A Rare Case Of Metastatic Insulinoma In A Patient With Huntington’s Disease

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A Rare Case of Metastatic Insulinoma in a Patient with Huntington’s Disease
Olesya Petrenko DO, Brinton Clark MD

Case Presentation
A 49-year-old female with Huntington’s disease presents to the ED after being found unresponsive at her assisted-living facility.

History of Present Illness
- Hypoglycemic, glucose 21, responsive to dextrose. Refused admission
- Other symptoms: dizziness, tremors, nausea; attributed to her psychiatric condition.
- Symptoms re-occurred, admitted.
- No other medical history.
- Only medication: Zyprexa

Exam
- Afebrile, pulse 102, BP 112/69, RR 18, SpO2 100% RA
- General: Young adult female, anxious, frequent spastic, choreiform movements.
- Abdomen: No abdominal masses palpable.
- Neuro: Poor coordination.

Work up and Hospital Course
- Started on D10 drip
- Given psych history, concern for surreptitious use of insulin
- Persistently hypoglycemic for days → work up

Work Up
- Fasting insulin 163 ↑
- C-peptide 7.0 ↑
- Cortisol, TSH normal
- 72-hour fast: gold standard for diagnosis
  - see table
- Pancreas CT protocol:
  - 4 cm mass in pancreatic tail
  - With multiple hyper-vascular heterogeneously enhancing hepatic masses, largest 9 cm
- Pathology: metastatic well-differentiated neuroendocrine tumor grade 3, stage IV with bilar liver metastases.

Treatment
- NOT candidate for surgery
- Diazoxide: reduces insulin secretion
- Octreotide 200 mcg TID
- Y90 therasphere radio-embolization to left + right hepatic lobar tumors
- Continued on Octreotide injection q4 weeks lifelong

72-Hour Fasting Protocol Results

<table>
<thead>
<tr>
<th>Lab value</th>
<th>Result</th>
<th>Diagnosis of Insulinoma</th>
</tr>
</thead>
<tbody>
<tr>
<td>Glucose</td>
<td>23</td>
<td>&lt;45</td>
</tr>
<tr>
<td>Insulin</td>
<td>101 ↑↑</td>
<td>Inappropriately elevated</td>
</tr>
<tr>
<td>C-Peptide</td>
<td>4.1 ↑</td>
<td>Inappropriately elevated</td>
</tr>
<tr>
<td>Pro-insulin</td>
<td>782 ↑↑</td>
<td>&lt;8.0</td>
</tr>
<tr>
<td>Insulin/C-peptide ratio</td>
<td>0.25</td>
<td>&lt;1.0</td>
</tr>
<tr>
<td>Beta-hydroxybutyrate (ketones)</td>
<td>Undetected</td>
<td>&lt;2.7</td>
</tr>
</tbody>
</table>

CT Abdomen W/WO Contrast Findings

Insulinoma

Discussion
- **Insulinoma:** 1-4 cases/million/year; very rare.
- Causes hyperinsulinemic hypoglycemia
  - diaphoretis, tremors, personality changes, seizures.
- 90% benign
- **Malignant insulinomas:** extremely rare
  - Invade into soft tissue, lymph node, liver.
  - Unresectable; require targeted therapy.
- 5% of insulinomas associated with MEN1

DIAGNOSIS
- **Clinical:** Whipple’s triad
  1. Hypoglycemia; glucose <50 mg/dL
  2. Neuroglycopenic symptoms
  3. Relief with administering glucose

Gold Standard: 72-Hour Fasting Protocol
- Measure insulin, C-peptide, pro-insulin, ketones, glucose, ratio
- Detects 99% of insulinomas

Imaging: Abdominal CT, MRI, EUS

TREATMENT
- #1 Surgical resection: curative
- Other:
  - Injection of octreotide
  - EUS-aided alcohol ablation
  - Radio-frequency ablation or embolization
  - Targeted therapy: everolimus or sunitinib

Conclusion
- We present a 49 YO patient with severe persistent hypoglycemia believed to be from surreptitious use of insulin given her underlying psychiatric disease but ultimately diagnosed with a metastatic insulinoma.
- Diagnosis often delayed or missed;
  - symptoms attributed to psychiatric, cardiac, neurological disorders.
- Missed diagnosis can be fatal, deprives patient of curative treatment.
- Severe hypoglycemia not always due to medication overuse or misuse.
- Must evaluate for further causes of hypoglycemia such as insulinoma as this can save a patient’s life.

References