database, clinician referrals, patient groups, and social media. Interested participants were screened for eligibility prior to completing the GO-QOL. Subjects were also questioned about TED-related signs, symptoms, and treatments and underwent a cognitive interview following GO-QOL completion.

**Results:** Thirteen TED patients completed the assessments (mean age = 44.8 ± 11.5 years, range: 26-67); all were female. Mean TED duration was 4.6 ± 5.5 years (range: 0.4-20.7). Twelve patients (92.3%) had Graves’ disease and one had Hashimoto’s thyroiditis. Descriptions of how TED signs and symptoms impacted quality of life were consistent with GO-QOL items, and qualitative interviews indicated that patients found the GO-QOL content relevant and complete. Responses indicated that minor wording changes may be needed to account for US cultural and language conventions and prevent confusion (specifically related to a bicycling question [12/13 reported not regularly riding a bike before TED symptom onset]). Visual functioning impacts most commonly-reported during the interview were difficulty driving a motor vehicle (92% of participants), difficulty with electronic screens (e.g., televisions, smart phones, and computers; 77%), difficulty moving around outdoors (including issues with light sensitivity, uneven surfaces, and depth perception; 69%), and difficulty doing hobbies or pastimes (69%). Emotional/psychological impacts frequently reported by participants were change in appearance (92%), depression and anxiety (including fear and worry; 77%), and frustration and anger (including moodiness; 69%). Negative reactions from others (staring, asking questions), social impacts and isolation, and lack of self-confidence and embarrassment were also reported (each 62%).

**Conclusion:** This analysis of US patient interviews offered strong support for GO-QOL content validity. Therefore, the GO-QOL is appropriate to quantify TED-related QOL impact in a US population. However, a few slight wording modifications may be needed for future optimal use in the US. Reference: Terwee CB. Br J Ophthalmol 1998;82:773-779

**Adrenal**

**ADRENAL CASE REPORTS I**

**Paraganglioma in Two Unrelated Kinyarwanda-Speaking Patients in Anatomically Distinct Sites**

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**SAT-217**

**Introduction:** Capable of generating excess catecholamines, untreated extra-adrenal paragangliomas (PGL) result in severe cardiovascular morbidity and mortality. Increasingly, a hereditary basis can be identified to underlie PGLs, though such data is largely absent in non-Caucasian populations.

**Case 1:** A 43 yr. old Kinyarwanda-speaking woman from DR Congo presented with left lower extremity edema and hypertension, with blood pressure of 154/86 while on spironolactone, HCTZ and furosemide. Ultrasound was negative for a DVT; abdominal CT revealed a 3 cm necrotic mass, inferior to the duodenum and abutting the IVC and aorta, as well as 2 bladder wall lesions. EUS-guided FNA revealed a keratin-negative neuroendocrine tumor. Urinary 24hr norepinephrine (NE) was high at 185 mcg [15–80]. Urinary 24hr normetanephrine (NM) was high at 1404 mcg [119-451]; hypertensive <900]. MIBG scan confirmed avidity in the aortocaval mass. Despite lack of bladder uptake on MIBG, pathology similarly pointed to PGL. Surgery included excision of bladder, pelvic nodes, uterus, and aortocaval tumor. Post-op, urinary 24hr NE was 18 mcg and NM was 297 mcg, both normal. One year later, MIBG/SPECT and CT of the abdomen were negative for recurrence. A GeneDx panel of 12 PGL/PCC mutations was negative.

**Case 2:** An unrelated 41 yr. old Kinyarwanda-speaking woman from Rwanda, with prior history of preeclampsia and multiple miscarriages, presented with palpitations, headaches and hypertension. Echo showed a 4 cm mass posterior to the left atrium; the mass was 18F-FDG PET-avid. Video-assisted thoracoscopic was performed yet the tumor’s vascularity precluded a biopsy. Biopsy of mediastinal mass after performing thoracotomy was consistent with PGL. Plasma NM was high at 7.1 nmol/l (<0.90), consistent with PGL and she underwent complete removal of the tumor. Testing for SDHB mutation was negative. Symptoms resolved and antihypertensives were discontinued. Follow-up plasma NM was 0.55 nmol/l 1-year post op and remained normal for six years of follow-up.

**Discussion:** Less than 10% of PGLs are known to involve the mediastinum or bladder (1). In familial PGL, the most commonly identified non-syndromic mutations involve SDHD, SDHAF2, SDHB, SDHD, SDHC, VHL, and MAX. Tumorigenesis in a sizable fraction of PGLs is not well understood.

**Conclusion:** We present two cases of extra-adrenal PGL, both exhibiting similar age, sex and geographic ancestry. Our cases raise questions that require active investigation regarding additional environmental and/or genetic factors which might predispose to PGLs in uncommon anatomic sites.

**References:** (1) Erickson D et al. J Clin Endocrinol Metab, 2001 (2) Martins et al. Int J of Endocrinol, 2014

**Bone and Mineral Metabolism**

**BONE AND MINERAL CASE REPORTS II**

**Late Diagnosis in Adult Form of Hypophosphatasia**

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**MON-361**

**INTRODUCTION/BACKGROUND:** Hypophosphatasia is a rare inborn error of metabolism that presents with important foot and thigh pain due to stress fractures. The diagnosis of the adult form is routinely neglected, even though it presents symptomatic and with persistent low serum alkaline phosphatase (ALP).

**CLINICAL CASE (DIAGNOSTIC EVALUATION, TRATMANET AND FUP):** A 43-year-old amateur athlete woman presented with pain in the right femur without any local trauma. Physical examination evidenced prolonged right tight pain and no other findings. Bone mineral density evaluated by dual-energy
x-ray absorptiometry was unremarkable. Biochemical investigations showed normal inorganic phosphate, calcium, zinc, and magnesium, but low ALP. The patient had six previous ALP measurements and all of them were below the lower limit of normality (46 to 116 U/L). These measurements were done in regular ob-gyn checkups with no further investigation or follow-up. In light of the hypophosphatemia and pathologic fracture, the serum pyridoxal 5'-phosphate concentration was measured and found to be elevated 35.8 mcg/L (normal 5.0 to 30.0 mcg/L). 

CLINICAL LESSONS/CONCLUSION: Hypophosphatasia occurs due to a deactivating mutation (or mutations) of the gene encoding Tissue-Nonspecific Alkaline Phosphatase (TNSALP), leading to a global deficiency of TNSALP activity and inadequate skeletal mineralization and fractures. The adult form presents during middle age with stress fractures. The first complaints maybe foot pain, which is due to stress fractures of the metatarsals, and thigh pain, due to pseudo fractures of the femur. Our patient illustrates the importance of low serum ALP activity in the assessment of these patients. The correct diagnosis should help to avoid the use of traditional therapies for osteoporosis or osteomalacia, which would be ineffective or potentially harmful.

Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY II

Renal Cell Cancer Metastasis to the Pituitary Masquerading as a Non-Functioning Adenoma

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MON-263

Renal cell cancer (RCC) metastasis to the pituitary (MP) is rare with only a few case reports in literature. A 47 yr. old male with history of RCC and hypertension was seen in pituitary clinic for management of a pituitary mass. He initially presented to an outside hospital with persistent headaches, unintentional weight loss, severe fatigue, dizziness, cold intolerance, low libido and difficulty with erections. CT head showed a pituitary mass. Lab work revealed pan-hypopituitarism with adrenal insufficiency [random cortisol 0.7 ug/dL (4-19 ug/dL); ACTH 13 pg/mL (7-63 pg/mL)], hypothyroidism [T4 0.52 ng/dL (0.7-1.5 ng/dL); TSH 0.3 mU/mL (0.45-4.5 mU/mL)] and hypogonadism [total testosteron 15 ng/dL (200-1000 ng/dL); LH of 0.6 mIU/mL (1.5-9.3 mIU/mL); FSH of 3.5 mIU/mL (1.6-8.0 mIU/mL)]. Prolactin was slightly elevated at 34 ng/mL (2-18 ng/mL) from stalk compression. IGF-1 was 78 ng/mL (Z score -1.2).

When we saw him, he was already on hormone replacement with significant improvement in symptoms. He had no visual field defects or symptoms of diabetes insipidus (DI). As for his history of RCC, he underwent a partial left nephrectomy in 2015, followed by a completion nephrectomy shortly after. Pathology was consistent with clear cell RCC. He did not receive any adjuvant therapy; he underwent a left nephrectomy in 2015, followed by a completion nephrectomy shortly after. Pathology was consistent with metastatic clear cell RCC. A follow up CT showed innumerable lung nodules and a 3.2 cm right adrenal nodule consistent with metastases. He completed stereotactic radiation to the sella and is currently on Iplimumab and Nivolumab for immunotherapy.

Thyroid

THYROID CANCER CASE REPORTS I

A Diagnosis of Thyroid Cancer Reveals a Triple Threat

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SUN-491

Introduction: The risk of secondary malignancies is increased in patients with papillary thyroid cancer (PTC). It is not completely clear if this risk is due to radioactive iodine treatment or due to other causes. We present a case of a patient diagnosed with papillary thyroid cancer (PTC) found to have lung cancer and small lymphocytic lymphoma which appear to be unrelated to radioactive iodine treatment.

Case Presentation: A 72 y/o woman with a history of Graves' disease, atrial fibrillation, and hyperparathyroidism initially presented to care for weight loss. She was found to have hyperthyroidism and was treated with methimazole. Thyroid ultrasound revealed multiple nodules including a 17mm right lower pole nodule with irregular borders for which she underwent FNA. Pathology demonstrated atypical-cells of undetermined significance (AUS), but thyroseq revealed a BRAF V600E mutation. She underwent total thyroidectomy with pathology showing multifocal thyroid cancer, 12mm and 0.8mm with 3/11 involved lymph nodes and a 3.2 cm right adrenal nodule consistent with metastases. Pathology was consistent with metastatic clear cell RCC. A follow up CT showed innumerable lung nodules and a 3.2 cm right adrenal nodule consistent with metastases. He completed stereotactic radiation to the sella and is currently on Iplimumab and Nivolumab for immunotherapy. MP is uncommon, and seen in 1-4% of patients with cancer in autopsy studies. Breast (40%) and Lung cancer (25%) are the most common tumors causing MP in women and men respectively, with very few case reports of RCC MP (2.6%). DI is the most common symptom of MP, which is thought to be due to direct vascular supply from hypophyseal arteries to the posterior pituitary. Interestingly in RCC MP, anterior pituitary dysfunction is more prevalent (90%) compared to DI (24%). There is also a tendency for suprasellar extension, causing vision loss. Radiologic features that could help distinguish MP from an adenoma include a rapidly enhancing sellar mass, erosion of sellar floor or posterior clinoid process, infundibular thickening, and the presence of intra-tumoral vascular flow voids. There reportedly is no benefit to using FDG-PET to diagnose MP from RCC. There are no standardized treatment guidelines and the choice of treatment is also influenced by the extent of systemic disease.

Thyroid ultrasound revealed multiple nodules including a 17mm right lower pole nodule with irregular borders for which she underwent FNA. Pathology demonstrated atypical-cells of undetermined significance (AUS), but thyroseq revealed a BRAF V600E mutation. She underwent total thyroidectomy with pathology showing multifocal thyroid cancer, 12mm and 0.8mm with 3/11 involved lymph nodes and right parathyroid adenoma. Given a questionable lower left lung nodule on preop CXR, she underwent...