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Case Report

Hypokalemic Periodic Paralysis Due To Distal Renal Tubular Acidosis

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ABSTRACT

Hypokalemic periodic paralysis (HPP) is a disorder that characterized by attacks of skeletal muscle paralysis depending on the changes in serum potassium levels, and can occur due to primary and secondary causes. One of the secondary causes of HPP is distal renal tubular acidosis (DRTA). DRTA is a disorder that characterized by hypokalemia or hyperkalemia hypercalciuria, metabolic acidosis and alkaline urine. DRTA's clinical symptoms are listed as constipation, nausea, vomiting, kidney and skeletal muscle complications, nephrocalcinosis, urolithiasis and severe hypokalemia crisis. In this case report, we reported a patient who admitted to emergency department with complains of nausea, vomiting, and periodic muscle weakness and was diagnosed with hypokalemic periodic paralysis due to DRTA was presented.

Key words: *Renal tubular acidosis, hypokalemic, periodic, paralysis*

Distal Renal Tübüler Asidoza Bağlı Hipokalemik Peryodik Paralizi

ÖZET

Hipokalemik Peryodik Paralizi (HPP), serum potasyum düzeyindeki değişikliklere bağlı olarak iskelet kaslarında paralizi atakları ile karakterize primer ve sekonder nedenlere bağlı oluşabilen klinik tablodur. Ataklar, günler veya aylar içinde tekrarlayabilir ve birkaç saat veya birkaç gün sürebilir. Atak sırasında serumdaki potasyum seviyesi düşük olup, ataklar arasında serum potasyum seviyesi genellikle normaldir. HPP'nin sekonder nedenlerinden biri de distal renal tubuler asidoz (DRTA)'dur. DRTA, metabolik asidoz, alkali idrar, hipopotasemi veya hiperpotasemi ve hiperkalsiüri kliniği ile karakterizedir. Kabızlık, bulantı, kusma, böbrek ve kas iskelet sistemine ait komplikasyonlar, nefrokalsinozis, ürolityazis ve ciddi hipokalemik kriz DRTA'nın klinik seyrinde görülebilir. Bu makalede bulantı, kusma ve periyodik kas güçsüzlüğü şikayetleriyle acil servise başvuran ve distal renal tübüler asidoza sekonder hipokalemi tespit edilen sporadik bir olgu sunulmuştur.

Anahtar kelimeler: *Renal tubuler asidoz, hipokalemik, periyodik, paralizi*

INTRODUCTION

Hypokalemic Periodic Paralysis (HPP) is a clinical entity characterized by temporary acute paralysis as a result of decreased serum potassium levels due to primary or secondary causes. There are familial or sporadic forms of HPP. Distal renal tubular acidosis (DRTA) is one of the rare secondary causes (1). DRTA is characterized by normal anion gap, hyperchloremic metabolic acidosis, bicarbonaturia, and decreased hydrogen (H+) secretion. DRTA may be primary or secondary. Primary DRTA may have an autosomal dominant or recessive trait. Secondary DRTA may co-occur with renal disorders causing tubular dysfunction, disorders of calcium metabolism (vitamin D intoxication, idiopathic hypercalciuria, and primary hyperparathyroidism), genetic disorders, and autoimmune diseases (2). In this paper we report a patient with HPP who presented to emergency department with muscle weakness and subsequently diagnosed with DRTA (Type 1).

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CASE

An 18-year-old female patient admitted to emergency department with a 2-day history of nausea, vomiting, and weakness in arms and legs. She had no history of drug use or any disease. She has admitted to emergency department with similar complaints for 3 to 4 times within the last 6 months. She was given some unknown drugs but no studies were performed for the causes of her complaints at those times. Her family history was not remarkable for any disease. At physical examination she was in moderate general condition, conscious, and cooperated. Her vital signs were as follows: Blood pressure: 100/70 mmHg, body temperature: 36.7 ºC, pulse rate: 76 bpm, rhythmic. She had muscle strength loss of 2/5 both in upper and lower extremities. Her deep tendon reflexes were hypoactive and Babinski reflex was indifferent. Laboratory tests were as follows: Hb: 12.8 gr/dL, Htc: 37.5%, platelet count: 231000/mm3, white blood cell count: 5600/mm3, urea: 20 mg/dL, creatinine: 0.86 mg/dL, Na: 135 mEq/L, K: 2.2 mEq/L, Cl: 118 mEq/L, Ca: 9.2 mg/dL, Mg: 1.9 mg/dL, parathormone: 24 pg/mL. Results of the arterial blood gas analysis were as follows: pH: 7.2, PCO2: 26 mmHg, PO2: 85 mmHg, HCO3: 8.5 mEq/L, BE: -20.5. Urinalysis results were as follows: pH: 7.0, density: 1003, Na: 142 mmol/L, K: 10 mmol/L, Cl: 104 mmol/L, urinary anion gap (sodium+potassium - chloride): 48 mmol/L. Four hours after oral ammonium chloride loading at a dose of 100 mg/kg urinary pH was reanalyzed and found to have increased to 6.5. Based on this result, the patient was diagnosed with distal renal tubular acidosis3.

Search for etiology of DTRA revealed normal thyroid function tests (free T3, free T4, TSH), cerebrospinal fluid analysis, C3, C4, cryoglobulin and immunoglobulin, serum copper, ceruloplasmin, plasma renin, and aldosterone levels. Anti-DNA, RF, ANA, SS-A, SS-B, and lupus cell were all negative. Electrocardiography and echocardiography, thyroid and abdominal ultrasonographic examinations, brain magnetic resonance imaging, and electromyographic examinations were within normal limits. Based on available information, no muscle biopsy was deemed necessary. The patient was admitted to intensive care unit with the diagnosis of DTRA-induced HPP. Her bicarbonate deficit was calculated with the following formula: bicarbonate deficit = (target bicarbonate level- patient's bicarbonate level) X body weight X 0.4. Accordingly, bicarbonate infusion was begun. ABG analysis 12 hours later revealed a pH:7.32 and HCO3:15.2 mEq/L, and the infusion was completed to 24 hours4. KCL infusion at a rate of 20 mEq/L/hour was also commenced5. Following a 2 hour infusion potassium level was 3.7 mEq/L. The patient was put on prophylactic acetazolamide 250mg/day 5. At 1 month follow-up serum potassium level was 4.2 mEq/L and the treatment was continued.

DISCUSSION

HPP is a heterogeneous disease characterized by attacks of skeletal muscle paralysis secondary to periodic drops in serum potassium levels. Hypokalemia may be secondary to potassium deficit as well as abnormal intracellular potassium shift. It may be familial or sporadic (3-8). The familial form is more common in Western societies, while sporadic disease is more prevalent in Asian countries. The familial HPP is the most common cause of HPP with a prevalence of 1/100000. Two-thirds of cases are autosomal dominant while 1/3 is sporadic. Clinical picture of sporadic cases resembles that of familial cases. The paralytic crises start at first or second decade. The frequency of the attacks increases between the ages of 15 to 30 years whereas they tend to occur less frequently with advancing age. The attacks are precipitated by food rich in carbohydrates and salt, emotional stress, and post exercise resting (9). Secondary causes of HPP include thyrotoxicosis (47.1%), diuretics (11.8%), DRTA, Gitelman syndrome, and liquorice consumption (each having a rate of 5.9%), and primary hyperaldosteronism (2.9%) (10,11). Our case was considered a sporadic case since she had no family history for HPP.

Patients with HPP may admit to emergency department with nonspecific complaints of muscle weakness, nausea, vomiting, constipation, and numbness. Symptoms secondary to hypokalemia become prominent with a serum potassium level below 2.5 - 3 mEq/L (12). The main finding in HPP is symmetric loss of muscle strength, especially in shoulder and pelvic muscles. Rarely, an asymmetrical involvement may also be observed, in which unilateral arm or leg is involved (13). Our patient had nausea and vomiting in addition to equal and symmetrical loss of muscle strength in both upper and lower extremities; this attack was also similar to previous ones our patient had experienced.

Renal tubular acidosis (RTA) is diagnosed by measuring pH from the first urine in the morning in addition to

simultaneous measurement of serum electrolytes, urea, creatinine, and ABG analysis. A positive urinary anion gap in addition to a pH>5.5 and hypopotassemia suggests DRTA (type 1) (13). Our patient had a low urine density, simultaneous hypopotassemia in the blood sample, and normal anion gap metabolic acidosis in ABG analysis. Among the differential diagnoses, Type 4 RTA was excluded since our patient had hyperchloremic metabolic acidosis in addition to a urine pH greater than 5.5, a low serum potassium level, and normal plasma cortisol, aldosterone, and renin levels. The proximal (Type 2) RTA was also ruled out because of a positive urinary anion gap. Urinary acidification test used to demonstrate renal acid (H+) secretion defect in DRTA can be done with oral ammonium chloride (NH4Cl, 100 mg/kg) or oral fluodrocortisone (0.1 mg) given with furosemide 40 mg. In normal subjects the urinary pH should be lowered to less than 5.2 with oral NH4Cl loading, while it remains unchanged or exceeds 6 in DRTA3. In our patient the diagnosis of DRTA was confirmed by a urinary pH of 6.5 measured 4 hours after oral NH4Cl loading test. The secondary causes of DRTA were also sought in our patient. As no other diseases explaining the clinical picture can be detected in our patient, we considered her a sporadic case. Since our patient had metabolic acidosis and bicarbonate deficit, we replaced potassium and bicarbonate levels until both of them were normalized (4).

Although DRTA rarely causes HPP, it should be remembered in differential diagnosis of patients with HPP who also have hyperchloremic metabolic acidosis, hypopotassemia, and a positive urinary anion gap.

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