

ACTH causes elevated levels of steroid hormone precursors & chronic stimulation of the adrenal glands resulting in hyperplasia. The most common form of CAH is due to 21OH deficiency (21OHD) with the classic, salt-wasting form causing glucocorticoid & mineralocorticoid deficiency & androgen excess.

Poorly controlled CAH causes increased production of androgens & progesterone & decreased fertility. Fertility in woman with 21OHD can be challenging due to decreased sexual interest & anatomical abnormalities. Despite these challenges, the pregnancy rate is not significantly lower in women with well controlled 21OHD. However, as these patients are uncommon recommendations for pregnancy can be challenging for endocrinologists.

Clinical Case:

A 27-year old G0P0 female with classic, salt losing CAH presented to discuss pregnancy. Her medications were hydrocortisone 10mg qAM & 5mg qHS & fludrocortisone 0.1mg daily. She denied symptoms of dehydration, nausea, vomiting, dizziness, or fatigue.

She was diagnosed with CAH while in-utero & started on steroid therapy after birth. She did not require any surgical genital reconstruction. Menarche occurred at age 11 and she had regular menses every 35-40 days, with no evidence of excessive androgen exposure including excess body hair, deepening of her voice, or cliteromegaly.

On physical exam she was normotensive & had no evidence of virilization or cushingoid features. She exhibited minor darkening of the palmar creases. Her labs were significant for free testosterone of 9.1 pg/ml (0.2-5.0 pg/ml), total testosterone of 115 ng/dl (2-45 ng/dl), ACTH of 780 pg/ml (6-50 pg/ml), androstenedione of 636 ng/dl (41-262 ng/dl), & 17 OHP of 1560 ng/dl (15-290 ng/dl).

Her hydrocortisone dose was increased to 20 mg qAM & 10 mg qHS & fludrocortisone 0.1mg daily was continued. Our objectives were to normalize the androgen level & suppress serum progesterone to less than 2 nmol/L. If the objectives were not reached she would be converted to prednisone BID.

Conclusion:

Endocrine providers are the primary resource for fertility recommendations for 21OHD patients & must understand the challenges in this very rare group of patients. Collaboration with the perinatologist is crucial for success. The goals of preconception endocrine assessment in a patient with classic CAH are to adjust hormone therapy to ensure optimal endocrine milieu for conception & risk assessment of having a child affected with 21OHD. The patient will need a higher dose of fludrocortisone during the later part of pregnancy. Counseling that stress doses of glucocorticoids for intercurrent illness and during labor and delivery are required for both the patient and the obstetrician.

Thyroid

THYROID CANCER CASE REPORTS II

Unusual Case of Metastatic Struma Ovarii Diagnosed at the Time of Hysterectomy

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Introduction: Struma ovarii is rare ovarian teratoma composed of more than 50% mature thyroid tissue, which can rarely transform to malignancy. There are fewer than 200 cases reported and no established treatment approach. We present a case of metastatic malignant struma ovarii. **Case Presentation:** A 41 year old female was diagnosed with metastatic papillary thyroid carcinoma (PTC) after it was found on uterine histopathology. History was notable for remote left ovarian cystectomy of a mature teratoma with prominent thyroid component. She had no personal or family history of craniocervical radiation or thyroid cancer. Eight years later, she had a total hysterectomy for menorrhagia. Pathology showed subcentimeter serosal deposits of follicular variant PTC, staining positive for thyroglobulin, CK19, and HBME1. Endometrial and cervical pathology were normal; the myometrium had many leiomyomata. Post-hysterectomy TSH was 2.3 (0.34 - 5.66 μ IU/mL), and thyroglobulin 44.3 (\leq 33.0 ng/mL). Thyroid ultrasound showed a 7 mm isoechoic nodule without lymphadenopathy. Whole body PET/CT showed multiple hypermetabolic masses in the pelvic peritoneum and liver. The right ovary had many cystic lesions and was enlarged to 5.3 x 4.5 cm. She underwent partial hepatectomy, oophorectomy, salpingectomy, and omental resection with no residual disease. Pathology showed follicular variant PTC in both ovaries, peritoneum, colonic mesentery, and omentum. Thyroglobulin fell to 6.9 ng/mL 3 weeks later. A 0.2 cm focus of follicular variant PTC with capsular invasion was found on pathology after total thyroidectomy. She underwent radioiodine ablation with 150 mCi. Postablative scan showed residual activity in the thyroid bed and right hemipelvis, but no new foci of activity. CT abdomen showed resolution of perihepatic lesions and thyroglobulin declined further to 0.5 ng/mL. **Discussion:** A low risk of recurrence (7.5%) has been reported in patients with malignant struma ovarii, with survival rates of 96.7% at 5 years and 84.9% at 20 years, despite a variety of surgical and adjuvant management strategies. Unilateral cystectomy, unilateral salpingo-oophorectomy, or total abdominal hysterectomy and bilateral salpingo-oophorectomy may be sufficient for patients with well differentiated thyroid cancer arising in struma ovarii without metastases. Thyroid ultrasound should be performed to exclude primary thyroid malignancy. Patients with distant metastases may benefit from aggressive treatment including resection of gross abdominal and pelvic disease and total thyroidectomy to facilitate radioactive iodine ablation and surveillance for recurrence. **Conclusion:** Due to its rarity, there is no consensus on optimal treatment of malignant struma ovarii. More research in this field is warranted.

Adipose Tissue, Appetite, and Obesity RARE CAUSES AND CONDITIONS OF OBESITY: PRADER WILLI SYNDROME, LIPODYSTROPHY

Presence of LMNA p.R582H Pathogenic Variant in Homozygous State Demonstrates Gene Dosage Effect on the Severity of Fat Loss in Lipodystrophy

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