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Renal Tubular Acidosis with Muscle Paralysis

Pages with reference to book, From 260 To 263

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Introduction

Four patients are described who were admitted at The Aga Khan University Hospital exhibiting muscle paralysis and hypokalemia in association with renal tubular disease. All four patients responded well to treatment. The purpose of this report is to describe the clinical features demonstratedby these patients, to discuss etiology and mechanism of the disease and to emphasize the importance of early diagnosis and treatment.

Case 1: 19 year old gentleman came to emergency room of the Aga Khan University Hospital on March 18, 1989 with a two day history of progressive weakness leading to quadreparesis. There was no difficulty inbreathing and no history of bowel or urinary incontinence. Family history was negative for this kind of illness. Routine blood analysis showed a potassium of 0.7 mmol/L (repeated twice), chloride 117 mmol/L, bicarbonate 15.7 mmoIIL,. urine pH of 5.0. He was treated with intravenous potassium chloride which resulted in rapid regaining of the muscle strength. His hemogram, thyroid function tests, serum creatinine and 24 hr urinary potassium were normal. He was discharged on oral potassium. In September, 1992 he again developed muscle weakness with respiratory difficulty. He was given intravenous potassium by a general practitioner but had very little improvement, so he was brought to the emergency room where he was found to have a sodium of 138 mmol/L, potassium of 2.6 mmol/L, chloride 120 mmoIJL, bicarbonate 12.6 mmol/L, blood urea nitrogen 12.0 mg/dl, serum creatinine 0.9 mg/dl. arterial blood gas analysis showed pH of 7.254, p02 of 110,5 mmHg, pCO2 of 31.9 mmHg, base excess of -11.7, bicarbonate of 14.1 mmol/L, urine pH was 6 and specific gravity of 1.0 10. The electrocardiogram showed changes of hypokalemia, roentgenograms showed no evidence of osteomalacia or nephrocalcinosis. His muscle strength recovered on intravenous potassium chloride and he was later discharged on oral potassium and sodium bicarbonate.

Comments

This is a typical example of distal renal tubular acidosis (RTA). The unusual features were the presence of acidic urinary pH on first admission which delayed the diagnosis of the condition. The patient till the writing of the paper is enjoying relatively good health with no further recurrences.

Case 2:44 year old lady presented to The Aga Khan Hospital emergency room with the complaint of generalized weakness leading progressively to flaccid quadriplegia. She was a diagnosed case of rheumatoid arthritis and taking non-steroidal anti- inflammatory medications for pain. Her family histoiy was unremarkable for any illness. She appeared normal on examination except for a flaccid quadnplegia. The haemoglobinwas normal. Serum electrolyte showed a sodium of 139 mmoJIL, potassium 1.4 mmolIL, chloride 124 mmol/L, bicarbonate 13.3 mmolIL, blood urea nitrogen was 5.0 mg/dl, serum creatinine of 1.1 mg/dl, arterial blood gas analysis showed metabolic acidosis, examination ,of the urine disclosed a specific gravity of 1.0 10, pH of 6.5, electrocardiogram findings were consistent with that of hypokalemia. Roentgenograms findings were normal. She was started on intravenous potassium chloride, her muscle strength started to improve but on tl1e second day of the treatment she developed carpopedal spasm of both bands, her serum electrolytes at that time showed the potassium of 2.7 mmol/L, chloride 133 mmol/L, bicarbonate 12 mmol/L, calcium 7.5 mg/dl, phos-

phorus 1.3 mg/dl. She was given intravenous calcium gluconate which resolved her carpopedal spasm and she gained full muscle strength within 48 hours. She was fully ambulant at the time of discharge with the serum potassium of4. 1 mmolJL; her discharge medication included oral potassium supplements and sodium bicarbonate tablets.

Comments

This is a typical case of renal tubular acidosis with potassium depletion and paralysis. Carpopedal spasm developed during the recovery period because of decreased lionized calcium secondary to alkali therapy which resolved on calcium administration.

Case 3: 26 year old, 7 month pregnant lady started feeling lethargic and weak, which she initially attributed to the pregnancy, but weakness progressed to the extent that she was unable to sit-up in bed or lift her hand against the gravity. Her past history was unremarkable, inparticularshe was not taking any medications apart from the iron supplements. There was no family history of similar illness. Examination revealed a normal looking young lady, pulse rate 120/min, normotensive, afebrile and markedly tachypneic with respiratory rate of 40/min. On central nervous system examination she was conscious and oriented; her mental functions were normal but had dysarthria because of difficulty in opening the mouth; cranial nerves were normal except for the weak orbicularis oculi, decreased gag reflex and difficulty in protrusion of tongue. Tone and power were decreased in all four limbs, deep tendon reflexes sluggish with down going planters. Rest of the systemic examination was normal. Routine blood tests showed the haemoglobin of 9.5 gms/dl, hematocrit of 27.6 percent, erythrocyte sedimentation rate of 40 mm/hr., random blood sugar 107 mg/dl, blood urea nitrogen of6 mg/dl, serum creatinine 0.7 mg/dl, serum sodium 139 mmol/L, serum potassium 2.3 mmoI/L, chloride 120 mmol/L, bicarbonate 8.2 mrnol/L, calcium 7.2 mg/dl, albumin 2.4 g/dl, globulin 3.8 g/dl. Arterial blood analysis revealed a pH of 7.45, pCO2 16.3 mmHg, p02 115.4 mmHg, bicarbonate 11.5 mmolJL, base excess 8.3. Urinary pH was 7.0, specific gravity 1.015. Patient was treated with intravenous potassium chloride and sodium bicarbonate which resulted in marked improvement pf her symptoms. On the second day of the treatment, frank tetany appeared with carpopedal spasm and a positive Chevostek's and Trousseau's sign for 24 hours. Her speech became fluent. and comprehensive with the full return of muscle strength in 48 hours. She was discharged on oral potassium and Aibright solution. She subsequently underwent nonnal delivery with no recurrence of any symptoms.

Comments

The most interesting finding here was the overcompensated respiratory alkalosis which could be partially accounted forby associated pregnancy. The presence of low bicarbonate on several occasions during the hospital confirmed the diagnosis of acidosis. The carpopedal spasm was due to hypocalcemia which was precipitated by the alkali replacement.

Case 4: A 26 year old lady was admitted to The Aga Khan University Hospital in August 1993 because of the sudden onset of progressive flaccid quadriparesis. During this period she was fully conscious, continent, withno respiratory distress ordysphagia. Her past medical history and family history was unremarkable and she was not taking any drug. Her exaniination revealed a healthy looking woman, except she was not moving her limbs, was vitally stable with mild tachypnea. Neurologically, she was conscious, oriented and cranial nerve examination was normal excepta poor trapezius effort. Power was decreased and reflexes were sluggish. The haemoglobin was normal. The blood urea nitrogen was 14 mg/dl, sodium 137 mmol/L, potassium 1.5 mmol/L, chloride 116 mmol/L. bicarbonate 9.4 rnmol/L, calcium 8.7 mg/dl. Arterialblood gas analysis showed pH of 7.428, pCO2 21.1 mmHg, pO2 114.6 mmHg, bicarbonate 13.9 mmol/L, base excess 7.2. The urine pHwas6.0 units. Roentgenograms of chest and abdomenwere normal, The patient was treated with intravenous and oral potassium. There was complete return of muscle function within 48 hours. On the second day of admission she developed carpopedal spasm lasting for 24 hours during her course of treatment for which she was given intravenous calcium gluconate. At this time the serum potassium was 2.7 mmolJL, calcium was 7.6 mg/dl, total protein6.4 g/dll, albumin 3.0 g/dl, globulin 3.4 g/dl, AJG ratio 0.9. The patient became

completely asymptomatic on the third day with normal electrolytes, therefore, was discharged on oral potassium supplements. She was again admitted a year later with severe quadriplegia and hypokalemia (1.7 mEq/L). Again she was treated with intravenous potassium chloride and discharged on third day of admission with normal power and the potassium of 3.2 mmol/L. She has been well till the writing of this paperon therapy with alkalimzmg solution and potassium supplements and has had no further difficulty.

Comments:

Typical case of renal tubular acidosis, both the episodes were prompted by severe deficit of potassium. Patient gave this history of fever couple of days, approximately 10 days preceding each episode. Tetany again was the feature during the correction of hypokalemia and acidosis.

Discussion

The major clinical, chemical and urinary findings are tabulated in Table.

Table. Clinical features.

Patient	Case 1	Case 2	Case 3	Case 4
Clinical features				
Age (Yr) and Sex	22	44F	26F	26F
Family history	-ve	-ve	-ve	-ve
Nephrocalcinosis	-ve	-ve	-	-ve
Abnormal state of consciousness	-ve	-ve	-ve	-ve
Tetany	-ve	+ve	+ve	+ve
Blood chemical findings on admission				
Sodium (mmol/L)	138	139	139	137
Potassium (mmol/L)	2.6	1.4	2.3	1.5
Bicarbonate (mmol/L)	12.6	13.3	8.2	9.4
Chloride (mmol/L)	120	124	120	116
BUN/Creatinine mg/dl	12.0/1.0	5.0/1.1	6.0/0.7	14.0/1.0
Calcium (mg/dl)	-	-	7.2	8.7
Urine findings on admission				
pH	6.0	6.5	7.0	6.0
Sp. gravity	1.010	1.010	1.015	-
Spot potassium	-	18.7	15.0	21.0

Acute hypokalemic paralysis is an uncommon cause of acute motor weakness. Morbidity and mortality associated with unrecognized disease include respiratory failure and death. Hence, it is imperative for physicians to be knowledgeable about the causes of hypokalemic paralysis and consider them in the differential diagnosis. The hypokalemic paralysis represent a heterogeneous group of disorders with a final common pathway presenting as acute weakness and hypokalernia. Most cases are due to familial hypokalemic periodic paralysis; however, sporadic cases are associated with diverse underlying etiologies including thyrotoxic periodic paralysis, barium poisoning, renal tubular acidosis, primary aldosteronism, licorice ingestion and gastrointestinal potassium losses. The approach to the patient with hypokalemic paralysis includes a vigorous search for the underlying etiology and potassium replacement therapy. Further therapy depends on the etiology of the hypokalemia. Disposition depends

on severity of symptoms, degree of hypokalemia and chronicity of disease[1]. In the patients described, the cause of hypokalemia was distal renal tubular acidosis leading to muscle weakness. A diagnosis of renal tubular acidosis should be considered in the presence of a normal anion gap acidosis[2] in the absence of substantial renal impairment. In such circumstances, a urinary pH of >5.5 is suggestive of renal tubularacidosis, if urinary tract infection is excluded. Traditionally, renal tubular acidosis is classified numerically into fourtypes. Hypokalemia is a feature of type I and II. A urine pH of<5.6 excludes type I but not type II renal tubular acidosis. If a urinary infection with the urea splitting organism (e.g., Proteus) has been excluded, a urinary pH of >5.6 is suggestive of renal tubular acidosis[3]. By far, the commonest form is type I renal tubular acidosis also called the distal renal tubular acidosis[4]. The classic type I renal tubular acidosis is characterized by a hyperchloremic metabolic acidosis accompanied by a reduced net acid excretion and the inability to lower urinary pH below 5.5 pH (usually higher than 6.0) in the face of spontaneous acidemia or after acid loading[5-11]. The defect in distal acidification impairs hydrogen ions secretion into the urine in exchange for potassium ion; consequently, urinary potassium losses are raised even in the presence of hypokalemia[12]. There are many causes of type I renal tubular acidosis, it can be primary (idiopathic) as well as secondary to wide variety of diseases including the autoimmune disorders (Sjogren's syndrome, chronic active hepatitis, primary biliaiy cirrhosis, rheumatoid arthritis), multiple myeloma, primary hyperparathyroidism, marked volume depletion and drugs (amphotericin B, lithium, toluene, ifosfamide).

Clinical features and manifestation

As was seen in the cases the usual presenting complaints of patients include musculoskeletal symptoms such as joint or muscle pains, muscle weakness or low back pain[16] in addition to the symptoms of the underlying cause. Other symptoms of hypokalemia include paresthesias, polyuria and thirst. Rarely, severe hypokalemia may produce paralysis. Unless hypokalemia is recognized in these patients respiratory failure and death may occur. Potassium depletion will cause a renal concentrating defect with resulting volume depletion, as well as an increase in the synthesis of prostaglandins in the vessel endothelium. Both effects cause hypotension with consequent hyperreninemia and hyperaldosteronism, which aggravates potassium wasting. Treatment of the acidosis may correct the secondary hyperaldosteronism and potassium loss, thus normalizing the semm potassium[13]. At times, diagnosis of distal renal tubular acidosis is suspected from the incidental fmding of nephrocalcinosis on a simple x-ray study of the abdomen. Sometimes, a renal stone, usually composed of calcium phosphate, is the initial presentation of distal renal tubular acidosis. Severe complication of chronic acidosis, such as myocardial failure, lethargy and coma are rare. Buffering of acid by bone salts is markedly enhanced during chronic metabolic acidosis. The result of bone buffering acid is one of the most disastrous consequences of distal renal tubular acidosis. Bone demineralization results from release of calcium carbonate from bone to neutralize excess hydrogen ions[17]. The penalty for buffering the accumulating acid is metabolic bone disease. A negative calcium balance is usual and hypercalciuria is present in about 30% of patients. The negative calcium balance raises parathyroid hormone levels, leading to secondary hyperparathy roidism and osteomalacia[13].

Diagnosis

The diagnosis of renal tubular acidosis can also be suspected from knowledge of the various disorders that can cause the syndrome. For practical purposes, the diagnosis of distal renal tubular acidosis starts by exciusionofothercauses of hyperchioremic metabolic acidosis. It is useful to consider whether plasma potassium is nonnal. low or elevated. On the basis of plasma potassium, renal tubular acidosis can be separated into different types. The diagnosis of distal renal tubular acidosis is then confirmed by the finding of a urinaiy pH above 5.5 in the face of spontaneous or ammomum chloride-induced metabolic acidosis. In general, if the patient is spontaneously acidotic (pH <7.35), there is no need to induce severe acidosis in order to evaluate uriiiaiy acidification4. Ammonium chloride test is

performed by administering 0.1 g/kg ammonium chloride orally over 1-2 hour to produce a fall in plasma bicarbonate of 3-5 nunol/litre. Distal renal tubular acidosis is diagnosed if the urinary pH is persistently >5.51. When the animonium chloride test is contraindicated by liver disease, a calcium chloride test (2meq/kg) should be performed[12,13].

Treatment

The treatment consist of the correction of underlying cause whenever possible. Classic distal renal tubular acidosis requires oral administration of alkalinizing salts to correct the acidosis. The amount of bicarbonate is calculated from the following formula: (desired plasma bicarbonate minus observed plasnm bicarbonate) x0.6 body weight in kilograms. The alkali requirement of 1 to 2 mEq per kg body weight of sodium bicarbonate daily is sufficient in most cases to buffer dietary acid production. The alkalinizing agent may be sodium bicarbonate (lgm=2rnEq) or Shohl solution (140 gin sodium citrate and 100 gm citric acid per litre of solution; lml=lmEq base) and should be given in two to three divided doses. Sodium bicarbonate is considerably cheaper than Shohi solution. Shohi solution should be used only if patient cannot tolerate the gastric bloating caused by sodium bicarbonate[13]. Effective alkali therapy can normalize urinary calcium and citrate excretion, prevent nephrocalcinosis and nephrolithiasis and restore nonnal growth in children[18]. Any potassium deficit should be corrected acutely. Chronic potassium replacement may be necessary, but loss of potassium is generally decreased with treatment of the acidosis[18]. As was observed in the three of the four cases discussed, that during treatment tetany resulted due to the decrease in ionized serum calcium resulting from alkali therapy, monitoring of serum calcium concentration and prompt treatment is recommended. Semm potassium levels and motor power is returned to normal within twenty four to forty eight hours.

Conclusion

In the light of above discussion, we recommend that in all patients with hypokalernic paralysis, arterial blood gas analysis, serum bicarbonate and urinary pH should be done to make the diagnosis of renal tubular acidosis, since the correct treatment can reverse many of the consequences of hypokalemia and prevent many complications like nephrolithiasis and bone diseases.

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