RECURRENT ADRENAL DISEASE

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Endorama 2/19/2015
SR 2412318, SC 3421561
• Category: Adrenal
• Attendings: Angelos & Grogan
36yo woman with a hx of Cushing’s Syndrome and right adrenalectomy 6/14 at OSH. Path demonstrated adrenal neoplasm of unknown malignant potential.

**PMH:** OSA, htn, asthma, hypothyroidism, pituitary mass

**Meds:** neurontin, synthroid, cytomel, ativan

**Labs:** TSH: 1.68 (0.3-4.0 mcU/mL)

**HPI:** Prior to operation had 80lb weight gain, proximal muscle fatigue, facial hair growth and acne
HPI

Initial Operation 6/2014
Laparoscopic transabdominal right adrenalectomy. Capsule of the adrenal gland was violated, 7.5cm mass intact

Pathology
reviewed at multiple institutions
• Rush & University of Michigan: adrenal neoplasm of unknown malignant potential
• University of Wisconsin, UofC & Brigham and Women’s: low grade ACC

Repeat Imaging 8/2014
Recurrence of sx: CT 3cm soft tissue density in R adrenal bed, PET negative

Recommendations
observation w q6 mo imaging
**Oncology Clinic**

MRI: 2.8x0.6cm linear enhancing soft tissue mass R adrenalectomy bed posterior to the IVC

CT chest: no evidence of metastatic disease

**Urology Clinic/GU oncology tumor board**

Surveillance, imaging q3mos
Endocrine Surgery Clinic

Repeat labs – no evidence of hormonal excess
  DHEA: 32 (45-270 ug/dL)
  Testosterone: 20 (20-60 ng/dL)
  Ca/PTH: wnl
  ACTH: 12.9 (<52 pg/mL)
  Free Urine Cortisol: 40 (3.5-45)
  metanephrines/catecholamines: normal per OSH report

Preoperative anesthesia appointment

OR for resection of soft tissue mass
OPERATIVE MANAGEMENT

OR

• R subcostal incision, medial/anterior rotation of R lobe of liver
• Excisional bx of small liver mass: bile duct adenoma
• IVC and R Renal vein identified, mass resected, adrenal tissue apparent grossly

Pathology

• Microscopic foci (largest 2mm) of residual low grade adrenocortical carcinoma
• 0/3 lymph nodes
ADRENAL CORTICAL CARCINOMA

**Epidemiology**
- 0.5-2 cases/million annually in the United States
- Bimodal age distribution: childhood, 4th decade
- Associated with LiFraumeni, Beckwith Widemann, Congenital Adrenal Hyperplasia

**Clinical Features**
- 5 yr survival 20-35% overall, 61-82% if disease confined to adrenal gland
- 50% are functional masses
- If hormonally inactive, present with GI symptoms or back pain
- Significant necrosis may cause fever and simulate infectious process
- Metastasize to liver, regional LN and lungs
**Modified Weiss Criteria**

- Mitotic rate >5 per 50 high-power fields
- Cytoplasm (clear cells comprising 25% or less of the tumor)
- Abnormal mitoses
- Necrosis
- Capsular invasion
- Calculate: 2x mitotic rate criterion + 2x clear cytoplasm criterion + abnormal mitoses + necrosis + capsular invasion

Score of 3 or more suggests malignancy

- Specificity 96%, sensitivity 100%
- Interobserver agreement was excellent (r=0.94)
**PATIENT #2**

45yo male with von Hippel-Lindau and a history of laparoscopic adrenalectomy for pheochromocytoma in 2003.

**PMH**

VHL: pheo & multiple nervous system hemangioblastomas (cervical, thoracic and lumbar spine, cerebellum), meningioma, nephrolithiasis, hypertension

**Surgical History**

Rxn hemangioblastomas x5, R temporal lobe meningioma resection, tonsillectomy, R adrenalectomy

**Family History**

brother VHL (s/p b/l adrenalectomy), mother VHL (multiple RCC, pheos, HBs, panc tumors), father lung cancer, grandmother renal cancer, grandfather liver cancer
ROS: headaches, htn

Serum metanephrines: normal

Imaging

Abdominal CT: normal bilateral adrenal glands, 1cm lymph node adjacent and medial to the right adrenal gland posterior to the IVC.

PET CT: enhanced uptake in the bilateral adrenal glands, retrocaval lymph node and 7mm focus in the pancreatic head

MRI spine: small lumbar mass not abutting the cord

CT head: large cystic mass in the medial cerebellum
PREOPERATIVE WORKUP

Indication
PET-avid masses not biochemically active, however presence of LN concerning. Options:
1. Observation: serial PET/CT every 6 months
2. Operation: completion R adrenalectomy, lymphadenectomy
   a) Alpha blockade
   b) No alpha blockade

Anesthesia
Alpha blockade: lumbar and cerebellar hemangiomas, PET active lesions, hypertension
R subcostal incision
Exposure of R kidney and vena cava
Resection of residual adrenal gland and lymph nodes
• Autosomal dominant
• Mutation in vHL tumor suppressor gene on short arm of chromosome 3
• Predisposition for tumors
  – Retinal angiomas
  – CNS hemangioblastomas
  – Clear cell renal carcinomas
  – Pheochromocytomas
  – Pancreas NET, cysts