similar condition in her family, she has three siblings all are well. She developed T2DM with at the age of 14. She was started on Metformin 2 gram daily and them pioglitazone 30 mg was added when she was 16 years. She never had menarche. The clinical examination revealed an adolescent girl with normal BP 106/68 mmHg, and BMI 19.6kg/m^2. She scored 24 onFerriman-Gallwey hirsutism scoring system. She had severe acanthosis nigricans on both axillae. She also had back and upper limbs hyperpigmentation.Lab tests revealed normal thyroid function tests, prolactin, cortisol, DHEA-S, and 17 hydroxy progesterone. Fasting glucose 7.2, insulin 123 μU/ml (2.6-24.9), c-peptide 964, HbA1c 8.2%. Total testosterone 24.61 nmol/L (0.069-2.715), SHBG 184.9 nmol/L, and Free testosterone index 13.31 (0.51-6.53). Her LH 8.9 and FSH 4.7.Radiological investigations revealed polycystic ovaries on pelvic ultrasound. MRI abdomen showed normal adrenals, and mildly enlarged ovaries with peripherally located follicles consistent with polycystic ovarian syndrome. The patient was started in Diane-35 (cyproterone acetate and ethinyl estradiol) oral pills. She started to have menarche three months after using Diane-35. Her Total testosterone had dropped from 24.61 to 1.69 nmol/L (0.069-2.715), SHBG 579 nmol/L, and Free testosterone index 0.29 (0.51-6.53). She reported that the hirsutism is getting less than before starting the treatment.Conclusion: Primary amenorrhea might be a manifestation of in HAIR-AN syndrome due to sever hyperandrogenism. The management of such condition is challenging. In addition to controlling the metabolic parameters, combined oral pills with antiandrogen effect might be effective.

Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY II

Oral Contraceptive Pills Mask an Aggressive Crooke's Cell Pituitary Adenoma

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MON-255

Background

Crooke's cell tumors are rare and aggressive forms of pituitary adenomas. This variant of Cushing's disease requires prompt diagnosis to avoid life-threatening complications. We report a unique case of Crooke's cell tumor with longstanding history of irregular menstrual cycles, undiagnosed and later presented as acute unilateral ptosis and diplopia due to aggressive tumor invasion.

Clinical Case

23-year-old female presented to the ER with facial swelling, left eye droop and diplopia for 3 days. She had a past medical history of oligomenorrhea and hirsutism which was normalized by oral contraceptive pills (OCP)- a combination of ethinyl estradiol and drospirenone for the last 3 years. Years prior, workup of her oligomenorrhea showed normal androgenic profile with normal DHEA-S, testosterone and 17-OH progesterone. Current exam was also significant for elevated blood pressure 200/110mmHg, BMI 37, pigmented abdominal striae and terminal hair over her chin. Labs remarkable for hypokalemia K+ 2.7 mmol/L (3.5-5.3), elevated AM cortisol 51 mcg/dL (4-20), low TSH 0.152 mcUnit/mL (0.4-5.0), low IGF-1 170 ng/mL (222-566) and FSH 1.4 mUnit/mL (1.0-9.0), with normal prolactin 24.3 ng/mL (<0.5-25) and free T4 0.87ng/dL (0.8-1.8). MRI brain showed 2.8cm homogenous enhancing soft tissue mass involving the central skull base, sphenoid sinus, sella, suprasellar cistern, and parasellar regions; displacing the optic chiasm, and invading the cavernous sinuses bilaterally and orbital apices. Post trans-sphenoidal surgery (TSS) of the pituitary mass, her left eye ptosis and diplopia resolved. Post-op MRI showed subtotal resection of the extra-axial enhancing abnormality at the central skull base with extension to multiple other anatomic spaces. Pathology read consistent with aggressive Crooke's cell adenoma, showing invasive biologic behavior without an elevated proliferation index with positive ACTH immunohistochemistry supportive of corticotroph cell adenoma. Post-op ACTH level 73 pg/mL (6-50) and cortisol 12.5 mcg/dL (4-20), while on dexamethasone. Repeat TSS was performed for residual adenoma. Cortisol remains elevated at 15.7mcg/dL despite high dose dexamethasone taper by the neurosurgery team for post-op development of right eye ptosis. She is currently awaiting proton beam radiation therapy.

Conclusion

Crooke's cell tumors are an aggressive form of pituitary adenoma for which early diagnosis is crucial for its prognosis. Our case highlights the importance of maintaining a wide differential in evaluating young women with menstrual irregularities and to include screening for Cushing's syndrome. Empiric treatment with OCPs can mask symptoms in the earlier course of Cushing's disease as in our patient, causing recognition only after presentation with significant tumor growth. Earlier detection could have prevented adenoma invasion and potential neurological sequelae.

Thyroid

BENIGN THYROID DISEASE AND HEALTH DISPARITIES IN THYROID I

Assessing Content Validity of the Graves' Ophthalmopathy Quality of Life Questionnaire (GO-QOL) in the United States

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SAT-424

Introduction: Thyroid eye disease (TED) is an autoimmune condition that negatively impacts patient's quality of life (QOL). The GO-QOL questionnaire was originally developed in the Netherlands to quantify how TED and treatments affect patient QOL. This questionnaire includes eight questions each on visual functioning and appearance related QOL; the items are answered on a 3-point Likert scale and transformed to a 0 (worst) to 100 (best) scale. Though widely used and validated outside the US, the questionnaire has not been validated in the United States (US). Here we examine the content validity.

Methods: Patients with moderate or severe TED were identified using an existing market research patient

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 ^{18}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 tight pain and no other findings. Bone mineral density evaluated by dual-energy