
Case report

A Case of Pseudo-Bartter Syndrome and Summary of the Approach to Metabolic Alkalosis

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Abstract

Background. Pseudo-Bartter Syndrome is characterized with metabolic alkalosis without renal tubular defect. Here we present a patient with severe metabolic alkalosis and review the approach to metabolic alkalosis.

Case report. A 50 year-old male presented to the emergency unit with nausea, vomiting, decreased urine output and distorted consciousness. He had projectile vomiting for one month and anuria during the last two days. Abnormal laboratory results were: Urea: 95.7 mmol/L, creatinine: 689.5 μ mol/L, potassium: 2.28 mmol/L, chloride: 50 mmol/L, pH: 7.57, HCO₃⁻: 61.4 mmol/L and pCO₂: 64 mmHg and decreased urinary chloride concentration (17.5 mmol/L). Dimensions of both kidneys were normal with ultrasonography. Upper gastrointestinal endoscopy showed an ulcer causing pyloric obstruction. He was given isotonic saline followed by poliuria and normalization of laboratory abnormalities.

Discussion. Our patient presented with cyclic vomiting, uremia, uremic encephalopathy, severe metabolic alkalosis, hypokalemia and hypochloremia. In the differential diagnosis of metabolic alkalosis all causes that may lead to hypovolemia, hypochloremia and hypokalemia must be searched. Urinary chloride concentration is the cornerstone for diagnosis. Hypovolemia and hypochloremia induce maximum renal chloride conservation, usually lowering urine concentration to less than 25 meq/L as in our case. He was diagnosed as Pseudo-Bartter syndrome and acute renal failure resulting primarily from pyloric stenosis which has led to vomiting and so extracellular fluid volume contraction with prominent loss of chloride and hydrogen.

Conclusions. Metabolic alkalosis is the most common acid-base disturbance. It is important to recognize the possible causes and plan the treatment accordingly.

Keywords: Metabolic alkalosis, pseudo-Bartter syndrome, pyloric stenosis.

Introduction

Diseases associated with hypokalemic metabolic alkalosis like Bartter and Gitelman Syndromes are characterized by defects in renal tubular epithelial electrolyte

transport [1]. Pseudo-Bartter Syndrome is also characterized with metabolic alkalosis but without any renal tubular defect. It is very important to determine possible underlying diseases when dealing with such disorders. The list of them includes cystic fibrosis [2], inappropriate diuretic use [3], chloride poor diet, bulimia, cyclic vomiting and laxative abuse [4]. In contrary to Bartter syndrome, urinary chloride concentration is decreased in all cases except diuretic use [5]. The treatment depends on the management of the underlying disease and restoration of fluid and electrolyte balance. Here, we present a case of pseudo-Bartter syndrome and review shortly the approach to metabolic alkalosis.

Case report

Investigations

A 50 year-old male presented to the hospital with nausea, vomiting, decreased urine output and distorted consciousness. He admitted that he had cyclic vomiting for the last year and was hospitalized with the same clinical presentation three months ago in another hospital. With oral proton pump inhibitors he was symptomless until the last month during which projectile vomiting about one hour after meals started and increased in severity in the last ten days. He lost about four kilograms of weight during this period. His urine output decreased progressively to anuria during the last two days. He smoked forty pack years. His past medical and family histories were unremarkable. Pathological findings of the physical examination were distorted orientation for time and place, confusion, flapping tremor of the hands, decreased turgor of the skin, dry tongue, hypotension (90/50 mmHg), tachycardia (110 bpm) and clapping at the epigastrium. Abnormal laboratory examination results were as follows: urea: 95.7 mmol/L, creatinine: 689.5 μ mol/L, sodium: 135 mmol/L, potassium: 2.28 mmol/L, chloride: 50 mmol/L, pH: 7.57, HCO₃⁻: 61.4 mmol/L, pO₂: 74mmHg and pCO₂: 64 mmHg. Urinary system ultrasonography revealed that the dimensions and echogenicity of both kidneys were normal; and the bladder was empty and there was no dilatation in the collecting system. The urine analysis which could be performed only after sufficient urine output was maintained two days after admission to the hospital; revealed

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a specific gravity of 1.025, pH of 5.0 with unremarkable sediment findings and no protein, glucose and bilirubin. Urinary chloride concentration was 17.5 mmol/l. The upper gastrointestinal endoscopic examination showed an ulcer causing pyloric obstruction. The ulcer was proven to be benign with pathological examination.

Diagnosis

He was diagnosed as Pseudo-Bartter syndrome due to pyloric stenosis which has led to hypovolemia, metabolic alkalosis and compensatory respiratory acidosis and prerenal azotemia.

Treatment

After restoration of fluid and electrolyte balance with isotonic saline (beginning with 50 ml/hour; and then increasing the infusion rate according to the urine output and nasogastric drainage and vital findings) and potassium (the dose adjusted according to the serum potassium levels); biochemical parameters returned to near-normal levels within one week without need for hemodialysis. His final laboratory findings were as follows: urea: 46 mg/dl, creatinine: 1.5 mg/dl, sodium: 137 mmol/L, potassium: 3.6 mmol/L, Cl: 96 mmol/L, pH: 7.42, HCO_3^- : 31 mmol/L and pO_2 : 82 mmHg. He was given pantaprazol 40 mg/day orally for his peptic ulcer. Then he was transferred to the department of surgery for a surgical intervention.

Discussion

Our patient presented with cyclic vomiting, uremia, uremic encephalopathy, severe metabolic alkalosis, hypokalemia and hypochloremia.

A primary elevation in plasma bicarbonate (HCO_3^-) concentration is usually induced by hydrogen loss from gastrointestinal tract; or in the urine due to diuretic therapy (6). These hydrogen ions are derived from H_2CO_3 ($\text{H}_2\text{O} + \text{CO}_2 \leftrightarrow \text{H}_2\text{CO}_3 \leftrightarrow \text{H}^+ + \text{HCO}_3^-$) and there will be an equimolar generation of HCO_3^- for each milliequivalent of H^+ that is lost. Metabolic alkalosis can also be produced by administration of HCO_3^- ; by movement of H^+ into the cells as in hypokalemia; and by contraction as in diuretic use, gastric losses in patients with achlorhydria and sweat losses in cystic fibrosis.⁶ Volume contraction alkalosis occurs when the fluid that is lost contains chloride but little or no HCO_3^- by the way of secondary elevation of serum aldosteron level.

Normal kidneys possess the ability to correct metabolic alkalosis by excreting excess HCO_3^- in the urine. In case of reduced renal functions and increased HCO_3^- absorption (depletion of effective circulating volume and secondary elevation of aldosteron), this compensation can be overwhelmed and HCO_3^- level rises. Increased proximal tubular reabsorption of HCO_3^- and more importantly decreased distal tubular secretion of HCO_3^- (due to secondary hyperaldosteronism) account for the decreased urinary HCO_3^- [7,8]. Concurrent chloride depletion and hypokalemia also increase distal HCO_3^- reabsorption [9]. Hypochloremia suppresses the activity of

Na-K-2Cl co-transporter leading to NaCl loss and secondary hyperaldosteronism and so increased distal H^+ secretion [9]. Hypochloremia also increases directly luminal H^+ -ATP-ase activity in α -intercalated cells leading to H^+ secretion [10]. When sodium is reabsorbed without chloride, luminal negativity increases leading to H^+ secretion. The net effect is the paradoxical finding of acidic urine despite extracellular alkalemia [6]. Hypokalemia is of primary importance mainly in states of primary mineralocorticoid excess [6]. All these changes are reversed with correction of fluid and electrolyte deficits.

In the differential diagnosis of metabolic alkalosis all causes that may lead to hypovolemia, hypochloremia and hypokalemia must be searched. Urinary chloride concentration is the cornerstone for diagnosis [6]. Hypovolemia and hypochloremia induce maximum renal chloride conservation, usually lowering urine concentration to less than 25 meq/L. In patients with alkali loading and mineralocorticoid excess, who are generally volume expanded, the chloride excretion is equal to intake [6].

Urinary chloride concentration is less than 25 meq/L in cases of vomiting, low chloride intake (in little children), diuretic use (in late stages when hypovolemia ensues), factitious diarrhea, cystic fibrosis and post hypercapnia. Our patient had no history of diarrhea, profuse sweating or use of a diuretic or laxative agent. Post hypercapnia could not be responsible, because the patient had primary metabolic alkalosis and compensatory respiratory acidosis at the time of presentation. He had hypovolemia, hypochloremia and hypokalemia all of which are known to induce metabolic alkalosis with mechanisms just described. Being middle-aged without previous history during childhood, good response to isotonic saline and existence of a clear underlying disease made us think that there was no tubular defect and so we excluded Bartter and Gitelman Syndromes. He was diagnosed as Pseudo-Bartter syndrome and acute renal failure resulting primarily from pyloric stenosis which has led to vomiting and so extracellular fluid volume contraction with prominent loss of chloride and hydrogen.

The treatment of Pseudo-Bartter syndrome depends primarily on that of the underlying disease, with particular attention paid to correction of hypovolemia and hypokalemia. Isotonic saline should be infused at a rate of 50-100 ml/hour (greater than urinary and other sensible and insensible fluid losses) until urinary chloride rises to more than 25 mEq/L [11].¹ Patients with severe metabolic alkalosis sometimes require more urgent correction of serum pH. Hemofiltration and hemodialysis is an option especially in volume overloaded and uremic patients [11]. Potassium losses should be replaced with oral or intravenous KCl. The K^+ deficit is 200-400 mmol in patients with mild to moderate metabolic alkalosis induced by upper gastrointestinal chloride losses. When nasogastric drainage must be continued, H^+ and Cl^- losses can be reduced by administration of drugs that inhibit gastric acid secretion [12]. Acetazolamide in-

creases HCO_3^- excretion, and may be useful especially in volume overloaded patients with diuretic-induced metabolic alkalosis and those with post hypercapnic metabolic alkalosis [11]. Hydrochloric acid may be used cautiously with frequent control of arterial blood gases.

Conclusion

Metabolic alkalosis is one of the most common acid-base disturbances. Clinical presentation may be atypical as encephalopathy or severe acute renal failure. Hence it is important to recognize the possible causes especially those associated with gastrointestinal tract and plan the treatment accordingly.

Conflict of interest statement. None declared.

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