calvarium, long bones, ribs, and compression fractures of T8, T10, and T12 vertebral bodies. A 1.5 cm left adrenal nodule was also noted. She was treated with bortezemib, cyclophosphamide, lenalidomide, and dexamethasone, and clinically improved. Four months after initial presentation, patient was to undergo chemotherapy with melphalan and autologous stem cell transplantation. Further imaging was performed, and CT Chest revealed an enlarging left adrenal mass measuring 3.0 x 3.2 cm with increased attenuation at 37 Hounsfield units and lobulated borders with no invasion of adjacent structures. The endocrinology team was consulted for evaluation of the adrenal mass, as patient's disease from multiple myeloma was presumed to be in remission. Differential for the adrenal mass included pheochromocytoma, primary adrenal carcinoma, and metastatic disease from multiple myeloma vs other primary. Plasma fractionated metanephrines and DHEAS were within normal range. CT guided core biopsy was performed. Cytology revealed diffuse infiltrate of atypical plasma cells. IHC studies were positive for CD138, CD56, and showed lambda light chain restriction. Cyclin D1 stain was negative, consistent with plasmacytoma. Clinically, the patient started developing progressive disease, including cutaneous plasmacytomas, and she was treated with additional chemotherapy.
Conclusion:
Adrenal plasmacytoma is extremely rare, and in a patient with multiple myeloma, should be considered in the differential diagnosis of a rapidly enlarging adrenal mass with high CT attenuation. CT guided biopsy is the definitive test for diagnosis.
References:
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Reproductive Endocrinology

CLINICAL STUDIES IN FEMALE REPRODUCTION II

Beyond PCOS - Ovarian Neoplasms Presenting with Hirsutism and Virilization

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SUN-011

Background: PCOS is the most common cause of hirsutism in women of reproductive age. The presence of virilization in addition to hirsutism should alert to the possibility of less common causes of hyper-androgenization (HA) in this population including otherwise uncommon functional ovarian neoplasms (FON). We present 3 cases of women initially thought to have PCOS in whom virilization was the prime clue to the correct diagnosis of FON. **Clinical Case series:** Case 1 is a 40yr old woman with obesity and dysmetabolic syndrome referred for hirsutism presumed due to PCOS. She had noted symptoms over 2–3 yrs with amenorrhea and associated infertility. Examination revealed marked hirsutism and virilization with Ferriman-Galleway score (FGS) of 20. Lab tests confirmed marked male range HA. Multiple imaging tests revealed no adrenal or ovarian mass lesions. FDG-PET scan finally revealed a left ovarian focus for which she has left oophorectomy that revealed a 1cm Leydig cell tumor, Her HA resolved post-op and spontaneous periods resumed. Case 2 is a 45yr old woman referred with possible PCOS who had 5 mth history of progressive hirsutism and generalized hypertrichosis, dull lower abdominal pain and amenorrhea. Examination revealed marked hirsutism with generalized hypertrichosis and virilization. FGS was 25 and clitoral index was 935mm2. Lab tests confirmed marked male range HA and abdominopelvic imaging show no adrenal lesions but a 5.2cm left ovarian mass. Left salpingo-oophorectomy revealed a steroid cell tumor and postoperatively her androgen levels normalized. Case 3 is 37 yr old woman with SLE and obesity with prior gastric bypass referred with presumed PCOS but presenting with 1 yr history of progressive hirsutism. She was initially thought to have non classical CAH and treated with oral glucocorticoids with no symptom improvement. Examination revealed marked hirsutism, virilization with elevated FGS and clitoromegaly. Lab tests showed marked male range HA but multiple imaging studies revealed no apparent adrenal or ovarian lesions. Patient had no fertility interests and so had elective total hysterectomy and bilateral salpingo-oophorectomy. Histopathology revealed a 2.5cm left ovarian Leydig cell tumor not apparent at surgery and post op her androgen levels normalized. **Conclusion:** The distinction between PCOS which is ubiquitous and FON which is rare hinges on careful history and examination. Rapid onset hirsutism with virilization should prompt suspicion of FON. Marked male range HA (total serum testosterone >250ng/dl) is another "red flag" finding. Persistent radiologic search for such lesions should continue as they may not be immediately apparent on routine abdominopelvic imaging.

Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY

A Rare Case of IgG4-Related Hypophysitis

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SUN-273

Introduction:
Hypophysitis is an acute or chronic inflammation of the pituitary gland and is an important diagnostic consideration in a patient with a sellar lesion. The annual incidence of hypophysitis is estimated to be 1 in 7–9 million and it accounts for approximately 0.4% of pituitary surgery cases. The following highlights a rare case of isolated IgG4-related hypophysitis
Clinical Case:
A 63-year-old Caucasian female presented with sudden onset of diplopia and decreased visual acuity. This was associated with a 3-month history of headaches and 5-lbs weight loss. Past medical history was significant for hypertension and a 1.5cm sellar/suprasellar mass incidentally discovered during the work-up for persistent headaches 1-month prior. Initial

anterior pituitary hormone evaluation was normal and the patient was scheduled for endoscopic endonasal resection of a presumed non-functioning pituitary adenoma. Family history was negative for pituitary tumors or hyperparathyroidism. Physical examination was notable for medial deviation of her left eye but neurologic examination was otherwise normal. Laboratory studies were notable for a normal TSH [1.769 uIu/ml (normal: 0.3–5.0)] and low free T4 [0.44 ng/ml (normal: 0.89–1.78)] consistent with central hypothyroidism; an inappropriately normal FSH for a postmenopausal woman [5.6 mIu/ml (normal: 0.3–10.5), and a normal prolactin level [16 ng/ml (0.6–20)]. An 8am cortisol was low at 2mcg/dL (5–21) with an ACTH level of 10 pg/mL (9–46). IGF-1 was normal at 89 ng/mL (41–279). Repeat pituitary MRI imaging demonstrated a homogenously enhancing sellar/suprasellar mass measuring 3.8 cm with displacement of the optic chiasm. Serum IgG4 levels were normal. The patient was started on 50mg IV hydrocortisone every 8 hours for central adrenal insufficiency and levothyroxine 88 mcg daily for central hypothyroidism and underwent an endoscopic endonasal biopsy of the lesion. Surgical pathology was notable for plasma cell-rich lymphohistiocytic hypophysitis and IgG4 plasma cells constituted >40% of the total plasma cell population. The patient subsequently received 1g of rituximab and repeat imaging one week later showed marked improvement in the size and extent of the lesion. The patient was discharged on prednisone and levothyroxine and received a second dose of rituximab at follow-up. The patient reports a decrease in the frequency of her headaches but continues to endorse diplopia.
Conclusion:
IgG4-related hypophysitis typically presents as part of a multifocal systemic process. This case highlights a rare entity of IgG4-related hypophysitis without other features of systemic disease and with normal serum levels of IgG4. Although glucocorticoids are universally regarded as the first line of therapy, an immunosuppressive agent or B-cell depletion therapy such as Rituximab may improve remission and decrease the risk of relapse.

Diabetes Mellitus and Glucose Metabolism

CLINICAL AND TRANSLATIONAL STUDIES IN DIABETES

Hypoglycemia Following OGTT Is More Frequent and Pronounced in CF Compared with Controls

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MON-660

Glucose homeostasis is often abnormal in people with pancreatic insufficient cystic fibrosis (PI-CF). This dysfunction is viewed on a continuum from "normal" glucose tolerance to cystic fibrosis-related diabetes (CFRD), and may also include postprandial and oral glucose tolerance test (OGTT)-related hypoglycemia. This study aimed to delineate the mechanism(s) underlying OGTT-related hypoglycemia. We compared extended OGTT with frequent blood sampling of glucose and insulin in adolescents and young adults with PI-CF [CF(+)] to historical data from a healthy cohort [CF(-)]. We hypothesized that the subset of CF(+) with hypoglycemia would demonstrate 1-hour glucose ≥ 155 mg/dL and impaired early phase insulin secretion (insulin secretion within first 30 min of OGTT). Hypoglycemia [hypo(+)] was defined as plasma glucose <65 mg/dL and was used to assign subjects to exposure groups. We restricted analyses to 180 minutes given available control data. Glucose and insulin incremental areas under the curve (Glc-AUC; Ins-AUC) for 30-minute intervals were calculated. One-hour glucose, nadir glucose, Glc-$AUC_{0\text{-}30}$, and Ins-$AUC_{0\text{-}30}$ and were compared between CF(+) and CF(-) subjects using Student's t-test or Wilcoxon Rank Sum depending upon normality. Participants were 60.5% male, age: 25.4±4.8 years, with BMI-Z: 0.06±0.96kg/m^2 [no differences for CF(+) vs CF(-)]. FEV1%-predicted for CF(+) was 83±21. 69.6% of CF(+) participants self-reported prior episodes of hypoglycemia, 68.7% of whom reported confirmation via glucometer. Hypoglycemia occurred by 180 minutes [hypo(+)] in 15/23 (65%) CF(+) and 5/15 (33.3%) CF(-) subjects (p=0.028). For hypo(+), nadir glucose occurred on average at 180 minutes for both CF(+) and (-). Hypo(+) CF(+) had higher mean 1-hour glucose (197±49mg/dL vs 134±66mg/dL, p=0.035), lower mean glucose nadir (48±7mg/dL vs 61±4mg/dL, p<0.01), and lower early-phase insulin secretion (Ins-$AUC_{0\text{-}30}$: 263±168 versus 650±275 μU/mL, p<0.01) than hypo(+) CF(-). There was no difference in Glc-$AUC_{0\text{-}30}$ for hypo(+) CF(+) vs CF(-). Hypoglycemia is frequent in CF, and is associated with early glucose dysregulation (elevated 1-hour glucose) and compromised early-phase insulin secretion compared to controls with presumed non-pathologic reactive hypoglycemia. The mechanism of hypoglycemia in CF appears to be different than that seen in healthy individuals, and its association with progression to CFRD warrants further evaluation.

Bone and Mineral Metabolism

BONE AND MINERAL CASE REPORTS I

Novel CASR Gene Mutation as a Cause of Familial Isolated Primary Hyperparathyroidism

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SAT-370

Background: Primary hyperparathyroidism (pHT) is one of the most common causes of hypercalcemia. About 10%