HU, with an absolute and relative washout of 65% and 44% respectively, not consistent with a benign adenoma. Patient was placed on a high salt diet along with fluids and doxasozin. Once appropriately alpha blocked, metoprolol was added prior to surgery.
Clinical lessons:
In the setting of labile BP, one should keep a high clinical suspicion for pheochromocytoma. Orthostatic hypotension here was related to epinephrine predominance pattern of secretion and volume depletion. Steroids should be avoided in patients with known pheochromocytoma or in those with an adrenal adenoma without a negative biochemical workup for a pheochromocytoma as it could precipitate an adrenergic crisis possibly by inducing catecholamine synthesis and release and sensitizing target organs to the effects of catecholamines.
Reference:
1. Rosas AL, et al. Pheochromocytoma crisis induced by glucocorticoids: a report of four cases and review of the literature. European Journal of Endocrinology (2008) 158 423–429.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

An Unusual Presentation of Insulinomas-Profound Hypophosphatemia

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SUN-929

Background: Insulin induces intracellular shift of phosphorus, causing hypophosphatemia. This is mild, transient and quiescent in normal persons. Persistent hyperinsulinemia in patients with depleted phosphorus stores can cause profound symptomatic hypophosphatemia. **Clinical Case:** A 36yr old man with MEN 1 presented in follow-up. His original manifestation of MEN 1 was familial primary hyperparathyroidism (pHPT) for which he had parathyroidectomy in 2009. At one of his endocrine clinic visits, he was noted to have asymptomatic hypophosphatemia with serum phosphorus of 2.4 (2.7–4.5 mg/dL) and mildly elevated PTH of 89 (9–77 pg/mL). This was presumed to be due to mild persistent/recurrent pHPT, and he was advised to increase dietary phosphorus intake. About a year later, he presented to the emergency room (ER) with palpitations and marked asthenia. Serum phosphorus was 1.0. He was treated with intravenous phosphorus and discharged on oral phosphorus replacement with advice to increase his dietary phosphate intake. Despite this, he continued to have recurrent spells of dizziness, weakness, palpitations, and marked hypophosphatemia several requiring repeat ER visits. His degree of mild pHPT remained stable and imaging studies to identify a possible cause for tumor induced osteomalacia with phosphate wasting was unrevealing. He volunteered at a follow up visit that he had a long history of low blood sugar spells (confirmed by fingerstick) that he had learnt to manage with frequent carbohydrate rich snacks. This prompted the consideration of a possible insulinoma as the cause for his persistent hypophosphatemia. Despite significant hypoglycemia on an abbreviated supervised fast, an OGTT and mixed meal test showed profound severe insulin mediated hypoglycemia. CGMS also showed multiple extended early morning severe hypoglycemic spells <50mg/dl over a 2 wk period. Multiple imaging tests were negative but a Gallium Dotatate scan revealed multiple areas of uptake in his pancreas. Endoscopic ultrasound of the pancreas demonstrated multiple masses in the pancreas: one 7 x 5 cm mass in the body and one 4 x 4 cm mass near the neck of the pancreas. Fine needle aspiration biopsy revealed well-differentiated neuroendocrine tumors. He had distal pancreatectomy and tumor enucleation of 5 tumors identified on intraoperative sonography. Histopathology confirmed multiple neuroendocrine tumors, with immunostaining for insulin. He has had no further episodes of hypophosphatemia since despite being off oral phosphate supplements. **Conclusion:** Profound hypophosphatemia can be the dominant presentation of persistent hyperinsulinemia seen in patients with insulinoma. Insulinomas and other causes of persistent hyperinsulinemia need to be considered in the differential diagnosis of persistent/recurrent hypophosphatemia.

Adrenal

ADRENAL - HYPERTENSION

Genetic Spectrum Of A Canadian Cohort Of Sporadic Pheochromocytomas And Paragangliomas: Higher Prevalence Of Germline Mutations In PGL And NGS Assay With A Multigene Panel Increases The Mutation Rate

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MON-212

Background: Pheochromocytomas (PHEOs) and paragangliomas (PGLs) (PPGLs) are rare tumors with a high heritability. The prevalence of germline mutations in sporadic PPGLs varies depending of series.
Objective: To determine the prevalence and spectrum of germline mutations in our cohort of patients with apparently sporadic PPGLs.
Method: We retrospectively reviewed the charts of patients with sporadic pathology-confirmed PPGLs who underwent genetic testing after genetic counselling at our Quaternary center from 2005–2019. Genetic analysis included sequential gene sequencing by Sanger method from 2005–2014 (n = 89) and a multigene sequencing by NGS with a panel (14 susceptibility genes for PPGLs) from 2015–2019 (n = 34). Some patients underwent both (n = 12).
Results: Among 230 patients that were treated for PPGLs from 2005- 2019, 135 patients underwent genetic testing (77 females; 58 males and 77.8% French Canadians). There were 60 PGLs (29 head and neck, 21 abdominal and 10 thoracic) and 75 PHEOs, 4 being bilateral. The prevalence of pathogenic germline mutations was 27.4% (37/135).

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/m^2 required higher doses of T 2% gel to achieve physiological T levels than men with BMI ≤29.1 or 29.2–32.4 kg/m^2.(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 ($C_{avg0\text{-}168h}$) 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 (C_{min}) and $C_{avg0\text{-}168h}$ 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 C_{min} 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 $C_{avg0\text{-}168h}$ was 12.0 ng/dL, 7.2 ng/dL, and 5.7 ng/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 C_{min} and $C_{avg0\text{-}168h}$, 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) Dobs 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,