ADULT-ONSET NESIDOBLASTOSIS

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ADULT-ONSET NESIDIOBLASTOSIS

Objective:
1. Discuss how to diagnose Insulinoma
2. Understand the localization studies for Insulinoma
3. Adult-onset Nesidioblastosis
Chief complain: light headed/sweating/palpitation with fasting

HPI:
- 70 yrs old M with neuroglycopenic sx for 5 years.
  - W/U found to have Low serum glucose/Elevated C-peptide/Insulin.
- PMH:
  - HL, HTN, PCKD, 2\textsuperscript{nd} hyperparathyroidism, OSA.
- PSH:
  - Tonsillectomy, AV fistula.
- Meds:
  - Sevelamer, Octreotide, Simvastatin, Cinacalcet, Alprazolam.
• Physical Exam:
  – Unremarkable
• Lab Ix:
  – C-peptide: >13.pmol/ml (Ref: 0.3- 2.35)
  – Pro-insulin: 310 (Ref: 3-20)
  – Insulin: 110 mcu/ml (Ref: 2.6-24.9)
  – Glucagon: 84
  – Sulfonylurea panel negative
• No endosonographic evidence of pancreatic mass
CALCIUM STIMULATION TEST
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<th>Draw Time</th>
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<td>Splenic Artery (SPA4)</td>
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• Operation 10/18/17
  – Robotic splenic sparing distal pancreatectomy.

• Post-operative course:
  – Uneventful d/c to rehab HD # 9
  – Lab: C-peptide 2.25 pmol/ml
GA
INSULINOMA

• Neuroendocrine tumor
  • Excessive secretion of insulin
  • Sporadic vs syndromic (MEN)
  • Sx Whipple’s triad
    – Hypoglycemia during fastin/exercis
    – Blood sugar < 45 mg/dl
    – Sx improved with administration of glucose

• Diagnosis:
  – Low serum glucose
  – Elevated Serum Insulin/pro-insulin/c-peptide
  – Negative sulfanyl panel test
INSULINOMA
INSULINOMA
Nesidioblastosis

- Hyperinsulinoma hypoglycemia
- First described by George Laidlaw in 1938
  - Greek words “nesidion” islet and “blastos” builder
    - Neoformation of Langerhans islets from pancreatic duct epithelium
  - Most common cause of neonatal hyperinsulinemia

- Adult-onset Nesidoblastosis
  - First described in 1975 by Sandler
  - 0.5% to 7% of all cases of hyperinsulinemic hypoglycemia
  - 16% at the University of Chicago
NESIDIOLASTOSIS

• Aetiology:
  – Unknown
  – 2 genetic alteration SUR1 and Kir 6.2 on Chromosome 11
    • Code for Potassium channel on Beta cell membrane
  – Bariatric population after “Roux-en-Y gastric bypass”

• Symptoms:
  – Similar to Insulinoma; neuroglycopenic/adrenergic sx with fasting.

• Diagnosis of adult-onset Nesidioblastosis:
  – Hyperinsulinoma
  – Exclusion of insulinoma (Imaging, Macroscopic, Microscopic, IHC)
  – Pathological features of Nesidioblastosis
NESIDIOBLASTOSIS

Normal pancreatic islet

Hypertrophic islet
NESIDIOBLASTOSIS

Islet cell pleomorphism and hyperchromatic islet cell nuclei

Ductal Insular complex
NESSIDIOBLASTOSIS

• Management
  – Surgical:
    • Near total pancreatectomy (90-95%)
      – Insulin dependent DM 40%.
    • Distal pancreatectomy (60-80%)
  – Medical treatment
    • Diazoxide
    • Somatostatin
    • Calcium channel-blockers
NESIDIOBLASTOSIS
NESIDIOBLASTOSIS

Questions?