

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 =  $44.8 \pm 11.5$  years, range: 26-67); all were female. Mean TED duration was  $4.6 \pm 5.5$  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

#### *Paranglioma 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 spiro-lactone, 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 <sup>18</sup>F-FDG PET-avid. Video-assisted thoracoscopy 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 diagnose 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 thigh pain and no other findings. Bone mineral density evaluated by dual-energy