Metabolic Interplay : Renal Tubular Acidosis inducing Metabolic Bone Disease and Hypokalemic Periodic Paralysis

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INTRODUCTION

Renal disorders caused by disturbance in acid-base homeostasis caused by tubular dysfunction in acid excretion or bicarbonate ion reabsorption are collectively known as renal tubular acidosis (RTA). The subtypes are as follows:

Type 1: Distal RTA
Type 2 : Proximal RTA
Type 3 : Mixed RTA
Type 4 : Hyporeninemic hypoaldosteronism RTA

Renal tubular acidosis comprises a group of disorders that are characterised by low capacity of net acid excretion and persistent metabolic acidosis, despite a preserved glomerular filtration rate, showing frequent association with hypokalemia.

DIFFERENTIAL DIAGNOSIS

Provisional diagnosis elicited by the history was put forth as metabolic bone disease like osteomalacia or osteoporosis secondary to renal tubular acidosis in view of past urinary diversion procedure.

Other differentials that may be put forth based on history include Inflammatory arthritis, Acute inflammatory demyelinating neuropathy (AIDP), Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), Hypokalemic paralysis, etc.

FINAL DIAGNOSIS AND MANAGEMENT

Final diagnosis was put forth as acquired distal renal tubular acidosis causing Hypokalemic periodic paralysis and metabolic bone disease - osteoporosis, resulting in bilateral shaft of femur fracture (due to bilateral neck and shaft of femur insufficiency).

Potassium citrate was given to correct the underlying renal tubular acidosis and hypokalemia leading to normalization of potassium levels along with calcium carbonate for osteoporosis.

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3. Alexander RT, Bitzan M. Renal Tubular Acidosis. Pediatric Clinics of North America. 2019 Feb;66(1):135–57.
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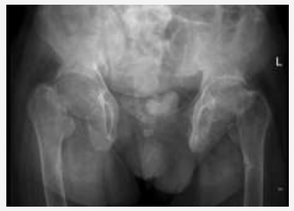
CASE PRESENTATION

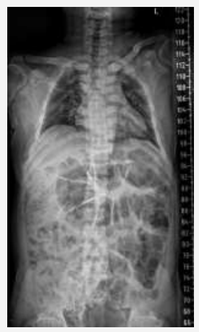
A 35 year old male presented with complaints of right hip, thigh and knee pain of mild to moderate intensity for the past 3 months which increased on weight bearing, walking and movement of right limb, and decreased on rest. Patient provided history of progressive lower limb deformity along with genu valgum and coxa vara since 6 years ago associated with pain and difficulty in walking.

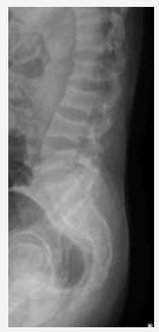
- Patient underwent urinary diversion procedure at the age of 10 years.
- He experienced 2 episodes of hypokalemic periodic paralysis in the last 10 years and was hospitalised previously due to febrile illness, atrial fibrillation, weakness of upper and lower limbs.
- On examination, bilateral thigh and calf wasting was present along with bilateral knee in valgus position and coxa vara. Range of movement of the hip was normal but painfully restricted. Tenderness was present over the right hip and knee region.
- Proximal muscle weakness of the bilateral lower limb was elicited. Further, there was reduced bulk in the bilateral lower limbs.



PLAIN RADIOGRAPHS OF LUMBAR SPINE

Biconcave deformity of visualised lumbar vertebrae noted with decrease in bone density - significant of osteoporosis

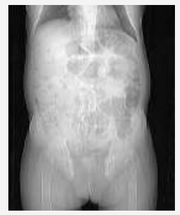


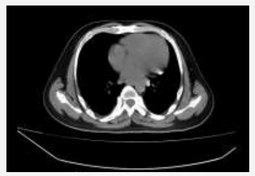


INVESTIGATIONS

Biochemical Analysis (Serum) :
Urea : 129 mg/dL
Creatinine : 2.14 mg/dL
Potassium : 2.2 mmol/L
Bicarbonate : 3.00 mmol/L
Calcium : 8.7 mg/dL
Phosphorus : 3.0 mg/dL

Arterial Blood Gas :
pO2 : 183.80 mmHg
pCO2 : 21.40 mmHg
HCO3 : 6.70 mmol/L
Total Hb : 10.74 g/dL
pH : 7.11





PLAIN CT STUDY OF KUB

Kidney : Bilateral hydroureteronephrosis (right moderate and left minimal) noted with dilated proximal ureters . Few air foci noted in the right renal pelvis. No calculus seen. Mild pelvicalyceal wall thickening seen

Urinary bladder : Not visualised. Post cystectomy status, continent urinary diversion

Basi-cervical fracture of the neck of the right femur, fracture proximal shafts of both femur is noted. Diffuse osteopenia of visualised bones noted-likely due to renal osteodystrophy.

DISCUSSION

Renal tubular acidosis (RTA) is a rare and frequently undiagnosed multiple electrolyte disorder associated with hypokalemia leading to hypokalemic periodic paralysis characterised by episodes of muscle weakness and arrhythmia concomitant to the variations in blood potassium levels. Further, acidosis results in metabolic bone disease. This case is one of Type 1 - Distal RTA resulting as a consequence of urinary diversion procedure the patient underwent in his childhood. Common presentations include osteomalacia, growth failure, hypercalciuria resulting in nephrocalcinosis and recurrent UTIs.

CONCLUSION

- Renal tubular acidosis should be considered in patients presenting with metabolic bone disease.
- It is an important differential diagnosis for patients presenting with hypokalemic periodic paralysis.
- Due to high index of clinical suspicion and following thorough clinical and biochemical assessment, appropriate treatment should be started early on leading to significant improvement in quality of life.

ACKNOWLEDGEMENT

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