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-hydroxyprogrenolone

• 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 4-10 cm)

• 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
Main takeaways:
1. Large size (>4 cm diameter)
2. Heterogeneous
## Imaging Modality Ability to Detect Malignancy

<table>
<thead>
<tr>
<th>Imaging Modality</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>PPV</th>
<th>NPV</th>
</tr>
</thead>
<tbody>
<tr>
<td>Non-contrast CT</td>
<td>100</td>
<td>33</td>
<td>72</td>
<td>100</td>
</tr>
<tr>
<td>18 FDG-PET/CT</td>
<td>87</td>
<td>84</td>
<td>85</td>
<td>86</td>
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## Radiation/Cost of ACC Work-up

<table>
<thead>
<tr>
<th>Imaging Modality</th>
<th>Radiation</th>
<th>Comparable to natural background radiation of</th>
<th>Cost</th>
</tr>
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<tbody>
<tr>
<td>POCUS</td>
<td>None</td>
<td>n/a</td>
<td>$233 (104-641)</td>
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<tr>
<td>CT abdomen/pelvis</td>
<td>10 mSV</td>
<td>3 years</td>
<td>$1,515 (512-5,055)</td>
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<tr>
<td>CT chest</td>
<td>7 mSV</td>
<td>2 years</td>
<td>$780 (440-2,464)</td>
</tr>
<tr>
<td>Chest XR</td>
<td>0.1 mSV</td>
<td>10 days</td>
<td>$72 (29-472)</td>
</tr>
<tr>
<td>FDG PET-CT</td>
<td>7 mSV</td>
<td>2 years</td>
<td>$2,605 (2,084-6,513)</td>
</tr>
<tr>
<td><strong>TOTALS</strong></td>
<td><strong>24.1 mSV</strong></td>
<td><strong>7 years, 10 days</strong></td>
<td><strong>$5,205 (3,169-15,145)</strong></td>
</tr>
</tbody>
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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](https://www.healthcarebluebook.com/ui/consumerfront))
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


