ADULT-ONSET NESIDIOLASTOSIS CAUSING HYPERINSULINEMIC HYPOGLYCEMIA: DIAGNOSIS AND TREATMENT CHALLENGE. A CASE REPORT

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Nesidioblastosis is defined as a diffuse proliferation of primitive pancreatic islet cells budding from ductal epithelium.1

A 41-year-old male operated for a duodenal ulcer (1999) was referred to our center via emergency room with suspicion of an insulinoma after an episode of loss of consciousness with a glycemia (G) of 24mg/dl. Current symptoms started 4 days prior to admission with dizziness, blurred vision, sweating, tremor in the upper limbs which resolved after ingestion of foods with high glycemic index.

Further questioning revealed 2 similar episodes in September 2012 investigated in a cardiology and respectively neurological service and one more in March 2012 during surgery for intestinal adhesion with G=18mg/dl, no further investigated.

At admission: altered general status with retrograde amnesia, dry skin and mucous membrane and deep tenderness in the epigastrum, BP=125/75mmHg, Pulse=65b/min.

<table>
<thead>
<tr>
<th>Laboratory findings</th>
<th>72-hour fast</th>
<th>72-hour fast (under 300mg HCH)</th>
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<tbody>
<tr>
<td>sO2 = 92.2%</td>
<td>Started 09:30</td>
<td>Started 08:00</td>
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<tr>
<td>Ca²⁺ = 1.03 mmol/l</td>
<td>Stopped: 12:00</td>
<td>Stopped 12:00</td>
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<tr>
<td>Cl = 110 mmol/l</td>
<td>G = 32 mg/dl</td>
<td>08:00: G = 150 mg/dl</td>
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<tr>
<td>Lac = 24 mg/dl</td>
<td>Plasma insulin= 86.3 μU/ml</td>
<td>10:00: G = 118 mg/dl</td>
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<td>G = 102 mg/dl</td>
<td>12:00: G = 121 mg/dl</td>
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<tr>
<td>PTH = 81mg/dl</td>
<td>G/I = 0.37</td>
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Abdominal US
- no visualization of the pancreas;
- bilateral renal microlithiasis.

Contrast CT and MRI
- Excluded renal microlithiasis;
- normal pancreas.

Endoscopic US
- pseudolobular aspect in the pancreas tail and body.

The patient was guided to a surgical clinic with dietetic recommendation and Phenytoin 200mg/day for insulinoma localization and surgical treatment. He was admitted into the surgical clinic after 3 weeks from the previous hospitalization with only 2 moderate hypoglycemic episodes under Phenytoin treatment.

Short Octreotide test
Debut:

- 08:00 G=60mg/dl; Serotonin serum = 75μg/l
- 100 ug s.c. Octreotide Chromogranin A = 53 μg/l
- 09:00 G=94mg/dl 5-hydroxy-indoleacetic acid = 3mg/24h
- 10:00 G=34mg/dl

Due to repeated hypoglycemic episodes after Phenytoin treatment was stopped, exploratory laparotomy was performed with intraoperative ultrasonography which exhibited two possible tumors in the pancreas tail and body. A subtotal splenopancreatectomy was performed. Microscopic view was consistent with nesidioblastosis and no tumor was found. The patient remains euglycemic ten months post-operatively suggesting that the source of insulin excess had been removed.

Conclusions
Nesidioblastosis:
- is a rare cause of adult hypoglycemia;
- pre-operative differentiation from insulinoma is difficult;
- suspected when imaging studies are negative.

References: