OR30-03

Objective: There is limited data regarding the use of diabetes technology such as continuous glucose monitor (CGM) and continuous subcutaneous insulin infusion (CSII) among patients with type 1 diabetes (T1D) in a minority serving and safety-net hospital. We examined racial differences in the use of CGM and CSII in this setting.

Methods: A retrospective review of 227 patients ≥ 18 years of age with T1D seen in the Endocrinology clinic at a safety-net hospital from October 2016 and September 2017 was completed. Statistical analysis assessed the likelihood of diabetes technology use among different races.

Results: The mean age was 39, 59% male, mean duration of diabetes was 21 years, 30% overweight, 22% obesity, 80% English speaking, and 50% had government insurance. In terms of the distribution of race/ethnicity, 43% were Caucasian, 25% African American (AA), 15% Hispanic, 15% defined as other, and 2% Asian. Mean HbA1c ± standard deviation (SD) of any technology (either CGM or CSII or both) and non-technology users were 8.27 ± 1.58 and 9.49 ± 2.04, respectively. Patients who had government health insurance were found to have lower odds of using technology (odds ratio [OR], 0.43; 95% confidential interval [CI], 0.25 - 0.74) compared to patients who had private health insurance. Overall, 26% of the patients used CSII with 43% of this population Caucasian, 10.5% AA and 14.2% Hispanic. The overall CGM use was 30% with 47% of users Caucasian, 14% AA and 22% Hispanic. In a multivariable logistic regression model that adjusted for insurance and language, AA or other were found to have statistically significant lower odds of using technology (AA OR 0.25 [95% CI 0.11 - 0.53] and other OR 0.33 [95% CI 0.12 - 0.89]) compared to the Caucasian group.

Conclusion: Our study showed that the use of technology in the Caucasian group was statistically significantly higher than in the non-Caucasian groups except for the Asian group. After adjusting for insurance and language, AA and other demonstrated statistically lower rates of technology use. Racial differences in diabetes technology use were observed in our study as well as the association between technology use and lowered HbA1c. Given diabetes technology is a useful tool in reducing HbA1c and hypoglycemia, the barriers to accessing diabetes technology in non-Caucasian individuals should be addressed to decrease health disparities.

Bone and Mineral Metabolism

BONE AND MINERAL CASE REPORTS II

Cutaneous Skeletal Hypophosphatemic Syndrome (Cshs) Caused by Somatic HRAS p.G13R Mutation: Long Follow-Up of Two Brazilian Women

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MON-333

BACKGROUND CSHS refers to the association of epidermal nevus syndrome (ENS), skeletal dysplasia, and hypophosphatemic osteomalacia (OM) mediated by FGF23 resulting from post zygotic mutations in RAS signaling pathway, with known by relationship with human cancers. **CLINICAL CASE** Patient 1 presented ENS since birth at right hemibody. At 1.6-yr-old, she underwent treatment for a left inguinal rhabdomyosarcoma. At 3-yr-old, she had an atraumatic right femur fracture associated with muscle weakness, and laboratory data and X-rays suggesting OM. Phosphate and calcitriol were initiated, but with poor adherence, and no improvement; skeletal deformities got worse and the girl became wheelchair user at 13-yr-old. Skeletal CT scan at age 17 showed dysplastic lesions with lytic changes at right dimidium (skull, jaw, ribs, pelvis and femur) with systemic OM signs confirmed by bone biopsy. The progressive enlargement of the jaw lesion required surgical removal after 2 years; histopathology revealed giant cell tumor. Patient 2 also had congenital ENS on the right dimidium with complaint of bone pain and muscle weakness since 2-yr-old. She evolved with bone fractures and deformities at 4-yr-old, becoming wheelchair user after 2 years. Iliac crest biopsy confirmed OM, already suspected based on laboratorial and X-rays findings at age 7. She had few improvements with phosphate and calcitriol treatment also due to low compliance. During follow-up, symptomatic nephrolithiasis occurred and, in regions affected by EN, multiple basal cell carcinomas (BCCs) emerged requiring excisions. Skeletal CT scan at age 36 showed dysplastic lesions at right hemibody (skull, ribs, pelvis, and limbs) with diffuse bone rarefaction and signs of OM. Sanger sequencing of DNA from EN and jaw tumor samples of patient 1 and from EN and BCC samples of patient 2 disclosed heterozygous HRAS p.G13R mutation, and this mutation was absent in leukocytes DNA from both patients confirming CSHS mosaicism. Owing to the CSHS associated increase risk of cancer, screening with thyroid and breast ultrasound, mammography, CT of skull, chest, abdomen, and pelvis ruled out presence of tumors in patient 1. Patient 2 is waiting for similar screening. Nowadays, patient 1 is 25-yr-old and patient 2 is 36-yr-old; both women have maintenance of OM, characterized by persistent hypophosphatemia with elevated bone formation makers despite treatment with phosphate and calcitriol. **CONCLUSION** CHSC is a very rare syndrome with less than 10 cases with molecular characterization in literature. Although Collins et al suggest an age-dependent improvement in mineral abnormalities, we reported two women without OM recovery probably because of extensive bone dysplasia. These cases also reinforce association of CSHS with neoplasms, including first descriptions of patients with rhabdomyosarcoma and giant cell tumor of jaw and the longest follow-ups described until.

Adrenal

ADRENAL CASE REPORTS II

Clinical and Anatomopathological Characteristics of Two Atypical Aldosterone-Producing Adenomas

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SUN-178

Background: Aldosterone producing adenomas (APAs) are the most common cause of unilateral primary aldosteronism (PA). In most cases, APAs present as small (<2 cm in diameter) benign appearing nodules on computed tomography (CT). Up to 70% of APAs may harbor *KCNJ5* somatic mutations.

Clinical Cases: Case 1. A 33-yr-old man was referred to investigate resistant hypertension (HT). Biochemical evaluation revealed normal K levels, aldosterone (A) of 14.7 ng/dl, renin of 2.1 mUI/L (normal, 4.4-46.1) and A/R ratio of 24.8. Confirmatory testing confirmed PA diagnosis. Hypercortisolism investigation revealed a non-suppressible cortisol after an overnight 1 mg low-dose dexamethasone suppression (8.3 µg/dL), abnormal midnight salivary cortisol, and normal urinary free cortisol, plasma DHEAS and ACTH levels. Computed tomography (CT) scan showed a well-limited mass in left adrenal, measuring 5.8 cm with pre-contrast density of 30 HU and absolute wash-out of 72%. After left laparoscopic adrenalectomy, hydrocortisone was started and adrenal insufficiency confirmed by basal cortisol <3 µg/dL. He presented biochemical cure of PA and improvement in HT control. Histologic examination revealed an encapsulated tumor with glomerulosa-like cells predominance and a Weiss score 1 (clear cells <25%). CYP11B2 staining was positive in 10% and Ki67 in 5% of tumor cells. **Case 2.** A 58 yr-old woman was referred to investigate an adrenal mass. She had resistant HT and hypokalemia since 2010. Biochemical evaluation revealed hypokalemia (2.8 mEq/L), A of 16.9 ng/dl, renin <1.6 mUI/L and A/R ratio of 10.6. Confirmatory testing confirmed PA diagnosis. Hypercortisolism investigation was negative. CT scan showed a heterogeneous solid mass in the right adrenal, measuring 5.5 cm with pre-contrast density of 30 HU and absolute wash-out of 77%. After laparoscopic right adrenalectomy, histologic examination revealed an encapsulated tumor with glomerulosa-like cells predominance and a Weiss score 2 (clear cells < 25% and > 1/3 diffuse architecture). CYP11B2 staining was positive in 30% and Ki67 in 5% of the cells. She presented biochemical cure of PA and improvement in HT control. Genetic investigation for somatic *KCNJ5*, *ATP1A1, ATP2B3* and *CTNNB1* was negative in both cases.

Conclusion: We describe two rare cases of APAs that presented as large and suspicious tumors, without somatic mutations in genes associated with APAs.

Adrenal

ADRENAL CASE REPORTS II

A Case of Metastatic Merkel Cell Carcinoma Within a Cortisol-Producing Adrenal Adenoma

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SUN-181

Introduction:

Metastasis within another tumor is rare, and exceedingly more rare within a functional adrenal adenoma. Merkel cell carcinoma (MCC) is an aggressive and rare neuroendocrine carcinoma of the skin. We present a patient with history of MCC who presented with an adrenal mass, found to be metastatic MCC within a cortisol-producing adrenal adenoma.

Case description:

54 year old male with a history of type 2 diabetes, hypertension, MCC of right thigh, presented for adrenal mass. He was initially evaluated in the ER for severe abdominal pain for 4 days. He had a CT abdomen done, which showed 6cm right adrenal mass with extensive fat and soft tissue component within it. Hounsfield units within solid area measured up to 47 units, and surrounding area measured -3 Hounsfield units.

His history was significant for MCC of right thigh, type 2 diabetes, HTN, obesity and OSA. Pathology from MCC resection showed no evidence of invasion and clear margins, but he did have microscopic focus on inguinal lymph node biopsy. His PET scan was negative except for low uptake in a 6cm right adrenal lesion that was considered benign-appearing. He completed radiation therapy to right thigh and groin.

Physical exam was notably negative for any cushingoid features. Aldosterone, renin and plasma metanephrines were normal. Cortisol after dexamethasone suppression test was elevated, and ACTH and DHEAS were low, raising suspicion of Cushing's syndrome. He had right adrenalectomy and was then started on steroid replacement. Pathology showed adrenocortical adenoma with a 2cm well-circumscribed mass within the adenoma positive for metastatic MCC with lymphovascular invasion. ACTH stimulation testing was performed at follow up after holding hydrocortisone for over 24 hours, and was consistent with suppression of glucocorticoid axis in the contralateral adrenal gland.

Discussion:

MCC and metastasis within a functional adrenal tumor are both rare occurrences. Our patient had positive regional lymph node involvement at the time of diagnosis, and although the initial PET scan was interpreted as showing an benign adrenal adenoma, it is likely that this represented distant metastasis. Labs indicated that the surrounding adenoma was likely cortisol-producing. To our knowledge,