Patients carrying a germline mutation were younger than patients with no mutations (40.7 yo (20 - 67) vs. 49.6 yo (11 - 80)) and had a higher prevalence of metastatic tumors (26.6% vs. 20.4%). The prevalence of germline mutations was 43.3% (26/60) in PGLs and 14.7% (11/75) in PHEOs. In the 26 mutated PGLs, there were 13 SDHC (50.0%), 6 SDHB (23.1%), 4 SDHD (15.4%), 2 SDHA (7.7%) and 1 FH (3.8%) mutations. The recurrent pathogenic SDHC c.397C>T (p.Arg133*) mutation was found in 12 out of the 13 SDHC mutations reflecting the presence of a funder effect in the French Canadian population. In the 11 mutated PHEOs, there were 3 MAX (27.3%), 3 VHL (27.3%), 2 RET (18.2%), 1 SDHB (9.1%), 1 NF1 (9.1%), 1 FH (9.1%) mutations.

From 2015- 2019, we proposed NGS assay with the multigene panel to 12 patients (9 PHEOS and 3 PGLs) for whom the initial genetic test was negative. Novel germline mutations were found in 4 (33.3%) of these patients, representing 10.8% (4/37) of the mutation-carriers. Mutations were found in 2/9 PHEOs; a 28 yo female with bilateral PHEOs (MAX (deletion exon 1 and 2)) and a 33 yo male with malignant PHEO (MAX (c.3G>A)), and in 2/3 PGLs; a 31 yo woman with metastatic abdominal PGL (SDHA (c.985C>T)) and a 59 yo woman with a thoracic PGL (SDHA (c.1432_1432 + 1del)).

Variants of uncertain significance (VUS) were identified in 7/60 PGLs (11.6%) and 5/75 PHEOs (6.7%) but the significance of these variants remains to be determined.

Conclusion: In our cohort, the prevalence of germline mutations was of 44.3% in apparently sporadic PGLs and 14.7% in PHEOs. Genetic re-evaluation overtime using multigene sequencing by NGS assay in a subgroup of patients led to an increase of mutation rate in PHEOs and PGLs with the identification of germline MAX and SDHA mutations.

Reproductive Endocrinology

MALE REPRODUCTIVE HEALTH - FROM HORMONES TO GAMETES

Effect of Weight on Serum Testosterone with Subcutaneous Testosterone Enanthate in Men with Testosterone Deficiency
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SAT-037
Background: In men, obesity is often associated with low testosterone (T) levels, but information is limited as to how body weight affects the pharmacokinetic profile or dosing of testosterone therapy (TTh) in men with T deficiency. Historically, men with body mass index (BMI) >32.4 kg/m² required higher doses of T 2% gel to achieve physiological T levels than men with BMI <29.1 or 29.2–32.4 kg/m².(1) In a phase 3 trial (N=150) of subcutaneous (SC) testosterone enanthate (TE) administered weekly, concentration-guided dosing raised T levels to within physiological range in 92.7% of patients.(2) Here, we report a post hoc analysis evaluating the association between body weight and serum T levels attained with SC TE. Methods: SC TE was evaluated in an open-label, single-arm, dose-blinded, 52-week phase 3 trial (NCT02159469). Patients self-administered 75 mg SC TE weekly during the titration phase; blinded dose-adjustments in 25 mg increments occurred at pre-defined time points beyond the sixth dose. The primary endpoint of this study was the percentage of patients achieving an average serum T concentration (Cavg0-168h) of 300 to 1,100 ng/dL at week 12. For this post hoc analysis, a linear regression model with weight and dose as independent variables was used to assess differences in mean minimum T concentration (Cmin) and Cavg0-168h, at week 12. Results: For this analysis, 137 patients were included. Doses were 50 mg (n=25), 75 mg (n=93), and 100 mg (n=19). The mean weight was 84.4 kg, 102.2 kg, and 112.0 kg for the 50 mg, 75 mg, and 100 mg dose groups, respectively (range, 49.9–146.5 kg). The dose-normalized T Cmin was 9.2 ng/dL, 5.7 ng/dL, and 4.3 ng/dL per 1 mg of SC TE for the 50 mg, 75 mg, and 100 mg groups, respectively. The dose-normalized T Cavg0-168h was 12.0 mg/dL, 7.2 mg/dL, and 5.7 mg/dL per 1 mg of SC TE. In an overall linear regression model, 48.2% (P<0.0001) and 55.0% (P<0.0001) of the total variance in Cmin and Cavg0-168h, respectively, can be predicted from the independent weight and dose variables. Conclusion: Our results show an inverse relationship between body weight and T exposure. Men with higher mean body weights required higher doses of SC TE to achieve physiologic T levels compared with men with lower mean body weights. The available doses provide effective options to reach target exposures. These findings highlight the impact of weight and dose selection on SC TE exposure. References: (1) Dobbs et al., J Sex Med 2014;11:857–864; (2) Kaminetsky et al., J Urol. 2019;201:587–94.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

Adrenal Plasmacytoma in Multiple Myeloma Patient-An Unusual Presentation
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SUN-928
Title: Adrenal Plasmacytoma in Multiple Myeloma Patient: an unusual presentation
Introduction: Extramedullary plasmacytomas are plasma cell tumors that arise outside of the bone marrow. They are solitary lesions, and are most often located in the head and neck region, mainly in the upper aerodigestive tract. However, involvement of adrenal gland is extremely rare, with only nine case reports published to date. A mass in the adrenal gland carries a broad differential, and identification is important, as diagnosis drives treatment options. CT imaging with attenuation, timing of contrast medium washout, size, and shape, with biopsy is necessary for diagnosis of a high Hounsfield unit mass. Ruling out pheochromocytoma before biopsy of the adrenal glands is crucial.
Clinical Case: A 64-year-old female was diagnosed with multiple myeloma after presenting with back pain and altered mental status. Imaging revealed diffuse lytic lesions in clavicles, pelvis,
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 virilism 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-Gallwey 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