

RADY401 Case Presentation: Adrenocortical Carcinoma

Matt Gellatly, July 20, 2020

Patient History & Initial Work-up

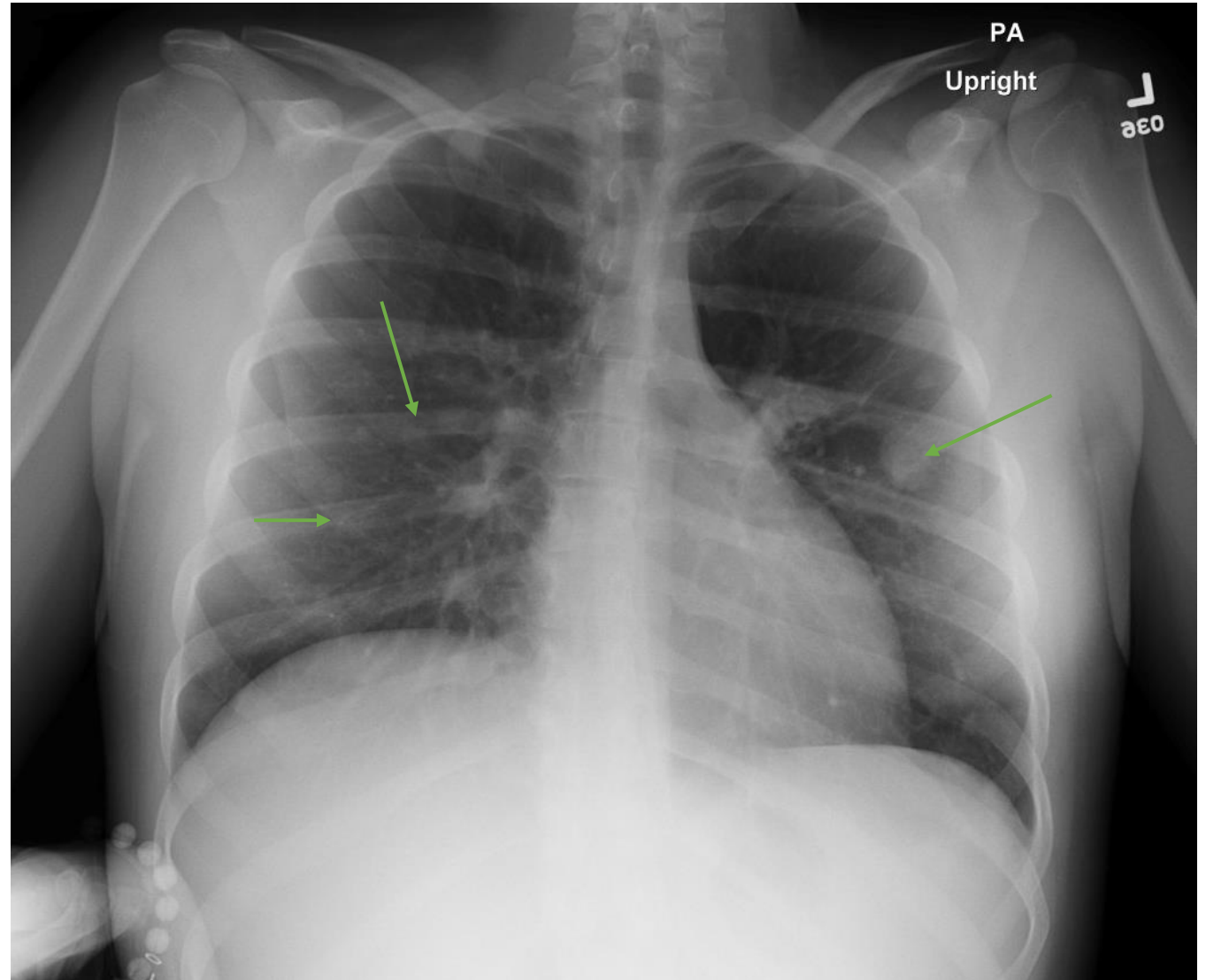
- Previously healthy 18 yo F presents with abdominal pain, chest pain, early satiety, unintentional weight loss (35 lbs)
- On physical exam, palpable abdominal mass, abnormal RUQ cystic mass on POCUS, then CT abdomen/pelvis further characterizes mass as likely from R adrenal origin
- Labs
 - Normal: cortisol, ACTH, LH, FSH, estradiol, testosterone, estrogen, prolactin, B-HCG, AFP, serum metanephrines
 - Elevated: DHEA-S, androstenedione
 - Pending (as of 7/19): VMA, DHEA, pregnenolone, 11-deoxycortisol, 17-hydroxypregnenolone
- DDX: Adrenocortical carcinoma, pheochromocytoma, adrenal hematoma, adrenal metastasis, adrenal adenoma, adrenal lymphoma

Imaging Studies from Workup

- POCUS (7/12)
- Chest X-ray (7/12)
- CT abdomen/pelvis with IV contrast (7/12)
- CT chest with IV contrast (7/12)
- 18-FDG PET/CT whole body (7/13)

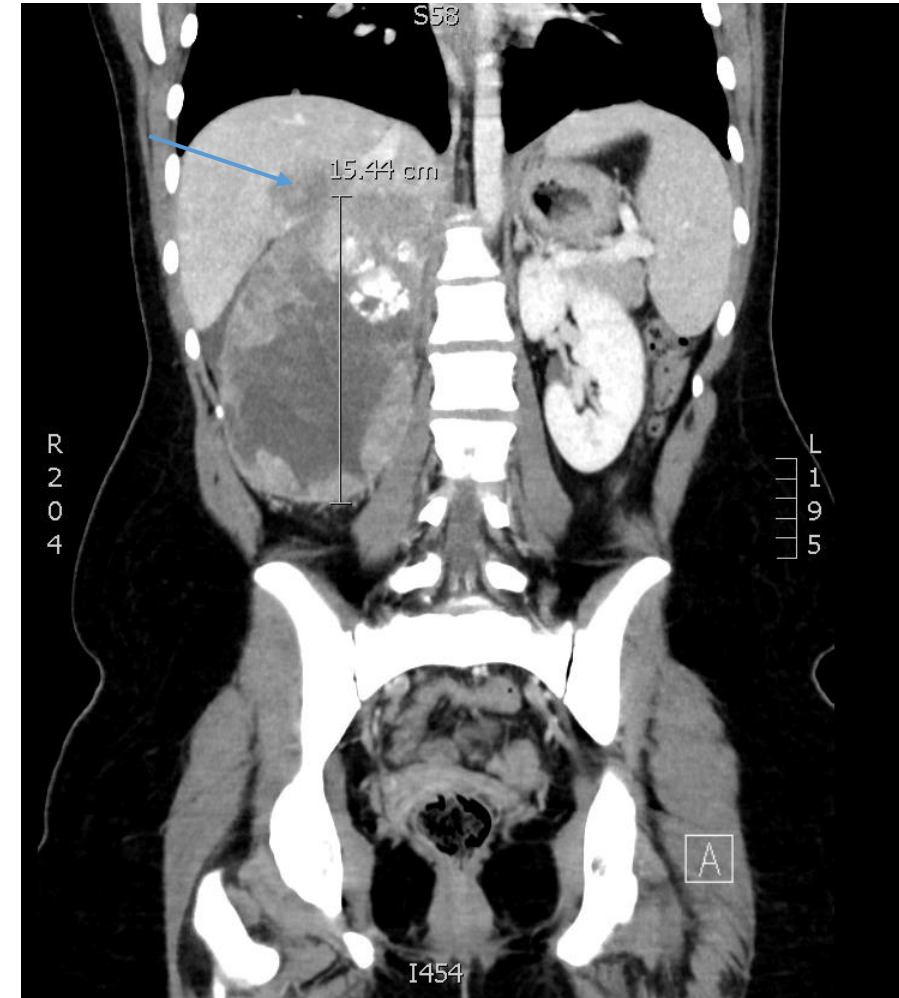
Chest X-ray

- Rounded mass in Left lung
- Multiple small lesions bilaterally



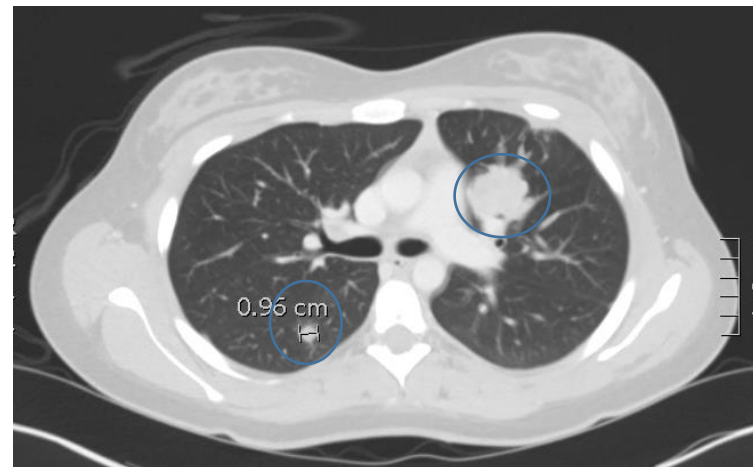
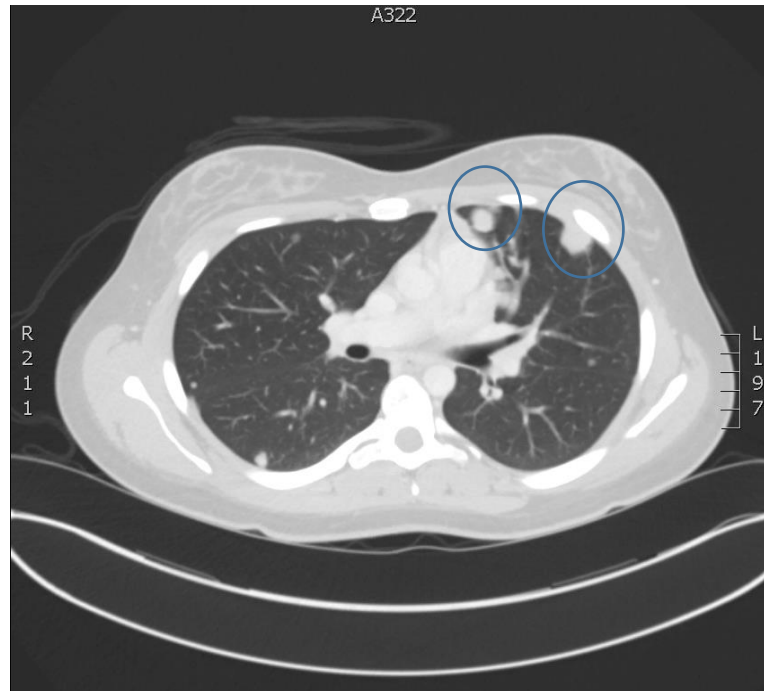
CT abdomen/pelvis with IV contrast

- Right adrenal gland
 - Heterogeneous enhancing
 - Irregular
 - Central necrotic portion (red arrow)
 - Scattered internal calcifications
 - Encasement of suprarenal and hepatic IVC
- Mass effect
 - Right kidney
 - Pancreas
 - Duodenum
 - Liver
 - Gallbladder
 - Colon
 - Small bowel
- Right hepatic lobe (blue arrow)
 - Irregular heterogeneously enhancing mass, communicating with adrenal mass
 - Effaced hepatic capsule



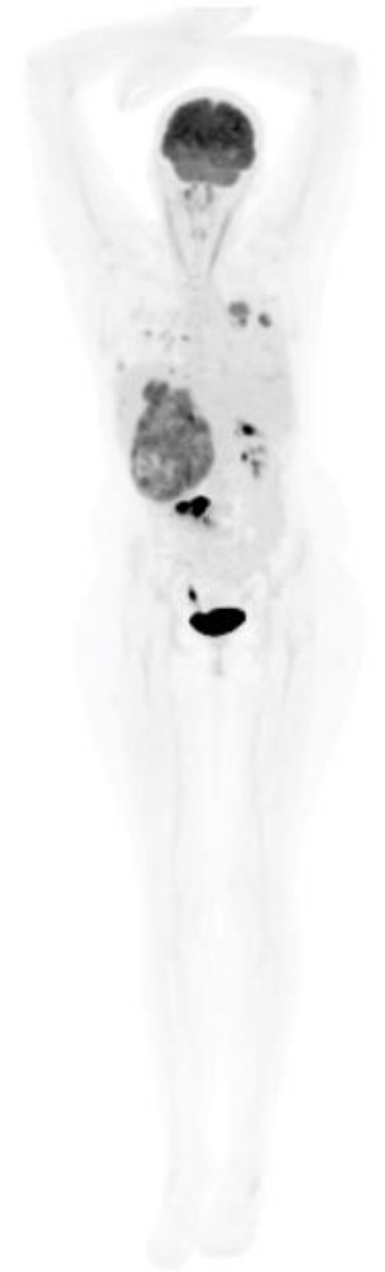
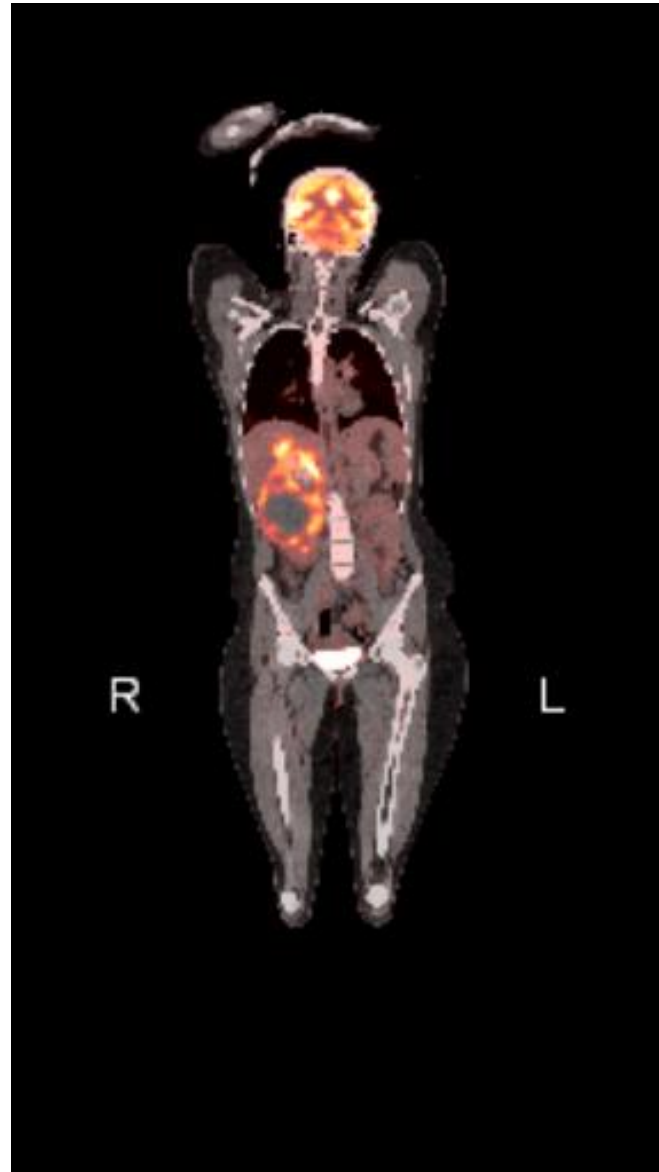
CT chest with IV contrast

- Innumerable pulmonary nodules bilaterally
- Anterior mass to left of hilum (2.7cm)
- Anterolateral L upper lobe subpleural nodule (1.7 cm)
- Posterolateral L lower lobe subpleural nodule (1.6)



18-FDG PET/CT

- FDG avidity
 - Pulmonary nodules and masses
 - Right hepatic mass
 - Adrenal mass with central necrosis
- Physiologic uptake: salivary glands, tongue base, vocal folds



Patient treatment or outcome

- Port placed by IR for chemotherapy delivery (7/14)
- Core-needle biopsies of adrenal mass obtained under ultrasound guidance (7/16)
 - FNA cannot distinguish adrenal carcinoma from benign adrenal lesion, but can differentiate adrenal tissue versus metastatic tumor
 - Critical to rule out pheochromocytoma before adrenal mass biopsy
- Enrolled in ARAR0332 Phase III clinical trial
 - Treatment of ACC with Surgery + LN dissection + Multiagent chemotherapy (cisplatin, etoposide, doxorubicin, dexrazoxane, mitotane, pegfilgrastim)
- Consult genetics to assess for neoplastic syndrome (MEN-1, Li-Fraumeni, Lynch, Beckwith-Wiedemann, Carney complex)



Typical ACC Imaging Work-up

- CT or MRI is best initial imaging procedure for this abdominal mass
- MRI advantageous in some situations for characterizing local invasion of ACC, specifically vascular invasion
- Plain abdominal radiographs not very helpful, but can show evidence of mass effect
 - Calcifications present in about 30% of ACC, but hard to see on plain films
- FDG-PET/CT useful for identifying ACC versus benign adenomas, which can sometimes present with elevated Hounsfield units (HU) or delayed washout values
- C-MTO PET can distinguish adrenocortical tumor versus noncortical lesion, but not malignant versus benign
 - Metomidate (MTO) inhibits 11-beta hydroxylase and aldosterone synthesis, with high affinity for cortical enzymes

Classic ACC Imaging Findings

- Max diameter >4 cm highly suggestive of malignancy (commonly 4-10cm)
- Additional features on CT/MRI:
 - Heterogeneity
 - Irregular borders
 - Calcifications
 - Invasion of surrounding structures
 - LN enlargement
- Cortical adenomas are typically more 'lipid-rich' than ACC
 - CT attenuation of benign adenoma usually <10 HU, suggesting nearly 100% that tumor is benign adenoma

Additional ACC Cases

Main takeaways:

1. Large size (>4 cm diameter)
2. Heterogeneous



Imaging Modality Ability to Detect Malignancy

	Sensitivity	Specificity	PPV	NPV
Non-contrast CT	100	33	72	100
18 FDG-PET/CT	87	84	85	86

Radiation/Cost of ACC Work-up

Imaging Modality	Radiation	Comparable to natural background radiation of	Cost
POCUS	None	n/a	\$233 (104-641)
CT abdomen/pelvis	10 mSV	3 years	\$1,515 (512-5,055)
CT chest	7 mSV	2 years	\$780 (440-2,464)
Chest XR	0.1 mSV	10 days	\$72 (29-472)
FDG PET-CT	7 mSV	2 years	\$2,605 (2,084-6,513)
TOTALS	24.1 mSV	7 years, 10 days	\$5,205 (3,169-15,145)

Not included: CXR #2 (worsening SOB), pre-chemotherapy echocardiogram, port placement under fluoroscopy, CT abdomen/pelvis #2 (bladder dysfunction)

Price estimates per Healthcare Bluebook (<https://www.healthcarebluebook.com/ui/consumerfront>)

UNC Top Three

- Adrenocortical carcinoma is **rare** (1-2 per 1,000,000 patients/year)
- **CT or MRI** usually distinguishes ACC from benign adrenal adenomas, but FDG-PET/CT is best when suspicion of malignancy is high
- Main features on images/clinical presentation: **large size** (>4cm diameter), **heterogeneous**, **irregular borders**, and clinically can present as functional (Cushing's syndrome) or non-functional (mass effect)

References

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