Case Report

Journal of the Royal Society of Medicine Open;
14(6) 1–2
DOI: 10.1177/20542704231183266

Diabetic ketoacidosis in a patient known with Gitelman syndrome

Ida Ringsing Nielsen[1], Patricia Almine Skat-Rørdam[1] and Ivan Werner Jensen[2]
[1]Department of Medicine, Nykoebing Falster Hospital, Nykobing Falster, Denmark
[2]Steno Diabetes Center, Nykoebing Falster, Denmark
Corresponding author: Ida Ringsing Nielsen. Email: idjoh@regsj.dk

Summary

Gitelman syndrome is a rare hereditary nephropathy, which causes chronic metabolic alkalosis with low potassium and magnesium levels. There is no known coherence between Gitelman syndrome and Type-1 diabetes but patients with both diseases that develop diabetic ketoacidosis might present with normal acid status and receive incorrect treatment. In our case report the patient was known with both diseases and quickly diagnosed and treated but the condition is rare and previously only described in two other case reports.

Keywords

Diabetes<endocrinology<CLINICAL, fluid electrolyte and acid-base disturbances<renal medicine<CLINICAL, metabolic disorders<endocrinology<CLINICAL

Introduction

Diabetic ketoacidose and Gitelman syndrome (GS) might be tricky to discover and treat because of preceded alkalosis and electrolyte derangement.[1]

Case

A 20-year-old woman with known GS and T1D, treated with an insulin pump, was admitted to hospital with nausea and vomiting. Upon arrival the patient had an arterial blood gas that showed: pH = 7,24 (7,35–7,45), pCO2 = 3,4 kPa (4,5–6,2 kPa), pO2 = 15,3 kPa (11,1–14,4 kPa), glucose = 28,0 mmol/L (2,9–8,3 mmol/L), potassium = 3,4 mmol/L (3,5–4,6 mmol/L), sodium = 142 mmol/L (137–145 mmol/L), lactate = 2,9 mmol/L (0,5–1,6 mmol/L), bicarbonate = 14 mmol/L (21–27 mmol/L), chloride = 100 mmol/L (98–106 mmol/L), blood ketones = 7 mmol/L (< 3 mmol/L), urine ketones = >15,6 mmol/L, anion gap = 35 mmol/L (10–20 mmol/L) and magnesium = 0,60 mmol/L (0,71–0,94 mmol/L).

The patient was treated after a ketoacidose regime with 10 IU Novorapid intravenously (IV) and 1 L NaCl initially, following 6 IU Novorapid IV per hour and a sodium/potassium mixture IV 500 ml/hour. After 4 h the patient was clinically better and was then treated with a glucose-insulin-potassium IV drop. The day after the patient's blood sugar had normalized. During the patient's hospitalization her blood potassium fell to 2,4 mmol/L despite intake of her usual potassium chloride supplement of 3000 mg four times daily. The patient was given an extra 3000 mg potassium chloride the day before her discharge. The patient was discharged from the hospital after 3 days of treatment for diabetic ketoacidosis with no known cause.

Upon the patient's follow up in nephrologic regime nearly 2 months later her electrolytes were still deranged with blood potassium = 3,0 mmol/L and magnesium = 0,49 mmol/L despite her usual potassium chloride supplement of 3000 mg four times daily, Amiloride 10 mg per day, Spironolactone 100 mg three times daily, Magnesium Citrate 1600 mg four times daily, Magnesium Depot tablet 750 mg four times daily and HbA1c = 95 mmol/mol. Blood samples taken during hospitalization and at follow up can be seen in Table 1.

Discussion

Diabetic ketoacidosis in a patient with GS is rare and has only previously been described in two other case reports respectively from the USA and Tunisia.[2,3]

In our case report the woman was already known with T1D and GS. She was hospitalized with a diabetic ketoacidosis, which quickly was confirmed and treated. A diabetic ketoacidosis can easily be overlooked in patients with GS, as they normally have a tendency to metabolic alkalosis which combined with a diabetic acidosis can make the metabolic status appear normal.[1,4] Therefore, a measure of high blood glucose, high acetate levels and high anion-gap might be important values to discover diabetic ketoacidosis in patients with GS. In our case report the high anion-gap was caused by high levels of acetate, which reflect the high non-

Table 1. The patient's electrolyte balance during hospitalization and at following controls.

Electrolyte balance	01.10.21 5:31 PM	01.11.21 8:38 PM	01.12.21 8:32 AM	01.25.21 11:57 AM	03.01.21 11:57 AM	05.03.21 2:20 PM
Potassium;P (3,5–4,6 mmol/L)	4,4	2,4 ↓	2,4 ↓	3,0 ↓	3,0 ↓	2,9 ↓
Sodium;P (137–145 mmol/L)	132 ↓	138	135 ↓	134 ↓	137	134 ↓
Ionized calcium free;P*(1,18–1,32 mmol/L)				1,16 ↓	1,19	1,30
Magnesium;P (0,71–0,94 mmol/L)	0,60 ↓			0,48 ↓	0,49 ↓	0,54 ↓
Phosphate;P (0,76–1,41 mmol/L)				1,07	0,88	1,26

* Measured at pH = 7,4
↓ indicating values below lower limit of reference intervals

measured anions. Inadequate treatment of T1D might worsen symptoms of GS. Osmotic diuresis caused by high blood glucose levels might worsen electrolyte derangement as hypokalaemia and treatment with Insulin might induce further hypokalaemia in patients with GS. In our case report the potassium levels were near normal at the time of hospitalization but dropped to 2,4 mmol/L during treatment with Insulin and the patient's hypokalaemia persisted despite large doses of per oral potassium supplements. Analysis of the pH-value is important in many life-threatening conditions, but a normal value can still disguise a serious metabolic disturbance as in patients with T1D and GS and mislead the treatment. Even the correct treatment of a diabetic ketoacidosis in patients with GS can further complicate the electrolyte derangement which make diabetic ketoacidose in a patient with GS a complex situation to treat.

Declarations

Declaration of conflicting interests: The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Funding: The author(s) received no financial support for the research, authorship, and/or publication of this article.

Ethical approval: Written informed consent has been obtained from the patient.

Guarantor: Ida Ringsing Nielsen

Contributorship: IRN and PAS contributed equally as primary authors. IWJ contributed as senior author.

Acknowledgements: None.

ORCID iD

Ida Ringsing Nielsen https://orcid.org/0000-0003-0117-3918

References

1. Knoers NV and Levtchenko EN. Gitelman syndrome. *Orphanet J Rare Dis* 2008; 3: 22. doi: 10.1186/1750-1172-3-22. PMID: 18667063; PMCID: PMC2518128.
2. Biagioni M, Marigliano M, Iannilli A, et al. Diabetic ketoacidosis complicated with previously unknown Gitelman syndrome in a Tunisian child. *Diabetes Care* 2011; 34(6): e107. doi: 10.2337/dc11-0127. PMID: 21617100; PMCID: PMC3114359.
3. Zangeneh F, Chiang M and Zangeneh F. Pitfalls in the laboratory diagnosis of diabetic ketoacidosis in Gitelman's syndrome. *Diabetes Care* 2003; 26(3): 955. doi: 10.2337/diacare.26.3.955. PMID: 12610073.
4. He G, Gang X, Sun Z, Wang P, Wang G and Guo W. Type 2 diabetes mellitus caused by Gitelman syndrome-related hypokalemia: a case report. *Medicine (Baltimore)* 2020; 99(29): e21123.