HYPERKALEMIC DISTAL RENAL TUBULAR ACIDOSIS WITH HYPORENINEMIC HYPOALDOSTERONISM IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS — A CASE REPORT

T K Lim
K Y Fong

Medical Unit II
National University of Singapore
Singapore General Hospital
Outram Road
Singapore 0316

T K Lim, MBBS, M Med
Lecturer

K Y Fong, MBBS
Medical Officer

SYNOPSIS:

A 43-year old woman with lupus nephritis was admitted in acute hyperchloremic metabolic acidosis. Her serum potassium was 6.8 mmol/L, blood pH 7.28 and urine pH 6. She had impaired urinary potassium excretion associated with attenuated renin and aldosterone response to salt depletion and ambulation. She appeared to have both "classic" distal RTA and hyperkalemic, hyporeninemic hypoaldosteronism.

INTRODUCTION

Distal, "Classic" renal tubular acidosis (RTA) or Type-1 RTA is a well defined disorder of renal tubular function and the early description by Fuller Albright in 1946 remains the most lucid (1). The syndrome is characterised by hypokalemia, hyperchloremic metabolic acidosis and inability to lower the urine pH below 5.5 (2). The acquired variety had been associated with a host of autoimmune diseases including Systemic Lupus Erythematosus (SLE) (2).

Acquired distal RTA is a heterogenous group of disorders — it includes the "classic" hypokalemic group but the hyperkalemic or type-4 RTA has attracted the most interest recently and is considered to be the more common variety (3). Distal hyperkalemic RTA has been described with diabetes mellitus, obstructive uropathy, potassium — sparing diuretics, hemoglobin S disease, Addison's disease, postrenal transplantation and isolated aldosterone deficiency (4, 5, 6).

This is a case report of a woman who presented with lupus nephritis and was found to have hyperkalemic, hyporeninemic hypoaldosteronism and distal RTA.

CASE REPORT

A 43-year old Thai woman, was referred to this unit for the problem of intermittent epigastric pain of 6 months duration. She was under treatment by the referring physician for nephrotic syndrome. There was no history of joint pains or ingestion of potassium supplements.

On admission she was noted to be pale and have a blood pressure of 180/100 mm Hg. There was periorbital and bilateral ankle edema. The rest of the cardiovascular and respiratory systems were normal. Palpation of the abdomen revealed epigastric tenderness but no guarding. Per rectal examination showed brown stools. There was no facial rash or arthritis.

Investigations showed her haemoglobin (Hb) level to be 5 gm/dl. She was transfused and the Hb rose to 8.3 gm/dl. Her total white cell count was 2,500/mm^3, platelet count was 210,000/mm^3 and reticulocyte count of 1.2%. A collagen screen showed the presence of LE cells (22/100 Polys) and anti-nuclear antibody. Serum complement level (CM 50) was 16 units/ml. The direct Coomb's test was negative.

Urine microscopy showed 20–25 red blood cells, 10–12 white blood cells, occasional epithelial cells, few granular casts and the presence of albumin ($^{++}$). Culture of the urine grew no organisms. Her serum total protein was 4.9 gm/dl (N-6.2–8.2) and albumin was 1.7 gm/dl (N-3.7–5.1). 24 H urinary protein showed 1.12 g/specimen (Vol 700 ml) and the calculated creatinine clearance was 29.5 ml/min (Vol 1700 ml). A diagnosis of systemic lupus erythematous with nephrotic syndrome from lupus nephritis was made and she was started on prenisolone.

A gastroduodenoscopic examination was normal. Ultrasound examination of the abdomen showed both kidneys to be approximately 10 cm in bi-polar length with no other abnormalities.

A bone marrow biopsy was also performed which showed a hypoplastic marrow. No fibrosis was seen.

Serial determinations of electrolytes and acid-base:

Days in Hospital	1st	3rd	7th	12th	17th
Urea (mg/dL)	89	67	137	135	167
Na^+ (mmol/L)	137	134	139	133	135
K^+ (mmol/L)	6.8	6.6	4.4	4.1	4.2
Cl– (mmol/L)	121	116	113	99	102
Creatinine	1.4		1.9	1.4	1.5
HCO_3 (19–31 mmol/L)	15.5	19.2	22	29	20.9
Blood pH	7.28	7.35			7.41
Urine pH	6	5			

Patient was given Resonium A and intravenous sodium bicarbonate when initial results were known and subsequently started on Shol's solution ($NaHCO_3$ and citrate) on the 3rd post admission day.

Other laboratory results obtained were: Serum Calcium of 6.0 mg/dL (N-8.4–10.4) and serum phosphate of 6.2 mg/dl (N-2.4–4.3). Her 24 hour urinary electrolytes showed: Na^+ — 172 mmol/spec (Vol 2100 ml), K^+ — 30 mmol/spec. and Ca^{2+} — 25.2 mg/spec. The renin and aldosterone levels were also determined 2 hours after i/v fursemide and ambulation. Plasma renin activity was 0.37 ng/ml/H; and plasma aldosterone was 25 pg/ml.

She was discharged with Tab Frusemide, Prednisolone, Methyldopa and Shol's solution.

DISCUSSION

This patient presented with lupus nephritis and "classic" distal RTA. She had frank hyperchloremic metabolic acidosis and was unable to acidify her urine. Her persistent hyperkalemia was puzzling and lead us to suspect metabolic acidosis initially and to study her urinary electrolyte excretion and renin-aldosterone activities subsequently. She had impaired potassium exretion at 14 mmol/l of urine when her serum potassium was 6.6 mmol/dl. This was accounted for by her attenuated renin and aldosterone response to salt depletion and ambulation.

She appeared to have two distinct disorders, renin-aldosterone deficiency and hyperkalemic distal RTA. Hyporeninemic hypoaldosteronism alone could explain the impaired renal potassium excretion but not the impaired renal acid secretion (her urine pH was 6 when blood pH was 7.28 and HCO_3 15 mmol/l) which was strongly suggestive of distal, "Classic" RTA (3, 4). A very similar patient had recently been discussed at a Nephrology Forum (7).

She responded well to resonium and sodium citrate but developed mild prerenal azotemia as a result of both fursemide therapy and mineralocorticoid deficiency, we were unable to study her renal tubular function in more detail because she returned to Thailand for further follow up.

Renal tubular damage is a prominent feature of lupus nephritis and the association with "Classic" distal RTA is well documented (2). Both unexplained hyperkalemia and hyporeninemic hypoaldosteronism have been described in patients with SLE. These are uncommon associations and have described only in isolated case reports (8, 9).

Our patient appears to have a combination of both "Classic" distal RTA and the hyporeninemic, hypoaldosteronism in association with hyperkalemia. We are not aware of and previous such association with SLE.

References

1. Albright F, Burnett Ch, Parson W: Osteomalacia and late rickets. The various etiologies met in the United States with emphasis on that resulting from a specific form of renalacidosis, the therapeutic indications for each etiological subgroup, and the relationship between osteomalacia and Milkman's syndrome. Medicine, 1946; 25: 339–479.
2. Cogan MG, Retor FC, Seldin DW. Acid-base disorders, in Brenner B, Rector FC (eds): The Kidney, Philadelphia, WB Saunders Co, 1981: 841–907.
3. De Fronzo RA, Hyperkalemia and Hyporeninemic hypoaldosteronism. Kidney Int, 1980; 17: 118–38.
4. Batlle DC, Jose A, Kurtzman NA, Hyperkalemic Distal Renal Tubular acidosis associated with obstructive nropathy. N Engl J Med: 1981; 304: 373–80.
5. Sebastian A, Schambelan M, Findenfield S, Morris RC Jr. Amelioration of metabolic acidosis with fludrocolisone therapy in hyporeninemic hypoaldosteronism. N Engl J med, 1977; 297: 576–83.
6. McSherry E. Renal Tubular acidosis in Childhood. Kidney Int. 1981; 20: 799–809.
7. Kurtzman NA. Acquired distal renal tubular acidosis. Kidney Int. 1983; 24: 807–19.
8. De Fronzo RA, Cooke CR, Goldberg M, Cox M, Myers AR, Agusz S. Impaired Renal tubular potassium secretion in systemic lupus erythematous. Ann Intern. Med. 1977; 86; 268–71.
9. Kiley J, Zager P, hyporeninemic hypoaldosteronism in two patients with systemic lupus erythematosus. Am J Kidney Dis. 1984; 4; 39–43.