the emergency room where he was treated for pituitary apoplexy with steroids and surgery. Vision improved the next day. Despite uncomplicated post-operative course, patient developed proximal muscle weakness and exam notable for diffuse motor deficit in bilateral lower extremities with hyperreflexia. Endocrinology workup was negative for hypercortisolism and ophthalmology diagnosed him with optic neuropathy. Neurology evaluation led to a diagnosis of multiple sclerosis (MS). Patient was started on natalizumab with complete resolution of all visual and muscle symptoms.

Clinical lesson Our patient presented with complaints of fatigue, decreased libido and work up that showed a macroprolactinoma without MRI evidence of optic chiasm impingement. During treatment, he developed acute visual deficits that were attributed to pituitary apoplexy. This visual disturbance improved after surgery and use of high dose IV steroids, with the latter likely treating what had been an MS flare. In hindsight, ophthalmologic evaluation before surgery had shown new color blindness, a sign of optic neuropathy. Despite temporary relief, patient progressed to develop new muscle weakness and recurrent visual disturbance which led to the diagnosis of MS. Since being diagnosed and treated for MS, he has had complete resolution of his symptoms. This case stresses the importance of considering other etiologies for visual defects in patients with pituitary adenomas.

Neuroendocrinology and Pituitary

CASE REPORTS IN CLASSICAL AND UNUSUAL CAUSES OF HYPOPITUITARISM II

Histologically Proven Lymphocytic Hypophysitis with Marked Improvement on Glucocorticoid Therapy

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MON-240

Background: Lymphocytic hypophysitis (LH) is a rare autoimmune disorder characterized by lymphocytic infiltration of the pituitary gland. The disease predominantly affects women, with >50% of cases presenting during pregnancy or postpartum.[1] LH is often associated with other autoimmune conditions, primarily thyroiditis, and adrenalitis.[2]

Clinical case: A 27-year-old female presented with secondary amenorrhea for eight months. Workup revealed hyperprolactinemia (PRL 65 ng/mL) and a heterogenous pituitary mass measuring 3.3 cm in the largest dimension. Cabergoline was initiated for a presumed prolactinoma. Laboratory evaluation was significant for hypogonadotropic hypogonadism (estradiol <50 pg/mL, progesterone <1 ng/mL, FSH 2.9 mIU/mL, LH 0.45 mIU/mL) despite normalization in prolactin. She was also found to have Hashimoto's thyroiditis (FT4 0.7 ng/dL, TSH 8.2 uU/mL with positive TPO antibodies) and was started on levothyroxine.

Repeat imaging demonstrated a 2.4 cm heterogenous expanding sellar mass with soft tissue extension to the dorsum sella concerning for a meningioma. Visual field testing was intact without evidence of chiasmal compression. She underwent trans-sphenoidal pituitary decompression surgery which was terminated prematurely due to the presence of extensive fibrous tissue in the sella. Pathology was consistent with LH. Immunohistochemical staining was positive for lymphocytic markers CD3 and CD20, confirming marked infiltration of inflammatory B-cells and T-cells. Her postoperative course was notable for panhypopituitarism. In view of the pathological findings of LH, she was started on a high dose of 40mg of prednisone daily. Within two months, sellar magnetic resonance imaging revealed a homogenous normal-appearing pituitary with a reduction in soft tissue mass in the sellar and suprasellar region. Oral contraceptive therapy was initiated for sex hormone replacement with the resumption of menses. Prednisone was gradually tapered to 5mg/day, and she was subsequently transitioned to maintenance hydrocortisone for central adrenal insufficiency.

Discussion: LH is a rare chronic inflammatory disease that should be considered in the differential diagnosis of a non-secreting pituitary mass, especially if occurring in young women presenting during pregnancy or postpartum. The condition is associated with preferential destruction of corticotroph and thyrotroph cells.[3] Appropriate management remains controversial. High dose glucocorticoid therapy, to which our patient responded to dramatically, has been shown to be beneficial in reducing mass effect. Optimal treatment involves surgical resection of the pituitary mass to decompress surrounding structures.[3]

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Adrenal

ADRENAL CASE REPORTS I

Recurrent Co-Driver Mutation in CTNNB1-Mutant Aldosterone-producing Adenomas (APA), Causing Reversible Hypertension in Puberty, Pregnancy or Menopause

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SAT-224

Background: Three patients with a syndrome of LH/HCG-activated primary aldosteronism in pregnancy or menopause carrying somatic *CTNNB1* mutations were reported four years ago (Teo et al. NEJM 2015). This proved but half the story. Diagnosis of an APA in a 12-year old boy with severe hypertension revealed an apparently essential co-driver mutation. **Methods:** WES of tumour and blood was performed in the pubertal boy. Candidate genes were Sanger sequenced in other APAs from GB/Ireland, and France with known or suspected *CTNNB1* mutations. *LHCGR*, *GNRHR* and *CYP11B2* expression were measured in all available patients' APAs and the adjacent adrenal gland (AAG) by RT-PCR. RNA and gDNA from the zona glomerulosa (ZG) of the proband's AAG were collected by laser capture microdissection for Sanger sequencing of *GNA11* and *CTNNB1*. Function of mutant genes was assessed by measurement of aldosterone production and *LHCGR* expression by immunofluorescence (IFC) in NCI-H295R adrenocortical cells and primary human APA cells. **Results:** The proband's APA contained a p.(S45F) somatic mutation in *CTNNB1*, and a p.(Q209P) somatic mutation of the GTPase-activating residue (Q209) in *GNA11*. Mutations of Q209, to P or H, were also found in six other GB/Irish patients with previously identified mutations of *CTNNB1* (S33C, G34R, T41A, S45F, or S45P). All seven patients remain normotensive 2-12 years post-adrenalectomy, including some with long-standing pre-operative hypertension. Four of the 13 French patients with *CTNNB1* exon 3 mutant APAs have somatic mutation of Q209 of either *GNA11* (n=3) or *GNAQ*. In comparison with their own AAG, the GB/Irish double mutant APAs showed an increase in expression of *LHCGR*, *CYP11B2* and *GNRHR* by 32-166, 158-18980, and 1174-6642 fold, respectively. All four French double-mutants had >10 fold higher *LHCGR* than APAs with single mutations of *CTNNB1* or other genes. Hyperplasia of ZG was observed in the ZG of the boy's AAG but no APCC was detected. Homozygous or heterozygous Q209P mutation of *GNA11* was detected in multiple ZG samples in RNA and/or gDNA but WT in *CTNNB1* exon 3. H295R cells (*CTNNB1* S45P) were *GNA11* WT. Overexpression of *GNA11* Q209 mutation increased aldosterone secretion to 465% of *GNA11* WT overexpressing cells (n=6, P<0.001) and *CYP11B2* expression was also increased several-fold. Smaller increases were seen in primary human adrenal cells after double-transfection by *GNA11* and *CTNNB1* mutants (n=3, *P*<0.001). This also caused membrane expression of LHCGR, visualised by IFC. **Conclusions:** APAs with double mutation of *GNA11*/*GNAQ* Q209 and *CTNNB1* have a distinct phenotype, in which hypertension is triggered by high LH or HCG, and cured in all cases by adrenalectomy. *GNA11/Q* mediates the aldosterone response to ANGII, and the Q209 codon is analogous to the Q227 of GNAS, mutated in McCune Albright. Mosaicism for *GNA11* may cause ZG hyperplasia.

Reproductive Endocrinology

FEMALE REPRODUCTION: BASIC MECHANISMS

Steroid Cell Tumor, Not Otherwise Specified; A Rare Case of Hyperandrogenism

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MON-024

STEROID CELL TUMOR, NOT OTHERWISE SPECIFIED, A RARE CAUSE OF HYPERANDROGENISM

BACKGROUND:

Steroid cell tumor is a rare sex cord stromal tumor. There are 3 types including steroid cell tumor not otherwise specified (NOS), stromal luteoma and leydig cell tumors.

Steroid cell tumor (NOS) is the most common of all the subtypes. About 75% of the steroid cell tumor (NOS) are secretory. They can secrete androgens and estrogens. In a few cases, cortisol and renin secretion have been reported. The patient's clinical features depend on the hormone secreted.

CASE PRESENTATION

Here, we report a case of a 22-year-old woman who was seen at an outpatient clinic for hirsutism, irregular menstrual bleeding, and progressive weight gain. Examination revealed androgenic facial hair growth, clitoromegaly and obesity.

Initial differentials on presentation were PCOS, ovarian or adrenal pathology. Initial LH, FSH values were normal. Lab investigation showed elevated testosterone, DHEAS and 17-OH progesterone levels. Baseline labs showed Androstenedione of 3345 ng/dl (41-262 ng/dl), DHEAS of 595.5 ug/dl (110- 431.7 ug/dl), 17-OH progesterone was 2394 ng/dl (follicular: 15-70 ng/dl, Luteal: 35-290 ng/dl),