Tumor Biology
ENDOCRINE NEOPLASIA CASE REPORTS II
Undiagnosed Chronic Eczema as a Presentation of Glucagonoma in MEN 1 Syndrome
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MON-912
Background: Glucagonomas are pancreatic tumors arising from the islets cell of Langerhans that over-secrete glucagon. Necrotic migratory erythema (NME) is an important feature for the recognition of glucagonomas. Glucagonomas occurring in MEN1 is infrequent and seen in less than 3% of all glucagonomas.

Clinical Case: A 51-year-old male presented to the clinic multiple visits for rash affecting the legs and genital area of two months. His medical history include type 2 DM and HT. The rash was attributed to subacute eczema and treated with topical steroids but showed no improvement. The skin eruption initially appeared on lower extremities progressed to trunk, and face. The skin lesions were associated with weight loss and stomatitis. On physical examination, skin showed ill-defined erythematous plaque exhibiting annular pattern, scale, and erosion on all extremities and periorial area. When the skin lesions healed, the new cutaneous eruptions occurred. Laboratory testing revealed plasma glucagon decreased to 425 pg/dL, patient had complete resolution of the cutaneous lesions.

Conclusion:

Adrenal
ADRENAL CASE REPORTS II
Case Series of ACTH-Secreting Pheochromocytoma: Diagnosis and Management
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SUN-168
INTRO
ACTH-secreting pheochromocytoma is rare, posing challenges in diagnosis and management. Here we report two cases with different presentations and pre-operative approaches.

CASES
Patient 1 A 59 yo female who presented for basal cell carcinoma resection experienced hypertensive crisis intra-op and NSTEMI due to stress cardiomyopathy. The patient reported spells of palpitations, diaphoresis, tremors and pallor since 2015. She had elevated plasma metanephrine (M) 19.4 (0.0-49.9nmol/L), normetanephrine (NM) 24.7 (0.0-0.89nmol/L), 24-hour urine M 18711 (30-180mcg) and NM 11597 (128-484mcg). Imaging demonstrated a 5.6 x 5.8cm left adrenal mass. After pre-operative alpha and beta blockade, patient underwent left adrenalectomy. Pre-op 24-hour urine cortisol was checked as part of secondary HTN evaluation and found to be elevated at 219 (3.5-45mcg). She did not have any features of Cushing’s and this was initially felt to be an appropriate stress response. Due to persistent post-op hypotension and a low 10PM random cortisol level of 3.4mcg/dL, which was inappropriate given patient’s clinical status, hydrocortisone was subsequently added to the regimen with rapid and marked improvement. For patient 2 a 48 yo female was admitted for hypertensive urgency with symptoms of palpitations, facial and abdominal swelling, easy bruising and proximal muscle weakness. Her labs showed elevated 24-hour urine epinephrine 115 (0-20mcg), norepinephrine 366 (0-135mcg), plasma M 288 (0-62pg/mL) and NM 475 (0-145pg/mL), and an AM cortisol was 47.9mcg/dL. Aldosterone and renin levels were <1ng/dL and 0.775ng/mL/hr. Imaging revealed a 3.4 x 3.1cm right adrenal mass with portal venous phase of 53 HU and no washout. AM cortisol level post 1mg dexamethasone was 62.6mcg/dL with an ACTH level of 222pg/mL. An ACTH-producing pheochromocytoma was suspected. Despite progressive uptitration of alpha and beta blockade, her BP remained poorly controlled. Her blood glucose also increased to 300s range. Ketoconazole 400mg BID was subsequently added to the regimen with rapid and marked improvement in BP and glycemic control. She underwent appropriate given patient’s clinical status, hydrocortisone was subsequently added to the regimen with rapid and marked improvement in BP and glycemic control. She underwent appropriate given patient’s clinical status, hydrocortisone was subsequently added to the regimen with rapid and marked improvement in BP and glycemic control. She underwent
right adrenalectomy. Post-op plasma M, NM and ACTH were <0.20nmol/L, 0.36nmol/L and <5pg/mL.

**DISCUSSION**
Within the past 6 months, we have seen 2 cases of ACTH-secreting pheochromocytoma with different clinical symptoms and pre-op courses. Ectopic ACTH secretion may not cause classic Cushingoid features as in patient 1. Failure to recognize ectopic ACTH pre-op can lead to post-op complications such as AI. Patient 2 demonstrates that treatment of the hypercortisolism may be necessary in order to adequately control BP and glucose levels prior to surgery.

**Adipose Tissue, Appetite, and Obesity**
**MECHANISMS AND TREATMENT OF OBESITY IN HUMANS**

**Potential Role of Mutations in TBX3 in Human Weight Regulation**
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**OR33-06**
**Introduction:** Tbx3 has been shown to play a role in the terminal specification of hypothalamic melanocortin neurons during neonatal development & in maintaining the plasticity of their peptidergic role in adulthood in animal experiments (1). The absence of humans with biallelic mutations in TBX3 & the conservation of the critical domains across species emphasizes its essential role in life. Heterozygous mutations in humans have been associated with unlar mammary syndrome (UMS) with a spectrum of phenotype including obesity. Based on these observations, we hypothesized that heterozygous mutations in the conserved regions of TBX3 may play an important role in the weight regulation pathway in humans.

**Methods:** The Genetics of Early Childhood Obesity (GECO) study enrolls children with severe (BMI > 120% of 95th %tile of CDC reference) early onset (< 6 years) obesity. Whole exome sequencing (WES) was performed in a subset of proband-parent trios. Peripheral mononuclear cells (PBMCs) from selected families were collected to generate induced pluripotent stem cells using non-integrating Sendai virus. Differentiation into disease-relevant hypothalamic neurons suggest that a decrease in melanocortin signaling possibly explaining the phenotype in this family.

**Conclusions:** Mutations in TBX3 in humans may have a role as a monogenic cause of obesity and disease-relevant hypothalamic stem cells can serve as models to study them. Ref: 1) Quarta et al. Nat Metab 2019, 1(2), 222-35; 2) Wang et al. JCI, 125(2): 796-808.

**Diabetes Mellitus and Glucose Metabolism**

**TYPE 2 DIABETES MELLITUS**

**Effect of Night Shifts on Glycemic Variability in Patients with Type 2 Diabetes**
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**SUN-696**

Effect of night shifts on glycemic variability in patients with type 2 diabetes.
The analysis of indicators of carbohydrate metabolism and variability of glycemia in patients with type 2 diabetes mellitus working in night shifts was carried out. As model patients with impaired circadian rhythm, the study included 34 patients, railway transport drivers, with shift mode and the presence of night shifts, with work experience of more than 5 years, the duration of type 2 diabetes mellitus (DM2) from 1 to 7 years, who are on oral therapy with hypoglycemic drugs. Simulation of different working conditions (day-night) was carried out in the simulator “driver’s cabin”. All patients underwent a study of the main indicators of carbohydrate metabolism (fasting glycemia, postprandial glycemia, glycated hemoglobin (HbA1c)), as well as continuous daily glucose monitoring (CGMS) using Medtronic MiniMed iPro2 system (from 3 to 7 days).

Target glycemic levels were not achieved: fasting glycemia was 6.98±1.41 mmol/l; postprandial glycemia was 9.57±1.65 mmol/l; HbA1c was 7.23±1.62%.

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The period of hyperglycemia (glucose value above 7.8 mmol/l) according to the results of CGMS was 43.5% (min 19-max 56). The duration of hypoglycemic States in...