SUN-905

We present the case of a 31 year old female referred with weight gain, secondary amenorrhea, facial plethora and buffalo hump x6-9/12. Background medical history of caesarean section. She was on no regular medications. She rarely consumed alcohol. There was no relevant family history.

She experienced secondary amenorrhea x6/12. Her menarche was at the age of 14 and had regular periods thereafter. She had gained 13kg over the previous 9/12. Examination revealed an intrascapular fat pad, abdominal striae and facial plethora. Her BP was 150/87.

Initial investigations were as follows: Overnight DST: Cortisol 642 nmol/L, DHEAS <0.4 µmol/L, hCG <1 U/L, TSH 0.71 mIU/L, FT4 15.1 pmol/L, Prolactin 380 mIU/L, IGF-I 152 µg/L, FSH 4.4 IU/L, LH 3.0 IU/L, oestradiol <100 pmol/L, 17-OH Progesterone <1.0 nmol/L. She then underwent a low dose 48 hour dexamethasone suppression test the results showed: Cortisol Day 1(time 0) 704 nmol/L, Day 2 (time 24 hours) 702nmol/L, Day 3 (time 48 hours) 703 nmol/L and paired ACTH 1.4ng/L.

She was admitted from clinic with BP 189/107 and was started on metyrapone and ramipril. On this admission her bloods showed calcium 2.70 mmol/L, iPTH 113.5 ng/L, 25 (OH) D27nmol/L. Ct abdomen and pelvis revealed a 3.3x2.2 cm right adrenal lesion with hounsfeld units <10 and unremarkable left adrenal. ARR, plasma metanephrines and HbA1c were all normal. The case was discussed at MDM and referred for retroperitoneal laprascopic right adrenalectomy. She was discharged day 2 post op off anti hypertensives and on hydrocortisone 10mg/5mg/5mg. Histology confirmed adrenocortical adenoma and Ki67 <5%.

Synacthen test done one month post operatively showed time 0 cortisol 35 nmol/L, time 30 cortisol 56 nmol/L, time 60 cortisol 60 nmol/L and time 0 ACTH 51 ng/L. Post operatively her menses returned.

When vitamin D replete, we re-evaluated her hypercalcemia. This revealed 2.77 mmol/L, iPTH 100.7ng/L, calcium: creatinine ratio 0.72 mmol/mol. She had an ultrasound neck and sestamibi which both lateralised to right lower lobe of thyroid. A synacthen test was repeated which revealed time 0 cortisol 183 nmol/L and ACTH 44 ng/L, time 30 cortisol 258 nmol/L and time 60 cortisol 302 nmol/L. She was referred for 4 gland exploration with intra operative PTH. Her baseline intra operative PTH was 193 ng/L and her 10 minute post excision value was 55 ng/L which demonstrates a 65% drop in concentration and intra operative PTH returned to within the reference interval.

The histology was atypical displaying extension of the tumour through the capsule and possible vascular extension. Ki 67 was <2%. It has been sent to St. Guy's and Thomas' for a second opinion. Her calcium and iPTH returned to normal post operatively. MEN1 and CDKN1B genes were negative. We're awaiting gene sequencing on the following - RET, CDC73, CASR, CDKNIA, CDKN2C AND CDKN2B. This case represents a case of multiple endocrinopathies with no found genetic link.

Tumor Biology

ENDOCRINE NEOPLASIA CASE REPORTS I

Ectopic ACTH Syndrome: An Aggressive Presentation Due to Metastatic Liver Cancer of Unknown Primary

Ivan Alexander Serrano, MD[1], Sara Ahmad, MD[2], Ramona Dadu, MD[3], Paul Graham, MD[3], Steven Weitzman, MD[3].

[1]Methodist Hospital, Houston, TX, USA, [2]Baylor College of Medicine, Houston, TX, USA, [3]MD Anderson Cancer Center, Houston, TX, USA.

SUN-919

Background: Ectopic Cushing's Syndrome is a rare but often aggressive condition caused by ACTH-hypersecretion from non-pituitary tumors. In patients with metastatic cancer, as well as those with occult tumors, the diagnosis and management can be extremely challenging. **Clinical Case:** A 25-year old woman recently diagnosed with poorly differentiated metastatic liver carcinoma of unknown primary was admitted for lower extremity edema and worsening fatigue for the preceding month. Since being diagnosed with liver cancer, she developed uncontrolled hypertension, persistent severe hypokalemia and facial "puffiness". Physical exam was remarkable for moon facies and truncal obesity but no evidence of striae. An overnight 1-mg dexamethasone suppression test resulted in an elevated morning cortisol level of 93.4 mcg/dL and elevated ACTH of 299 pg/mL. A 24-hour urine cortisol was significantly elevated at 4,448 mcg/24 hours. These findings were consistent with hypercortisolism due to hypersecretion of ACTH. An MRI of the sella revealed no pituitary abnormality. A high-dose dexamethasone suppresion test (single 8 mg dose) was performed and her morning cortisol level remained elevated at 98.6 mcg/dL, consistent with ectopic ACTH secretion. She was treated for the underlying malignancy with carboplatin and paclitaxel. After a thorough discussion of therapeutic options, she was prescribed Ketoconazole with the plan to medically control the hypercortisolism potentially followed by bilateral adrenalectomy. Ketoconazole was up-titrated and Spironolactone was added resulting in significant improvement of hypokalemia and hypertension. Unfortunately, one week after discharge she was re-admitted due to worsening performance status, watery diarrhea and abdominal pain. A serum cortisol level was elevated at 124 mcg/dL and Metyrapone was added to her regimen. Unfortunately, her performance status continued to decline due to progression of cancer and uncontrolled hypercortisolism. As a result, she was deemed a poor surgical candidate for bilateral adrenalectomy. The patient's condition rapidly deteriorated and she developed malignant ascites as well as altered mental status. In accordance with her wishes, a DNR order was placed and she passed away shortly thereafter. **Conclusion:** Ectopic ACTH-syndrome is the etiology of 10–20% of cases of Cushing's syndrome. Clinical presentation is often sudden and rapidly progressive. Severe hypertension and hypokalemia are seen more commonly than in Cushing's disease. Cases secondary to occult tumors or metastatic cancer can be particularly challenging to treat when it is not possible to eliminate the source of ACTH hypersecretion via surgical or medical treatment. In patients such as this, early bilateral adrenalectomy should be considered after starting medical therapy in order to reduce morbidity and mortality due to hypercortisolism.

Thyroid

THYROID DISORDERS CASE REPORTS III

Bilateral Killian-Jamieson Diverticulum Mimicking Thyroid Nodules

Yu-Yi Lin, MD[1], Hsin-Kai Wang, MD[2], Harn-Shen Chen, PhD[3], Ching-Fai Kwok, PhD[1].