1. Soltani A, Pourian M, Davani BM. Does this patient have Pheochromocytoma? a systematic review of clinical signs and symptoms. *J Diabetes Metab Disord*. 2016 Mar 17;15:6. doi: 10.1186/s40200-016-0226-x. Erratum in: J Diabetes Metab Disord. 2017 Oct 16;16:42. PMID: 26998444; PMCID: PMC4797176.

Pediatric Endocrinology

PEDIATRIC ENDOCRINE CASE REPORTS II

Adrenal Hypoplasia Congenita - Is It Possible to Make This Diagnose in Previous "Idiopathic" Adrenal Insufficient Patients? Series of Cases.

Leila Pedroso de Paula, PhD[1], Nathalia Lisboa Gomes, PhD[2], Mauro Antonio Czepielewski, PhD[1], Berenice Bilarinho De Mendonca, PhD[2].
[1]Hospital de Clnicas de Porto Alegre, Porto Alegre, Brazil, [2]Hospital das Clnicas de São Paulo, São Paulo, Brazil.

MON-076

Background: Adrenal hypoplasia congenita (AHC) is a rare disease (1:70.000 men) characterized by reduction in all cortical adrenal hormones and also by hypogonadism. NR0B1 -related AHC includes both X-linked AHC and Xp21 deletion. AHC gene mutation was first described in 1994 and occurs in less than 1% of adrenal insufficiency (AI) cases and it is imperative that clinical endocrinologists increase their knowledge on this condition to recognize it as promptly as they were used to do so in other causes of pediatric AI.**Clinical Cases**: We describe here two sporadic cases and three familial cases from one family, all of them attending at our Endocrine Unit at HCPA, in the South of Brazil. The median age of AI first symptoms was 1 month old in two patients (acute infantile onset due to a salt wasting crises), and eleven years old in the remaining patients (childhood onset). Their exams confirmed the clinical suspicion of primary AI because of low plasma cortisol (from < 0.5 to 4 mcg/dl - reference range from 7-24 mcg/dl), as well as all adrenal steroids levels, high ACTH levels (from 672 to 2225 pg/ml - reference range from 7-63 pg/ml) and high plasmatic renin (from 92 to 448 mcUI/ml - reference range from 2.8-39.9 mcUI/ml). They were also extensively investigated searching for the AI pathophysiology. After some years of lost follow-up, one of the familial cases that had first been seen in 1993, returned to our hospital and was diagnosed with hypogonadism, leading to NR5OB1 gene evaluation.Regarding the genetic etiology, one of the sporadic cases developed a clinical picture including profound developmental delay, seizures and strabismus suggestive of a contiguous gene syndrome that involves Duchenne muscular dystrophy, glycerol kinase deficiency and AHC. A CGH array was performed and identified a Xp21.3-p21.1 deletion. The affected family had an extensive history suggestive of a genetic disease with X-linked inheritance pattern, as shown by the premature death of 3 uncles and a male cousin. A deleterious variant (c.131_212delinsTGAGACCTGTACCGT) in NR5OB1 gene was identified by Sanger sequencing in hemizygous state in the 46,XY affected patients.**Conclusion**: Albeit a rare disease, it is crucial that endocrinologist all around the World could be aware about clinical characteristics of this condition in order to properly diagnose it. And it is even possible, as it occurred with one of our patient, to make a late pathophysiologic diagnosis, making possible to treat the associated hypogonadism. Reference: Acherman JC, Vilain EJ. NR0B1- related adrenal hypoplasia congenita. GeneReviews 1993-2019.

Bone and Mineral Metabolism

OSTEOPOROSIS: DIAGNOSIS AND CLINICAL ASPECTS

Role of QUS and Functional Tests in Evaluating Fracture Risk Based on Frax Tool

Eduardo Medeiros Ferreira Gama, MD[1], Leandro Kasuki Jomori de Pinho, MD,PhD[2], Miguel Madeira, PhD[1], Carlos Duarte, PhD[3], Girlene Canhete Pessoa, FE[4], Aline Barbosa Moraes, MS[5], Maria Caroline Alves Coelho Amaral, Master[6], Francisco Paula Paranhos Neto, PhD[1], Maria Lucia Fleiuss de Farias, PhD[1].
[1]UFRJ/CENTRO CIENCIAS DE SAUDE, Rio de Janeiro, Brazil, [2]Instituto Estadual do Crebro Paulo Niemeyer, Rio De Janerio, Brazil, [3]UFRJ, Rio de Janeiro, Brazil, [4]UFRJ/Gedopa, Rio de Janeiro, Brazil, [5]UFRJ, Rio De Janeiro, Brazil, [6]Universidade Federal do Rio de Janeiro, Rio de janeiro, Brazil.

SUN-388

Background: Osteoporosis is common among elderly people, and identifying those at high risk for fracture is very important.

Aim: To evaluate whether the use of quantitative calcaneal ultrasound (QUS), the Sitting–rising test (SRT) and handgrip test (HT) are additional tools for tracking fragility fracture risk when compared to FRAX and NOGG.

Methods: During the national campaign against osteoporosis, held in 2018 in Rio de Janeiro, participants were randomly selected to perform QUS, SRT and HT, besides categorization of the risk of major and hip fractures by FRAX and NOGG. The following adequacy values were used: *QUS* T-score> -1.05 (adequate) or ≤ -1.05 (inadequate); Sitting – rising test (*SRT*) (composite score): age-reference values at quartiles 3 and 4 (adequate); quartiles 1 and 2 (inadequate); best result 3 attempts of the dominant arm *handgrip test*, according to age and gender: percentile ≥50 (adequate) and <50 (inadequate); *FRAX* tool: suggests high risk for major osteoporotic fractures if > 20% and for hip fractures when > 3%; *NOGG (*complement to FRAX): patient´s risk for major and for hip fractures considered as low (green zone), medium (yellow zone) or high (red zone). Qui square test was used for associations.

Results: We included 162 individuals: 118 females, mean age 66.8 years and 44 males, mean age 71.8 years. High risk of hip fractures by FRAX was observed in 51% of those patients with a QUS T-score ≤-1.05 while it was observed in 28% of those with a QUS T-score> -1.05 (p=0.005). An inadequate QUS T-score was also associated with a higher risk of hip fracture by NOGG (p=0.007). An inadequate SRT and HT were not associated with a high fracture risk.

Conclusions: As densitometry, a method established in clinical practice for the diagnosis of osteoporosis, has limitations in its use, other tools are necessary for tracking the risk of fragility fractures in these events. Quantitative calcaneal ultrasound was a good predictor of hip fracture

risk, while SRT and HT were not capable of evaluate for fracture risk stratification in our study, reinforcing the need for QUS for screening in large populations. Having strength and functional ability did not eliminate the need for investigation.

Neuroendocrinology and Pituitary

PITUITARY AND NEUROENDOCRINE CLINICAL TRIALS AND STUDIES

Opportunistic Assessment of Pituitary Gland with Routine MRI and PET/CT Can Guide in Earlier and Increased Identification of Hypophysitis in Patients Treated with Combination Checkpoint Inhibitors

Anna Galligan, FRACP[1], Amir Iravani, FRACP, FRANZCR[2], Arian Lasocki, FRANZCR[2], Roslyn Wallace, MBBS[2], Alison Weppler, FRACP[2], George Au-Yeung, FRACP[2], Nirupa Sachithanandan, MD, phd[2], Cherie Ying Chiang, MBBS,FRACP,FRCPA[2], John Wentworth, FRACP, PHD[2], Peter Grahame Colman, MBBS,MD[2], Thomas William Kay, MBBS,PHD,MD[1], Balasubramanian Krishnamurthy, MD, DM, FRACP[1], Shahneen Sandhu, FRACP[2].
[1]ST VINCENT'S INST OF MED RSRCH, Fitzroy Vic, Australia, [2]Peter MacCallum Cancer Centre, Melbourne, Australia.

OR32-06

Background: Hypophysitis is one of the commonly reported adverse events related to immune checkpoint inhibitors (ICI), and the incidence is expected to rise with increased use of combined programmed cell death protein 1 (PD1) and cytotoxic T lymphocyte associated protein 4 (CTLA4) blockade. The clinical diagnosis can be delayed due to non-specific symptoms. At our centre, subjects undergo periodic imaging to assess tumour response to ICI. We reviewed whether neuroimaging studies can guide us in the diagnosis of hypophysitis and whether early changes can be detected before the onset of the clinical syndrome. Methods: We retrospectively reviewed the medical charts, biochemistry, structural brain imaging and whole-body positron emission tomography (PET) with specific reference to hypophysitis in 162 patients treated with combination ICI at a tertiary melanoma referral centre. Suspected cases were identified based on meeting one or more of the following criteria: 1) A documented diagnosis of hypophysitis or pituitary dysfunction found on chart review, 2) A relative change in pituitary size or appearance from baseline on neuroimaging studies, or 3) An increase in pituitary maximum standardized uptake value (SUVmax) greater than 25% from baseline on ^{18}F-FDG PET. Results: 58/162 patients (36%) met criteria for suspected hypophysitis. Only 4 patients were identified on routine screening of early morning cortisol. 14 patients presented with symptoms leading to biochemical work up. A further 40 patients were found to have suspicious imaging changes, 13 of which went on to receive a formal diagnosis of hypophysitis. Of the remaining 27 patients, 23 were receiving high dose glucocorticoids for concomitant immune related adverse events at the time of the abnormal imaging study.Conclusion: We report the highest incidence to date of suspected hypophysitis in cohort of patients treated with combination ICI. This study highlights the important role of structural and functional neuroimaging in the early recognition of hypophysitis. Imaging may also play a role when the clinical syndrome is masked by concurrent glucocorticoid use.

Neuroendocrinology and Pituitary

CASE REPORTS IN UNUSUAL PATHOLOGIES IN THE PITUITARY II

Hyperprolactinemia: An Unusual Initial Presenting Manifestation of Multiple Sclerosis

MaKenzie Hodge, MD, Vijaya A. Ganta, MD, FACE, Gail Louise Nunlee-Bland, MD, Wolali Akua-Sabia Odonkor, MD, Anteneh Woldetensay Zenebe, MD, FACP, FACE, Kehinde Matilda Folawewo, MbChB, Nada Osman, MD.
Howard University Hospital, Washington, DC, USA.

MON-258

Hyperprolactinemia and multiple sclerosis (MS) have a direct relationship and hyperprolactinemia may precede clinical signs of MS as a heralding manifestation of disease. Prolactin has significant pro-inflammatory effects in addition to its lactotrophic properties and can also lower the body's immune tolerance, inducing autoimmunity. High levels of prolactin have been thought to contribute to the inflammation of multiple sclerosis. However, elevated levels of prolactin, especially in pregnant women, can be protective for MS patients and induce remission. Prolactin is neuroregenerative and stimulates the precursors for oligodendrocytes, the cells responsible for myelination. Our hypothesis is that an elevated prolactin level detected during an MS flare should not be treated with dopamine agonist, but rather allowed to decrease as the MS improves with treatment.

Case Presentation

A 24 year old woman with a history of marijuana use is referred to our clinic for elevated prolactin levels associated with galactorrhea for 3 months duration. In addition to marijuana use, patient was also sexually active and having regular menses, with menarche at age 11 years old. On physical exam, the patient was found to have bilateral nipple discharge with stimulation, and visual fields were intact to confrontation. At the time of referral, the patient's prolactin was 92.3 ng/dL (4.8–23.3 ng/mL) TSH was normal, and pregnancy test negative.An MRI showed multiple areas of enhancement compatible with active demyelination, concerning for multiple sclerosis. The pituitary gland was enlarged, without evidence of adenoma. A follow up prolactin level was 101 ng/dL and upon further discussion, patient also admitted to some "funny feeling" and weakness in her right hand and a feeling of being "off balance" diagnosed as a left ear infection. Patient was advised to seek urgent treatment for multiple sclerosis. She was admitted, where she was seen by neurology and diagnosed with relapsing remitting multiple sclerosis. She was initially treated with a course of IV methylprednisolone. She was discharged after this course and followed with neurology as an outpatient. For a few months our patient went into remission and her prolactin improved to 24 ng/dL. A few months later, she had a significant increase in her prolactin to 71.5 ng/dL accompanied by evidence of disease progression on MRI and symptoms of weakness