an insidious course with a definite history of muscle pains and polyuria preceding muscle paralysis. In addition, patient with RTA may have skeletal abnormalities and growth restriction as was seen in our patient. Although hypokalemia and decreased resting membrane potential which blocks the action potential is the underlying mechanism for muscle paralysis in both RTA and FPP, the genesis of hypokalemia is different in the two entities. While in the former, there is loss of potassium; in the latter, there is redistribution of potassium between the intra and extracellular compartment (1, 3, 6). It will be imperative to emphasize here that both the entities require different modalities of intervention to prevent life threatening hypokalemia. Alkali and potassium replacement is lifesaving in distal RTA but alkali alone can be life threatening in FPP as it facilitates intracellular potassium influx and worsen hypokalemia. Similarly, acetazolamide, which is important in prophylaxis of FPP, can worsen acidosis in distal RTA (1, 3). Rhabdomyolysis due to hypokalemia has been reported in literature but is very rarely seen in RTA, as majority of the patients are diagnosed earlier due to other manifestations of disease. Rhabdomyolysis in our patient was evidenced clinically by severe muscle cramps and biochemically by marked elevation of muscle enzymes. The genesis of rhabdomyolysis in hypokalemia is believed to be hypokalemia-induced impairment of muscle perfusion and metabolism, which together contribute to muscle dysfunction (11, 12).

Review of literature has revealed that most hypokalemic periodic paralysis (HPP) patients with RTA are adult females (7, 8). Our patient is unique as he is an adult male and has presented in the second decade of life by which time, other stigmata of RTA in the form of skeletal deformities and nephrocalcinosis are already present. If the patient would have been diagnosed earlier, these complications could have been prevented as it has been advocated that early recognition and initiation of alkali supplementation can prevent the progression of disease in RTA (2, 10).

Conclusion

Non-specific complaints like aches and pains should not be ignored in pediatric and adolescent age groups. A high degree of suspicion for RTA should be kept in such patients especially in the presence of restricted growth and skeletal abnormalities as early diagnosis and initiation of therapy can prevent long term sequalae and fatal outcome.

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