At 6 years of age, he started to demonstrate bilateral testicular enlargement of 4-6 mL with pubic hair Tanner stage I. Bone age was concordant with chronologic age. Gonadotropins and testosterone level were pre-pubertal. Testicular ultrasound showed bilateral enlargement with punctuate densities without evidence of hydrocele or mass. By 8 years of age, he had further testicular enlargement to 8 mL bilaterally, normal penile length 5.5 cm (6.2 ± 1.0) and no pubic hair. Biochemical evaluation showed normal thyroid function and pre-pubertal gonadotropins and testosterone. Repeat testicular ultrasound showed heterogeneous echotexture representing microlithiasis, without a focal mass.

Conclusion: Macroorchidism can be present in the absence of gonadotropin independent precocious puberty in MAS. Testicular ultrasound evaluation is important, as hyper-hypoechoic lesions, bilateral microlithiasis, focal calcifications, are the most common testicular abnormality in males with MAS.

Microlithiasis, due to calcium deposits within the seminiferous tubules, have been described often in this condition, and should be considered a marker of male MAS. The natural history of male fertility and testicular cancer risk in the context of microlithiasis and testicular calcifications is unknown. Close observation and testicular imaging at baseline and after age 5, to characterize subclinical involvement, are an appropriate course to follow.

Pediatric Endocrinology

PEDIATRIC OBESITY, THYROID, AND CANCER

Hyperosmolar Hyperglycemic State as Initial Presentation of Type 1 Diabetes in Children

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MON-084

INTRODUCTION: We present 2 pediatric patients with Hyperosmolar Hyperglycemic State (HHS) at diabetes onset.

CASE 1:

3 year old African American female presented to the pediatrician office with a 5 day history of polydipsia, polyuria and emesis. POCT glucose read high and patient was transferred to the PICU. Laboratory studies were significant for serum glucose of 1032 mg/dl, Na 128 mMOL/L (corrected 142mMOL/L), VBG showed pH 7.36, HCO3 20 mMOL/L, Serum osm 331 mOsm/Kg. Patient received a 20ml/kg bolus of 0.9%NaCl, followed by 2 times maintenance IV fluids and glargine 2 units. Glucose dropped from 418 to 122 mg/dl in 3 hours. Due to this Dextrose was added and IVF rate was decreased.

CASE 2:

8 year old African American obese male was admitted to the PICU for management of new onset diabetes. He presented with 3 days of flu-like symptoms and worsening drowsiness. Patient had increased consumption of large quantities of sugary beverages due to increased thirst. Laboratory workup: serum glucose of 2309 mg/dl, Na 133 mMOL/L (corrected 168 mMOL/L), pH 7.13, HCO3 10 mMOL/L. Patient was given 30 cc/kg NS bolus followed by an insulin drip of 0.1 u/kg/hour. Repeat studies 3 hours later showed a serum glucose of 1,414 mg/dl, Na 152 mMol/L (corrected 184 mMOL/L), pH 7.19, HCO3 17 mMOL/L, and serum osmolality of 408 mOsm/Kg. IVF were adjusted to correct the water deficit and insulin drip was decreased to 0.05u/kg/hour.

DISCUSSION: HHS continues to be a challenging diagnosis due to its low frequency compared with Diabetic Ketoacidosis especially when presenting at a very young age. Most practitioners will mistake the presentation for DKA and start an insulin drip. The early use of insulin is not necessary in the setting of HHS due to the risk of complications. A fast drop in glucose decreases the osmotic pressure and compromises the circulatory status with a higher chance of thromboembolism. In mixed HHS and DKA, the management aligns more with the DKA management but the amount of fluids needed is higher and insulin infusion may cause fast drop of glucose with potential decrease of intravascular volume as in our second patient. It is imperative that the diagnosis of HHS is made early so that the appropriate treatment can be instituted.

CONCLUSION: Appropriate fluid administration and delay in insulin administration are key in the management of HHS. The awareness of this possible presentation and the early recognition and appropriate fluid management are needed to improve outcomes.

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Adrenal

ADRENAL CASE REPORTS II

Bilateral Aldosterone-Producing Adenomas: A New Subtype of Bilateral Primary Aldosteronism?

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SUN-195

Background: Primary aldosteronism (PA) is the most common cause of endocrine hypertension (HT). PA subtypes include aldosterone-producing adenomas (APA) and bilateral adrenal hyperplasia. To date, few PA patients with bilateral adenomas have been reported, but only one case was well characterized by anatomopathological analysis and clinical outcome after adrenal sparing surgery (1).

Clinical case: A 53-year-old woman was referred to investigate resistant HT and hypokalemia. (3.0 mEq/L). PA

screening revealed aldosterone (A) of 37.9 ng/dL, renin (R) < 1.6 (4.4-46.1 mUI/L), A/R ratio of 24.8. Confirmatory testing confirmed PA diagnosis: seated saline infusion test (A= 83.3 ng/dL) and intravenous furosemide test (R= 3.1 mUI/L; positive test <13 mUI/L). Hypercortisolism investigation revealed a non-suppressible cortisol after an overnight 1 mg low-dose dexamethasone suppression [cortisol (C)= 2.9 µg/dL and dexamethasone= 701 (>130 ng/dL)], and normal urinary free cortisol, midnight salivary cortisol, plasma DHEAS and ACTH levels. Computed tomography demonstrated bilateral adrenal nodules without adrenal thickening: 3.5 cm right nodule (pre-contrast density of 7UH density; absolute wash-out of 71%) and 2.5 cm left nodule (pre-contrast density of 8UH density; absolute wash-out of 78%). Sequential adrenal venous (AV) sampling (AVS) under continuous cosyntropin infusion showed a lateralization index of 3.4 (bilateral disease <4). Then, the patient underwent right adrenalectomy and left nodulectomy. In the postoperative period, she presented normalization of K^+ levels and complete HT remission. She remained under hydrocortisone replacement for 2 months. After 2 months, biochemical evaluation revealed normal basal cortisol levels (13.3 µg/dL) and biochemical cure of PA (A= 3.1 ng/dL and R= 15.3 mUI/L). Currently, she doesn't have symptoms of adrenal insufficiency after discontinuation of hydrocortisone. Anatomopathological analysis showed bilateral adenomas (Weiss score of 0) in both sides without adjacent hyperplasia. CYP11B2 immunohistochemistry displayed a strong staining in 50% of cells in the right adenoma and in 30% of cells in the left adenoma. Few aldosterone-producing cell clusters (APCC) were identified in the right zona glomerulosa, which is a frequent finding in normal adrenals.

Conclusion: We herein described a very rare case of PA caused by bilateral-producing adenomas, confirmed by AVS and CYP11B2 staining after adrenal sparing surgery.

Bone and Mineral Metabolism

BONE AND MINERAL CASE REPORTS II

Tumor-Induced Osteomalacia from a Hypervascular Thoracic Paraspinal Mass: Challenges in Diagnosis and Management

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MON-353

Background: Tumor-induced osteomalacia (TIO) is a paraneoplastic syndrome driven by ectopic production of fibroblast growth factor 23 (FGF23), resulting in renal phosphate wasting, hypophosphatemia, and bone demineralization. Successful therapy requires complete resection of the tumor, which is often challenging to localize. **Clinical Case:** A bedridden 37-year-old woman presented to Endocrine Clinic after 9 years of progressive pain and weakness in her back, hips, and extremities. She had previously been diagnosed with a neurodegenerative disease, though an MRI from 5 years ago showed bilateral subacute sacral ala fractures. CT scan on presentation showed generalized osteopenia and numerous subacute to chronic atraumatic fractures (involving ribs, scapula, pubic rami, and right femoral shaft) concerning for osteomalacia. On exam, she had profound lower extremity weakness. Laboratory testing was notable for serum phosphorus 0.6 mg/dL (normal (nl) 2.5-4.9), calcium 8.4 mg/dL (nl 8.5-10.5), PTH 216 pg/mL (nl 13-85), 25-OH vitamin D 26 ng/mL (nl 30-100), alkaline phosphatase 345 U/L (nl 45-130), $1,25\text{-}(OH)_2$ vitamin D 28 pg/mL (nl 18-72), renal tubular phosphorus reabsorption TmP/GFR 0.53 mg/dL (nl 2.97-4.45), and inappropriately normal FGF23 level 170 RU/mL (nl<180). Phosphate salts, calcitriol, and cinacalcet were initiated. Octreotide scan was negative, but ^{18}F-FDG PET/CT showed mild hypermetabolic activity in the lower thoracic paraspinal area correlating with a 3.4 cm hypervascular mass near the left 8th rib head and two adjacent nodules on MRI. Due to bleeding risk, biopsy was deferred and further confirmatory studies were pursued. Genetic testing for hereditary forms of hypophosphatemia (FGF23, CLCN5, DMP1, ENPP1, FGFR1, PHEX) was negative. ^{68}Ga-DOTATATE PET/CT was not available, but selective venous sampling for FGF23 revealed a 3:1 ratio between venous drainage of the tumor bed and general circulation. Based on this localization, surgical resection was performed. Pathology was consistent with a phosphaturic mesenchymal tumor, but clear margins could not be confirmed due to intraoperative tumor fragmentation. Three months of medical management with phosphate salts and calcitriol normalized her phosphorus levels (2.5-3.3 mg/dL), and she underwent surgical repair of her femoral fracture. Although MRI revealed only postsurgical changes in the spine, her continued requirement for phosphate supplementation and persistently elevated FGF23 levels (258 RU/mL) are concerning for residual tumor. **Conclusion:** This case illustrates the challenges of TIO due to a hypervascular mass in a rare paraspinal site localized by selective venous sampling for the first time at our institution. The patient's continued FGF23 elevation underscores the importance of advancing medical therapies, such as burosumab, in patients with TIO that are not amenable to complete surgical resection.

Neuroendocrinology and Pituitary

NEUROENDOCRINOLOGY AND PITUITARY

Patient Health Questionnaire 9 (PHQ-9) and Barrat Impulsivity Scale (BIS-11); Tools to Identify Side Effects of Dopamine Agonist Treatment in Patients with Pituitary Adenomas

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