The Journal of Clinical Medicine

Case Report

A Practical Approach to Refractory Hypokalaemia: A Rare Presentation of Bartter Syndrome

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Received Date : Sep 19, 2023
Accepted Date : Sep 22, 2023
Published Date : Oct 26, 2023

ABSTRACT

Hypokalaemia could be a common finding in hospitalized patients. In most cases the cause are going to be obvious. However, during a subgroup of patients the cause is often unsure and establishing the designation could gift difficulties and might become a challenge. In such cases, measure of urinary indices e.g. excretion atomic number 19/creatinine concentrations along side blood acid/base parameters and assessment of urinary excretion potassium, chloride, metal and creatinine are utilized within the medical diagnosis. this text presents a case report of AN man with severe resistant hypokalaemia that AN initial atomic number 19 level of 1 mmol/L and related to limb weakness, alkalosis and hypercalcinuria. His atomic number 19 level was refractory to oral and blood vessel replacements. This patient showed a marked increase of humor atomic number 19 once administration of nonsteroidal anti-inflammatory. This patient had the total organic chemistry options of Bartter syndrome (BS), there was a severe hypokalaemia, with inappropriate excretion K+ loss, with severe urinary chloride excretion related to delicate alkalosis and traditional pressure level. He had conjointly hypocalcaemic hypercalcinuria, His excretion calcium/creatinine quantitative relation was zero.26 that was indicative of hypercalcinuria. His age and late symptom presentation would have steered Gitelman sickness however thanks to hypercalcinuria, the clinical designation of Bartter Syndrome was created. we have a tendency to describe here a case of Bartter Syndrome sort three whose designation was created in adult life. This patient is AN uncommon, probably extreme case of bachelor's degree. Being thirty years recent at the time of designation, he's out and away the oldest patient to date delineate with bachelor's degree. hard the urinary potassium/creatinine quantitative relation, urinary chloride excretion and urinary calcium/creatinine quantitative relation in conjunction with plasma acid/base values provided straightforward and reliable tests to tell apart Bartter syndrome from different medical diagnosis of hypokalaemia and to produce a correct management within the medical care unit till any advanced investigations to be done.

Keywords : Bartter syndrome; Gitelman syndrome; Refractory hypokalaemia; Trans-tubular potassium concentration gradient; Urinary potassium-creatinine ratio; Urinary calcium-creatinine ratio

INTRODUCTION

Hypokalaemia may be a frequent finding in hospitalized patients. In most cases the reason behind hypokalaemia is typically evident within the clinical history. However, in some patients the cause is often not clear and identification will become a challenge. In such cases, mensuration of urinary indices e.g. urinary excretion atomic number 19, chloride, metallic element and creatinine in conjunction with blood acid/ base parameters are used within the pathophysiological identification. this text presents a case report of AN man with severe persistent hypokalaemia that was refractory to oral and blood vessel replacements. This patient showed a marked increase of humour atomic number 19 when administration of nonsteroidal anti-inflammatory drug.

CASE REPORT

A 30-year-old male; no previous medical record, given to the emergency roll complaining of bilateral lower limb weakness with fasciculations in hands and legs and inability to run within the past in some unspecified time in the future. No history of trauma, fever, incontinence of stool or viscus, sensory complaints, or history of alcohol or abuse. within the sorting space, a blood gas was obtained and showed severe hypokalaemia (potassium of one mmol/L) with

gentle alkalosis. He was shifted to the revitalization space, placed on pads with two massive bore IV lines inserted, EKG obtained (Figure 1), and was started on atomic number 19 replacement.

On examination, BP: 98/40, Pulse: 90, SPO2: ninety nine on space air, Temperature: thirty seven degrees stargazer, and on medicine test his power within the right lower limb was 3/5 and also the left lower limb was 2/5. Otherwise sensation, deep connective tissue reflexes, and neural structure signs were traditional, pupils were two metric linear unit reactive briskly and his GCS was 15/15. His alternative examination parameters were traditional.

Apart from severe hypokalaemia, there was hypophosphatemia (1.6 mg/dL), gentle hypocalcemia (8.6 mg/dl), normomagnesemia (1.7 mg/dL), hypochloraemia (86 mmol/L), magnified random weewee Na (41 mmol/L), magnified random weewee atomic number 19 (58.9 mmol/L) and magnified weewee chloride (150 mmol/L); the science lab workup together with a septic exercise, nephritic perform, thyroid perform tests, and plasma and weewee osmolality was traditional. The patient was admitted underneath care of the medical team and remarked the medical care unit (ICU) for persistent low atomic number 19 and risk of arrhythmias because of prolongation of QTc. Also, EKG showed prolonged PR interval and proof of hypokalaemia (U waves in leads II, V2, V3, and V4, and ST-segment depression in leads II, III, aVF). A nephritic echography of our patient showed 2 adjacent tiny stones in left nephritic scroll with gentle pelvicalyceal dilatation with absence of renal lithiasis or alternative nephritic abnormalities.

While within the intensive care unit, the patient received massive doses of oral and IV Klorvess (KCL) through a central line daily and it had been noted that his weakness improves with the atomic number 19 replacement; but, his atomic number 19 blood levels were continuously low despite regular replacement (average 280 mmol daily) (Figure 2) and despite replacement of each metallic element and phosphate. The patient was persistently nephropathy.

A medical diagnosis of refractory hypokalaemia was investigated. In such case, mensuration of urinary indices like discharge rate of atomic number 19 and random weewee potassium/ creatinine concentrations in conjunction with blood acid/base parameters are used within the medical diagnosis [1]. Urinary atomic number 19/creatinine magnitude relation is employed as a straightforward acceptable index of potassium excretion and to determine a correct identification and management during this medical emergency. His main organic chemistry parameters, as shown in Table one, indicate that he had a severe hypokalaemia, with inappropriate weewee K loss. This patient had the total organic chemistry options of Bartter syndrome: There was severe urinary chloride excretion (>150 mmol/l) related to gentle alkalosis and traditional force per unit area, additionally he had hypocalcemic hypercalcinuria. His weewee metallic element (4.1 mg/dL): weewee creatinine (16 mg/dL) magnitude relation was zero.26 that was indicative of hypercalcinuria. the most medical diagnosis is with Gitelman syndrome, that additionally referred to as the hypocalciuric variant of Bartter syndrome however because of hypercalcinuria, the clinical identification of Bartter syndrome was created.

The patient had a resistant severe hypokalaemia in spite of huge atomic number 19 replacement. Hypokalaemia was marked improved when administration of nonsteroidal anti-inflammatory drug in dose of fifty mg double daily. when nonsteroidal anti-inflammatory drug, his atomic number 19 levels were maintained at a standard level and his daily KCL demand attenuated (Figure 3). The patient was shifted to the medical ward and later discharged to home and for advanced more workup of Bartter Syndrome to be worn out the patient clinic.

This patient is AN uncommon, presumably extreme case of Bartter syndrome, being thirty years previous at the time of identification, he's out and away the oldest patient to date represented with Bartter syndrome. His age and late symptom presentation would have recommended the a lot of Gitelman unwellness, however hypercalcinuria and clearance knowledge recommended Bartter syndrome.

DISCUSSION

Hypokalaemia may be a common clinical downside that the etiological designation will ofttimes be supported the patient history and therefore the clinical state of affairs [1]. The cause will typically be determined from the history such as hypokalaemia thanks to vomit, diarrhoea, or diuretic drug use and no any investigation are going to be necessary. However, in a very subgroup of patients the cause is often unsure and establishing the designation could gift difficulties [2].

In things wherever the reason behind hypokalaemia isn't clear, measuring of urinary atomic number 19 excretion and assessment of ara} and acid-base values are usually useful [3].

The causes of hypokalaemia will be merely divided into 2 categories: The one condition includes diseases inflicting urinary organ atomic number 19 wasting like diabetic diabetic acidosis, Cushing's syndrome, primary adenosis, Bartter syndrome, Gitelman syndrome, hypomagnesemia, and urinary

organ tube-shaped structure pathology, furthermore as intake of loop and diuretic drug diuretics, which ends in K loss by the kidneys; and therefore the alternative consists of these resulting in extra-renal excretion or transient shifting of K into cells, as in glandular disorder periodic disfunction, exogenous hypoglycemic agent infusion, hyperinsulinemia, vomiting, and diarrhea [4].

The traditional approach to differentiate between urinary organ and extrarenal causes of hypokalaemia relies on urinary atomic number 19 excretion measured in twenty four hours water samples or random urinary atomic number 19 concentration values. A urinary atomic number 19 concentration that's over fifteen to twenty mmol/day or over fifteen to twenty mmol/L suggests that there ar urinary organ causes for atomic number 19 wasting. Hypokalaemia within which these values ar but fifteen mmol/day or fifteen mmol/L severally indicates that there ar extrarenal causes for atomic number 19 depletion. getting a twenty four hours atomic number 19 excretion rate isn't sensible in a very medical emergency as a result of atomic number 19 replacement should run promptly which might compromise the accuracy of twenty four hours urinary atomic number 19 [5].

A spot water assortment before atomic number 19 replacement ought to be accustomed facilitate for establishing the designation [5]. In our patient there was high urinary atomic number 19 concentration (58.9 mmol/L) that indicates there ar urinary organ causes for atomic number 19 loss. However, random measurements is also deceptive because the water atomic number 19 concentration is decided by each the quantity of atomic number 19 within the water and therefore the water volume [6]. typically there ar several overlapping values of the spot urinary atomic number 19 concentration as nephropathy is common in patients with hypokalaemia. The nephropathy is also thanks to thirst or defective urinary organ concentration in patients with chronic hypokalaemia that ends up in a coffee price of the urinary atomic number 19 concentration albeit a major urinary organ atomic number 19 loss is gift. Therefore, a additional reliable take a look at is needed [5].

The trans-tubular atomic number 19 concentration gradient (TTKG) is associate index reflective the degree of atomic number 19 excretion within the animal tissue collection ducts of the kidneys and indirectly assessing adrenal cortical steroid bioactivity in patients World Health Organization have hypo- or hyperkalaemia [7,8]. measuring of the TTKG is taken into account superior to measuring of urinary atomic number 19 alone for assessing the contribution of urinary organ excretion to atomic number 19 levels. (9) TTKG ar helpful within the pathophysiological medical diagnosis of hypokalaemia [1].

The trans-tubular atomic number 19 gradient estimates the magnitude relation of atomic number 19 within the cannular fluid at the tip of the animal tissue collection tube-shaped structure to it within the peritubular capillaries and may be calculable from:

TTKG=(urine atomic number 19 × Osmolality plasma)/(plasma potassiumM× Osmolality urine).

For this formula to be correct, water osmolality should exceed plasma osmolality and water Na ought to be larger than twenty five mmol/L [7,8]. In our patient, the water osmolality was 381 mOsm/kg/H2O that exceed the plasma osmolality of the patient (281 mOsm/kg/H2O) and therefore the water Na was forty one mmol/L (greater than twenty five mmol/L).

The normal urinary organ response once hypokalaemia is thanks to non- urinary organ causes may be a trans-tubular atomic number 19 gradient but a pair of that indicates intracellular shift of atomic number 19. whereas a trans- cannular atomic number 19 gradient over three indicates hypokalaemia of urinary organ origin (renal atomic number 19 wasting) [9]. In our patients there was a TTKG was terribly high (27.15) that is indicative of magnified secretion of K+ within the animal tissue collection ducts.

However, there ar recent studies that advise to shift in stress from the TTKG to alternative variable reflective atomic number 19 excretion e.g. the urinary atomic number 19/creatinine magnitude relation thanks to intrarenal organic compound utilization that ends up in a better rate of urinary organ excretion of potassium [10,11].

The other variable reflective atomic number 19 excretion, the urinary potassium/creatinine magnitude relation (U(K/Cr)), has been accustomed measure the reason behind hypokalaemia [5]. measuring of the urinary potassium/creatinine magnitude relation is most well-liked as a result of it's not influenced by the water volume. U(K/Cr) is also a valuable marker to differentiate between urinary organ and extra-renal K loss [4,6].

Many studies have shown that the urinary atomic number 19/ creatinine magnitude relation will be accustomed differentiate between hypokalaemia caused by urinary organ atomic number 19 loss and hypokalaemia caused by extrarenal potassium depletion, as found in particle channelopathy hypokalemic periodic disfunction, by intracellular shift of liquid body substance atomic number 19 into musculus [4,6,12].

The effectualness of the urinary potassium/creatinine magnitude relation was evaluated in a very previous study of cardinal patients with severe hypokalaemia related to disfunction. The urinary atomic number 19/creatinine

magnitude relation was ready to differentiate between the thirty patients with hypokalemic periodic disfunction (HPP) (whose hypokalaemia was caused by an interior shift of animate thing atomic number 19 into the cells) and therefore the 13 patients with hypokalaemia largely thanks to urinary organ potassium wasting (Ten of them had urinary organ cannular pathology or Gitelman syndrome).

Urinary Ca excretion is vital as a result of it differentiate between the 2 syndromes. In distinction to the hypocalciuria of Gitelman syndrome, Bartter syndrome patients area unit typically documented to own symptom [18]. Although twenty four hours assortment of water Ca is best, random water Ca measure in relevancy creatinine are often performed. the conventional reference for the water Ca (mg/dl): water creatinine (mg/dl) magnitude relation is a smaller amount than zero.14. Values extraordinary 0.2 area unit found in patients with symptom [22]. In our patient, the water Ca (mg/dl): water creatinine (mg/dl) magnitude relation was zero.26 that was indicative of symptom along side severe urinary chloride excretion (150 mmol/L) and dogging severe hypokalaemia because of excretory organ atomic number 19 wasting. the most medical diagnosis is with Gitelman Syndrome, conjointly referred to as the hypocalciuric variant of Bartter Syndrome, however because of symptom the clinical designation of Bartter syndrome was created. Hypercalciuria predisposes Bartter Syndrome patients for lithiasis and renal lithiasis (calcification of the excretory organ tissue) [16]. A excretory organ echography of our case showed 2 adjacent little stones in left excretory organ whorl with gentle pelvicalyceal dilatation. Thus, redoubled water Ca excretion could also be thought-about because the doable risk issue for stone formation in our patient.

The persistent steady variety of hypokalaemia in bachelor's degree and GS might suddenly become life threatening beneath bound exacerbating conditions. Physicians got to be cognizant of such excretory organ cannular disorders and promptly treat patients in danger [18].

Distinguishing between bachelor's degree and GS isn't invariably easy because of composition variance. The genetic designation is currently doable however there area unit many limitations together with inconvenience and price. As mentioned before, GS is usually related to hypocalciuria and bachelor's degree with symptom, though some patients with sort 3 Bartter syndrome have traditional urinary Ca excretion [18,20,23].

At this, Bartter Syndrome can`t be cured, and treatment is especially directed for correction of dehydration and electrolytes disturbance, i.e. hypokalaemia and doable hypomagnesemia. merely increasing the intake of single electrolytes features a very little profit, since urinary excretion will increase proportionately [16]. The focus of chronic medical aid of bachelor's degree and GS patients includes electrolytes replacement furthermore as inhibition of secondary will increase of autocoid production and/or the renin- angiotensin-aldosterone axis, that exacerbate the urinary solution loss. Hypokalaemia conjointly induces autocoid production [18].

Indomethacin may be a anti-inflammatory drug that getting used in Bartter syndrome to assist improve growth in youngsters and reduce urinary atomic number 19 excretion. Indocin has been shown to get rid of vasoconstrictor insensitiveness to angiotensin II and to lower plasma renin activity and mineralocorticoid concentration in an exceedingly patient with Bartter's syndrome [24]. Indocin has been shown to inhibit autocoid synthetase, that support the hypothesis that autocoid excess may be a basic infective mechanism in Bartter's syndrome [25]. Indomethacin considerably reduced eGFR, plasma protease concentration and will increase plasma atomic number 19 concentration. Indocin conjointly decrease the speed of urinary excretion of atomic number 11 and phosphate [25,26]. Indomethacin will cause improvement in laboratory parameters generally seen in bachelor's degree patients. The dose of Indocin are often as high as 2-3 mg/kg/day. future gi aspect effects of Indocin together with gastritis and viscus ulcers are often dose restricted. Cox a pair of selective inhibitors could also be thought-about as another choice. a similar response to Indocin isn't seen with GS as a result of patients with GS don't generally show the redoubled autocoid production seen in patients with bachelor's degree [18]. Our patient had a resistant severe hypokalaemia in spite of enormous atomic number 19 replacement. Hypokalaemia was marked improved once administration of Indocin in dose of fifty mg doubly daily. Our patient showed marked increase of humour atomic number 19 once administration of Indocin.

This patient is Associate in Nursing uncommon, probably extreme case of Bartter syndrome. His designation remained unknown till adult life, being thirty years previous at the time of designation, he's far and away the oldest patient to this point represented with Bartter syndrome. His age and late symptom presentation would have instructed Gitelman sickness, however symptom instructed Bartter syndrome.

In summary, in our case report, there was a dogging severe hypokalaemia because of excretory organ atomic number 19 wasting and to determine a correct designation and management during this medical emergency, urinary atomic number 19-creatinine magnitude relation is employed as an easy applicable index of potassium excretion. symptom

along side high urinary chloride excretion in conjunction with plasma acid-base values facilitate for the clinical designation of Bartter syndrome (BS). Our case suggests that BS3 could also be presenting in adult life, as in G.S. symptom and clearance information ought to enable differentiating it from the additional common Gitelman sickness.

Conclusion

Calculating the urinary potassium/creatinine magnitude relation, urinary chloride excretion and urinary calcium/creatinine magnitude relation in conjunction with plasma acid/base values provided straightforward and reliable tests to tell apart Bartter syndrome from different medical diagnosis of hypokalaemia and to produce a correct management within the medical care unit till any advanced investigations to be done.

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