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CASE REPORT IV OPEN ACCESS

Metabolic Bone Disease due to Renal Tubular Acidosis as a Primary Manifestation in a Patient with Sjogren's Syndrome: A Case Report

Bisht A,[1] Shrestha PS,[2] Mall D,[3] Wasti K[4]

[1] Senior Resident, Diabetes and Endocrine Unit, Department of Internal Medcine National Academy of Medical Sciences, Kathmandu, Nepal, [2] Consultant, Diabetes and Endocrine Unit, Department of Internal Medicine National Academy of Medical Sciences, Kathmandu, Nepal,[3] Assistant Professor, Diabetes and Endocrine Unit, Department of Internal Medicine National Academy of Medical Sciences, Kathmandu, Nepal,[4] Senior Resident, Diabetes and Endocrine Unit, Department of Internal Medcine National Academy of Medical Sciences, Kathmandu, Nepal

Abstract

Background: Renal tubular acidosis (RTA) is a group of diseases where metabolic acidosis develops due to inability of renal tubules to maintain acid base balance despite normal glomerular filtration rate. Uncorrected acidosis in RTA can cause excess osteoclastic bone resorption and result in severe metabolic bone disease like rickets, osteomalacia and pathological fracture. Sjogren's syndrome can cause distal RTA(type 1). Here, we report a case of a 35 years old female who presented with metabolic bone disease secondary to RTA resulting from Sjogren's syndrome

Key words: Metabolic bone disease, Renal Tubular Acidosis, Sjogren's Syndrome

Introduction

There is inability of kidneys to acidify urine in distal parts of nephron in distal RTA. Distal RTA has association with autoimmune diseases like primary Sjögren syndrome and Systemic Lupus Erythematosus. Distal RTA should be considered if a patient comes with unexplained normal anion gap acidosis.[1] It can present as rickets, growth failure, osteomalacia, hypokalaemia and arrhythmia.[2]

CASE REPORT

A 35-year lady without prior comorbidities presented with complains of easy fatiguability and generalized body ache and difficulty in getting up for 3 months which were worsening. A month after onset of symptoms, she developed limping gait and pain over right hip which was moderate to severe and mechanical in nature. Gradually she developed similar pain over multiple sites. One week prior to hospital visit she had become bed ridden. There was no history of fever, diarrhea, early morning stiffness, photosensitivity, rashes and sicca symptoms. At presentation, the patient had tachycardia and deep labored breathing. Cardiovascular, respiratory, abdominal, higher mental functions, cranial nerves and sensory examinations were unrevealing. Power, tone and gait couldn't be assessed due to pain. Initial investigations are shown in table 1.

Correspondence Author

Dr Anjal Bisht, anjalbisht@gmail.com, Senior Resident, Diabetes and Endocrine Unit, Department of Internal Medicine, National Academy of Medical Sciences, Kathmandu, Nepal, Contact number +9779841820383

Table 1: Investigations before and after treatment

		Pre-treatment		Post-treatment	
	Normal range				
Sodium	135-145 mEq/l	141			
Potassium	3-5 mEq/l	2.9	2.6	4.6	4.0
Urea	15-45 mg%	32			
Creatinine	0.4-1.4 mg%	1.1			
Calcium	8-11mg%	8.2			
Albumin	3.5-5.5 gm%	3.8			
Phosphorous	2.5-4.5 mg%	3.9			
Vitamin D	30-100 ng/ml	15			
iPTH	10-65pg/ml	106			75
ALP	42-128 IU/L	650	550	330	
HCO3	22-28 mEq/L	9.0		14.7	20
pH	7.35-7.45	7.1		7.313	7.45
Serum chloride	96-106 mEq/l	123			
Anion Gap	8-12 mEq/L	9			

She had hypokalemia and her blood gas analysis showed hyperchloremic non anion gap metabolic acidosis. Urine potassium creatinine ratio was suggestive of renal potassium wasting. There was no hematuria or albuminuria. There was no nephrolithiasis or nephrocalcinosis. Hypokalemia was corrected with IV supplementation. After correcting hypokalemia 30 meq of citrate 4 times a day was started. As the patient had low bicarbonate, urine ph> 5.5 and improved after low dose bicarbonate, a diagnosis of type 1 RTA was considered. X-ray showed Looser's zone in bilateral scapula, ribs, bilateral superior and inferior pubic rami and old impacted fracture of right neck of femur. (Figure 1) DEXA scan showed severe osteoporosis. (Figure 2) ANA panel showed strongly positive Anti Ro and Anti La. The patient was then referred to rheumatology and nephrology where a diagnosis of Sjogren's Syndrome with distal type 1 RTA leading to osteoporosis was made. The patient was managed with vitamin D, calcium and bicarbonate supplements, steroids and mycophenolate. Later bisphosphonates was started. On subsequent follow up, the patient's condition has improved.

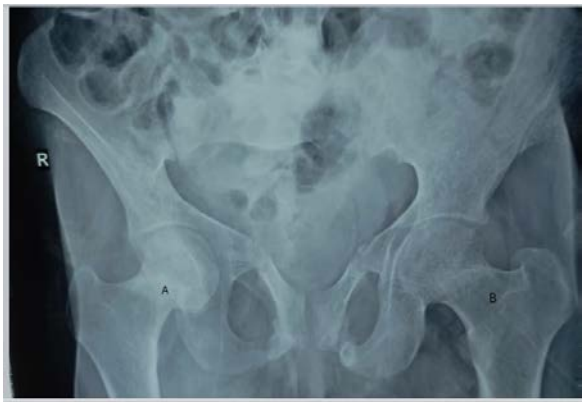

Figure 1. X-ray pelvis of the patient showing A. area of fracture B. area suggestive of osteoporosis

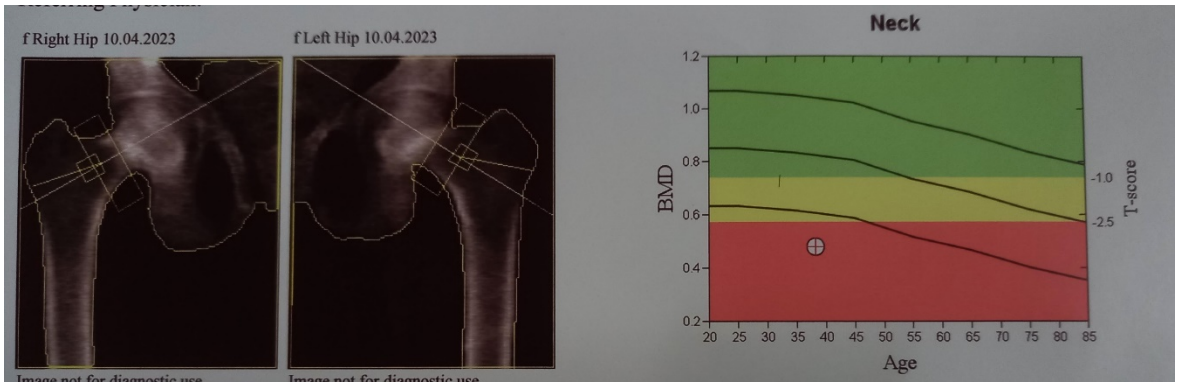

Figure 2. Dexa scan of bilateral hips showing osteoporosis

DISCUSSION

RTA is a group of diseases where metabolic acidosis develops due to inability of renal tubules to maintain acid base balances despite normal glomerular filtration rate.[3] RTA has normal anion gap metabolic acidosis. It results from net retention of hydrogen chloride or the net loss of sodium bicarbonate.[4] In distal RTA distal nephrons are unable to maximally increase secretion of protons in the presence of metabolic acidosis. RTA should be suspected in patients with normal anion gap who have no extra-renal bicarbonate losses. Urinary anion gap is an indirect measure of urinary ammonium. High urinary anion gap in the background of normal anion gap metabolic acidosis and hypokalemia can indicate distal RTA. Sometimes bicarbonate loading test is performed. Normally there is limited residual buffering capacity of the HCO3−/pCO2 couple in the distal nephrons. Increasing HCO3−delivery to distal nephrons make bicarbonate the preferred buffer.[5,6] Uncorrected acidosis in RTA can cause exaggerated osteoclastic bone resorption. Hypokalaemia, hypophosphataemia, vitamin D deficiency and secondary hyperparathyroidism can result due to tubular dysfunction resulting in severe metabolic bone disease like rickets osteomalacia pathological fractures and secondary osteoporosis.[7] Causes of distal RTA include autoimmune diseases like Sjogren's syndrome. Renal manifestations occur in lesser than 10 percent of primary Sjogren's syndrome.[8] This patient presented with weakness, generalized body ache and bone pain at various sites. The evaluation of patient revealed type 1 RTA with hypokalaemia, metabolic acidosis and metabolic bone disease in the form of fragility fracture and osteoporosis. The cause of underlying RTA was Sjogren's syndrome. Correction of metabolic acidosis, hypokalemia and treatment of underlying cause with immunosuppressant in the form of mycophenolate and steroids along with treatment of osteoporosis resulted in improvement.

CONCLUSION

Renal tubular acidosis can cause metabolic bone disease resulting in osteoporosis and fractures. RTA is a known manifestation of Sjogren's syndrome. A high index of suspicion is required to look out for Sjogren's syndrome in a young female who presents with unexplained hypokalemia, metabolic acidosis and metabolic bone diseases.

Consent

Case Report Consent Form was signed by the

patient's husband (guardian) and the original article is attached with the patient's chart.

Conflict of interest
None

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