Euglycemic DKA in MODY Patient: Empagliflozin to Blame.

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Euglycemic DKA in MODY Patient: Empagliflozin to Blame

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CASE PRESENTATION

42 year old female with mature onset diabetes of the young (MODY) type 2 presented to the emergency room with complaints of nausea, vomiting and epigastric abdominal pain worsening over four days. She was diagnosed with gestational DM at age 34 and post-partum was unsuccessfully treated with a variety of oral medications. She underwent testing which revealed a glucokinase mutation consistent with MODY type 2. Recently, she was started on empagliflozin-linagliptin in addition to a diet of less than 60 grams of carbohydrates per day. On admission, imaging was unrevealing, and she was diagnosed with gastroenteritis. Her serum glucose was 106 mg/dL, potassium was 4.6 mmol/L, bicarbonate was 21 mmol/L, anion gap was 14, and lipase was normal. Urinalysis revealed glucose of 500 mg/dL, ketones > 160 mg/dL, and specific gravity >1.030. Venous blood gas revealed metabolic acidosis. Beta hydroxybutyrate was 2.71 mmol/L. The patient was diagnosed with euglycemic DKA. Endocrinology discontinued empagliflozin, emphasizing that the only treatment required for MODY type 2 is diet restriction. Her symptoms improved, labs normalized, and she was discharged.

DISCUSSION

- This case demonstrates a deadly combination of inappropriate SGLT-2 inhibitor use causing decreased serum glucose levels in a MODY type 2 patient resulting in euglycemic DKA
- MODY Type 2 results from a defective glucokinase enzyme, therefore increased serum glucose levels are required to trigger insulin secretion
- In this case, empagliflozin resulted in decreased serum glucose and increased glucosuria. As a result, the body's demands for insulin were higher than the amount of insulin being secreted due to defective glucokinase enzyme, inducing DKA.

Table 1 - Genetic and Clinical Characteristics of MODY Subgroups

<table>
<thead>
<tr>
<th>MODY 1</th>
<th>MODY 2</th>
<th>MODY 3</th>
<th>MODY 4</th>
<th>MODY 5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genetics</td>
<td>HNF-4α (20q13)</td>
<td>Glucokinase (7p15)</td>
<td>HNF-3a/3b (19p13)</td>
<td>IF-1 (13q12)</td>
</tr>
<tr>
<td>Prevalence</td>
<td>5%</td>
<td>5%</td>
<td>70%</td>
<td>&lt; 1%</td>
</tr>
<tr>
<td>Severity</td>
<td>Progressive IGT</td>
<td>Nondiabetic hypoglycemia</td>
<td>Progressive IGT</td>
<td>Progressive IGT</td>
</tr>
<tr>
<td>Onset</td>
<td>12-35 years</td>
<td>Birth</td>
<td>12-28 years</td>
<td>14-40 years</td>
</tr>
<tr>
<td>Microvascular Complications</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Treatment</td>
<td>Progressive need</td>
<td>Progression need</td>
<td>Progressive need</td>
<td>Progressive need</td>
</tr>
</tbody>
</table>

• Diabetic ketoacidosis (DKA) occurs when the body’s requirements for insulin are higher than the insulin available for use, resulting in lipolysis, ketogenesis, and ketoacidosis
• DKA is classically associated with Type 1 diabetes mellitus (DM), but can occur in any patient with diabetes
• Euglycemic DKA is defined by ketosis, metabolic acidosis, and a blood glucose level less than 200 mg/dL
• Euglycemic DKA is concerning as it can be easily misdiagnosed and mistreated
• Few cases have demonstrated a possible relationship between empagliflozin and euglycemic DKA
• Empagliflozin is a sodium glucose co-transporter (SGLT-2) inhibitor in the proximal convoluted tubule of the kidney to reduce reabsorbed filtered glucose, resulting in decreased serum glucose levels.

BACKGROUND

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