40 yo Woman with Weight Loss

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Dr. Upala does not have any relevant financial relationships with any commercial interests
Learning Objectives

• Differential diagnosis of adrenal mass
• Review adrenocortical carcinoma
  – Presenting symptoms
  – Medical and Surgical treatment
HPI

• 40Yrs female with a past medical history of HTN, DM who presents with weight loss
• She presented 3 months ago with a chest pain to a hospital in South Illinois. Her chest pain was atypical and she was found to have high blood pressure and shingles. Her labs also was found to have a low potassium
• She was having symptoms of losing weight from 170 pounds to 127 pounds with feeling sometimes a panic attack, headache, high blood pressure and was placed on hydrochlorothiazide for that
HPI

• The patient was also diagnosed with diabetes mellitus around the same time and in the hospital she has been receiving low insulin sliding scale and sent home with metformin 500 twice daily with a good control. her A1c before was 6.1%, when she was diagnosed it was 9%
• For her low potassium, patient has been on supplements, lisinopril and Aldactone
• Patient reports rapid weight gain one year ago, along with acne and muscle weakness especially in the lower extremity. Her menses that were regular previously have ceased for the past year. She has not been on any hormonal therapy
• The patient denies any other family history of endocrine diseases except diabetes
Other History

• Past Medical History
  – HTN
  – T2DM
• Past Surgical history
  – None
• Family History
  – Father: T2DM
  – Mother: T2DM
• Allergy
  – No Known Allergies
• Social History
  – Never smoked
  – No alcohol intake
  – No recreational drugs
• Medication
  – Metformin
  – Spironolactone
  – Lisinopril
  – Potassium supplement
  – Acyclovir
Review of Systems

- Constitutional: No fevers, night sweats, appetite change, + weight loss, malaise, fatigue
- HEENT: No photophobia, +blurred vision, no pain, hearing loss, difficulty swallowing, thirst, hoarseness
- Resp: No cough, dyspnea, increased WOB
- CV: +CP, diaphoretic, palpitation, LE edema, no DOE, orthopnea, PND, palpitations,
- GI: No abdominal pain, nausea, vomiting, diarrhea, constipation
- GU: No dysuria, urgency, polyuria, hematuria
- MSK: No myalgias, joint pain, back pain
- Neuro: No syncope, No numbness, paresthesias, seizures, +tremors,
  +headaches
- Heme: No adenopathy or easy bruising/bleeding
- Endo: No heat or cold intolerance, dry skin, dry hair, hair loss
- Derm: No rashes, ulcers, abdominal striae, hirsutism, acne
- Psych: No anxiety or depression
Physical Exam

BP 155/83 | Pulse 87 | Temp 36.6 °C (97.9 °F) | Resp 16 | Ht 149.9 cm (4' 11") | Wt 57.7 kg (127 lb 1.6 oz) | SpO2 99% | BMI 25.67 kg/m2

Physical Exam:
Vital signs reviewed
Constitutional: no acute distress
HEENT: EOMI, oropharynx clear
Neck: supple, no thyromegaly, no acanthosis nigricans
Cardiovascular: regular rate, no extra heart sounds
Pulmonary/Chest: good respiratory effort, clear to auscultation bilaterally
Abdomen: bowel sounds present, soft, non-tender, no violaceous straie
Musculoskeletal: moving all extremities
Neurological: sensation intact to light touch
Skin: warm, dry
Psychiatric: not agitated
### Initial Lab

<table>
<thead>
<tr>
<th>Component</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>WBC</td>
<td>11.0</td>
</tr>
<tr>
<td>RBC</td>
<td>4.14</td>
</tr>
<tr>
<td>Hemoglobin</td>
<td>10.9 (L)</td>
</tr>
<tr>
<td>Hematocrit</td>
<td>34.1 (L)</td>
</tr>
<tr>
<td>MCV</td>
<td>82.4</td>
</tr>
<tr>
<td>MCH</td>
<td>26.3</td>
</tr>
<tr>
<td>MCHC</td>
<td>32.0</td>
</tr>
<tr>
<td>RBC Dist Width</td>
<td>13.3</td>
</tr>
<tr>
<td>Platelet Count</td>
<td>320</td>
</tr>
<tr>
<td>Mean Platelet Volume</td>
<td>10.2</td>
</tr>
</tbody>
</table>

Differential Method:
- No RBC or WBC abnormalities detected

<table>
<thead>
<tr>
<th>Component</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium</td>
<td>145</td>
</tr>
<tr>
<td>Potassium,</td>
<td>3.1 (L)</td>
</tr>
<tr>
<td>Ser/Plasma</td>
<td></td>
</tr>
<tr>
<td>Chloride</td>
<td>103</td>
</tr>
<tr>
<td>Carbon Dioxide</td>
<td>29</td>
</tr>
<tr>
<td>BUN</td>
<td>15</td>
</tr>
<tr>
<td>Creatinine</td>
<td>0.5</td>
</tr>
<tr>
<td>Glucose,</td>
<td>176 (H)</td>
</tr>
<tr>
<td>Ser/Plasma</td>
<td></td>
</tr>
<tr>
<td>Calcium</td>
<td>8.9</td>
</tr>
<tr>
<td>GFR Estimate</td>
<td>&gt;120</td>
</tr>
<tr>
<td>(Calc)</td>
<td></td>
</tr>
</tbody>
</table>

Lab Component

<table>
<thead>
<tr>
<th>Component</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total Protein</td>
<td>7.2</td>
</tr>
<tr>
<td>Albumin</td>
<td>4.2</td>
</tr>
<tr>
<td>Bilirubin, Total</td>
<td>0.3</td>
</tr>
<tr>
<td>Alk Phos, Serum</td>
<td>76</td>
</tr>
<tr>
<td>AST (SGOT)</td>
<td>19</td>
</tr>
<tr>
<td>ALT (SGPT)</td>
<td>31</td>
</tr>
</tbody>
</table>

TSH: 1.06, A1c 7.2
Imaging
Imaging

• A CT scan was done at that hospital which revealed a large left adrenal mass of 8.6×6.3 cm and urology service has recommended removal of this mass
• MRI was done as well for further evaluation which revealed a large left adrenal mass that has a big part of it does not wash out the contrast
DDX

Adrenal Mass Differential Diagnosis

- Adrenal Cortex
  - Adenoma
  - Nodular Hyperplasia
  - Carcinoma
- Adrenal Medulla
  - Pheochromocytoma
  - Ganglioneuroma
  - Ganglioneuroblastoma
- Metastases
  - Breast, Lung, Lymphoma
  - Leukemia, other
- Technical Artifacts
- Other
  - Myelolipoma
  - Neurofibroma
  - Hamartoma
  - Teratoma
  - Xanthomatosis
  - Amyloidosis
  - Cyst
  - Hematoma
  - Granulomatosis
- Pseudoadrenal
  - Renal, Pancreas, Spleen, etc
DDX?
DDX

Adrenal mass detected incidentally on CT scan or ultrasound examination

What is the size (greatest diameter) of the lesion?

- < 3 cm
  - Patient has no signs or symptoms, and screening laboratory test results are normal (see Table 5).

- > 3 to < 6 cm
  - MRI and additional endocrine evaluation

- > 6 cm
  - Surgical referral

Yes

Radiographic surveillance at 3 months, and then every 6 months for 2 years

No

Referral based on symptoms or laboratory test results
Lab

- Work Up?
Lab

- Aldosterone : 136 (<21)
- Urine Aldosterone : 128 (2-20)
- Cortisol 4 am : 38.5
- Salivary cortisol : 3230 (<100)
- Plasma metanephrines : < 0.2 (<0.50)
- Plasma normetanephrines: 0.25 (<0.9)
- Renin : 3.7 (0.6-3)
- 24 hrs urine free cortisol : 2856 (3.5-45)
- ACTH : 1 (<52)
HPI

- She has had a hormonal evaluation that reveals very elevated 24 hour UFC.
- Aldosterone level has been elevated but she had received aldactone while level was obtained.
- Her plasma metanephrine were normal, making pheochromocytoma unlikely.
Lab

1 mg overnight dexamethasone suppression test

- ACTH at baseline: 1 (<52)
- Cortisol 4 am: 40.4
- Cortisol 2 pm: 40
DDX

- Aldosterone secreting tumor
- Severe hypercortisolemia
Lab

• DHEA-S: 433.9 (60.9-337)

• Adrenal cancer
Brief History

• 40 yo female, has had a 1 year history of rapid weight gain associated with amenorrhea, muscle weakness, new onset hypertension and DM2, significant hypokalemia and biochemical work up consistent with Cushing's syndrome

• Am cortisol post low dose DEX suppression is every elevated at 40, her 24 hour UFC is very elevated at 2856 and her late evening salivary cortisol is very elevated at 3230. Plasma ACTH is suppressed at 1. This is all consistent with adrenal Cushings

• She also has a reported 8 cm L adrenal lesion that was to be resected. Considering the size of mass and abnormal wash out on CT, there is concern for adrenocortical carcinoma
Management

• Continue potassium supplement and spironolactone
• DVT prophylaxis
• Stress dose steroids during surgery and post operative exogenous glucocorticoid therapy until recovery of HPA axis
• Surgery (robotic-assisted adrenalectomy)
Pathology Report

• Left adrenal gland; robotic-assisted adrenalectomy:
  - Adrenocortical carcinoma (9.2 cm), high grade, invading into the peri-adrenal adipose tissue
  - Tumor involves the peripheral specimen
  - Tumor weight 152.8 grams
  - High grade (>20 mitosis/HPF)
  - Lymphovascular Invasion: Present
  - Regional Lymph Nodes: N/A
  - pTNM: pT3,NX
Potassium
Diabetes

Merchant et al. 2013
Epidemiology

• Estimated incidence of 0.5-2 per 106 patients per year
• Peaks of age distribution at age <5 and in the 4th and 5th decades
• Scattered reports of gene associations, but rarity of lesion limits studies – no clear associations
Clinical Presentation

- 60-65% of adrenocortical carcinomas are functioning lesions
- Cushings
- Virilization
- Feminization
- Hyperaldosteronism
Diagnosis

• Hormonal studies can be a first diagnostic test which confirms ectopic steroid hormone secretion, leading to an imaging and tissue diagnosis.
• They also can be a “tumor marker” which can be useful for monitoring response to therapy and suspicion of recurrence
Diagnosis

• 24 hour urinary cortisol excretion
• More than 90% of Cushinoid patients have free cortisol levels greater than 200mcg/24 hours
  97% of normals have levels less than 100mcg/24 hours
• ACTH measured with serum cortisol will demonstrate ACTH independent nature of hypercortisolism
Diagnosis

• Other steroids are elevated:
  – androstenediol and adrosetenedione
  – DHEA and DHEA-S
  – 11- deoxycortisol
  – urinary 17- ketosteroids
  – aldosterone

• Many intermediate enzymes are defective or dysregulated, leading to inefficient steroid production and precursor accumulation
Radiologic Findings

- CT detects 98% of adrenal carcinomas
- MRI scanning can also provide vascular invasion/tumor thrombosis information
- Many incidental findings?
- Malignant lesions tend to be > 5 cm, have irregular shapes/blurred margins, and be heterogeneously enhancing
Staging

• Hormonal studies directed at symptoms
• 24h urine studies to r/o pheochromocytoma
• CT scanning to determine extent and resectability of lesion
• MRI may clarify vascular invasion; right sided lesions have a propensity to form venous tumor emboli
### Staging

#### Adrenal cortical carcinoma TNM staging AJCC UICC 2017

<table>
<thead>
<tr>
<th>Primary tumor (T)</th>
<th>T criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>TX</td>
<td>Primary tumor cannot be assessed</td>
</tr>
<tr>
<td>T0</td>
<td>No evidence of primary tumor</td>
</tr>
<tr>
<td>T1</td>
<td>Tumor ≤ 5 cm in greatest dimension, no extra-adrenal invasion</td>
</tr>
<tr>
<td>T2</td>
<td>Tumor &gt;5 cm, no extra-adrenal invasion</td>
</tr>
<tr>
<td>T3</td>
<td>Tumor of any size with local invasion but not invading adjacent organs</td>
</tr>
<tr>
<td>T4</td>
<td>Tumor of any size that invades adjacent organs (kidney, diaphragm, pancreas, spleen, or liver) or large blood vessels (renal vein or vena cava)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Regional lymph nodes (N)</th>
<th>N criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>NX</td>
<td>Regional lymph nodes cannot be assessed</td>
</tr>
<tr>
<td>N0</td>
<td>No regional lymph node metastasis</td>
</tr>
<tr>
<td>N1</td>
<td>Metastasis in regional lymph node(s)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Distant metastasis (M)</th>
<th>M criteria</th>
</tr>
</thead>
<tbody>
<tr>
<td>M0</td>
<td>No distant metastasis</td>
</tr>
<tr>
<td>M1</td>
<td>Distant metastasis</td>
</tr>
</tbody>
</table>
Clinical Questions

• Treatment option (surgery, chemical therapy, monitor)
Treatment Overview

• Surgical resection
  – Mainstay Tx

• Chemotherapy
  – Mitotane

• Radiation
Surgical Treatment

- Complete surgical resection is the primary treatment modality

TABLE 7. Adrenocortical carcinoma: survival rates from reported series

<table>
<thead>
<tr>
<th>Study</th>
<th>Institution/group</th>
<th>Year</th>
<th>n</th>
<th>Overall</th>
<th>Complete resection</th>
<th>Incomplete resection</th>
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</thead>
<tbody>
<tr>
<td>Soreide et al.\textsuperscript{6}</td>
<td>Norway</td>
<td>1991</td>
<td>99</td>
<td>16</td>
<td>62</td>
<td>0</td>
</tr>
<tr>
<td>Icard et al.\textsuperscript{16}</td>
<td>French Endocrine Surgeons</td>
<td>1992</td>
<td>156</td>
<td>34</td>
<td>42</td>
<td>0</td>
</tr>
<tr>
<td>Zografos et al.\textsuperscript{17}</td>
<td>Roswell Park</td>
<td>1994</td>
<td>53</td>
<td>19</td>
<td>38</td>
<td>0</td>
</tr>
<tr>
<td>Haak et al.\textsuperscript{18}</td>
<td>Holland</td>
<td>1995</td>
<td>96</td>
<td>27</td>
<td>49</td>
<td>9</td>
</tr>
<tr>
<td>Crucitti et al.\textsuperscript{19}</td>
<td>ACC Italian Registry</td>
<td>1996</td>
<td>129</td>
<td>35</td>
<td>48</td>
<td>7</td>
</tr>
</tbody>
</table>

ACC, adrenal cortical carcinoma.
Surgical Treatment

• Complete resection is the strongest predictor of survival
Incomplete resection is associated with a uniformly poor prognosis, with less than a 1 year median survival

<table>
<thead>
<tr>
<th>Study</th>
<th>Institution</th>
<th>Year</th>
<th>No.</th>
<th>Median survival (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Crucitti [58]</td>
<td>Italy</td>
<td>1996</td>
<td>33</td>
<td>16</td>
</tr>
<tr>
<td>Lee [55]</td>
<td>MDACC</td>
<td>1995</td>
<td>7</td>
<td>8.5</td>
</tr>
<tr>
<td>Icard [27]</td>
<td>France</td>
<td>1992</td>
<td>28</td>
<td>&lt; 12</td>
</tr>
<tr>
<td>Icard [37]</td>
<td>France</td>
<td>1992</td>
<td>10</td>
<td>&lt; 4</td>
</tr>
<tr>
<td>Gröndal [60]</td>
<td>Sweden</td>
<td>1990</td>
<td>12</td>
<td>10</td>
</tr>
<tr>
<td>Henley [61]</td>
<td>Mayo Clinic</td>
<td>1983</td>
<td>14</td>
<td>&lt; 6</td>
</tr>
</tbody>
</table>

Patients with incomplete resections included those who underwent incomplete resection of the primary tumor and those who underwent complete resection of the primary tumor in the presence of unresectable distant metastatic disease.
Prognostic factors

• In a case review of 46 patients at MSKCC, 3 histologic factors correlated with survival:
  – Tumor > 12cm
  – 6 or more mitotic figures/10hpf
  – presence of histologic evidence of intra-tumoral hemorrhage

• 5 year survivals:
  – 0 factors: 83%
  – 1 factor: 42%
  – 2 factors: 33%
Radiation

• There are emerging case reports demonstrating improved outcomes when palliative XRT used for localized lesions
Chemotherapeutic Approach

- Mitotane
- Metyrapone (11B hydroxylase inhibitor)
- Ketoconazole
- Aminoglutehamide
Mitotane

- 1,1- dichloro-2-(o-chlorophenyl) ethane (o,p-DDD).
- Chemical relative to DDT
- Found to have adrenolytic activity in dogs in vivo (selectively destroyed the zonae reticularis and fasciculata)
Mitotane

- Inhibits the mitochondrial conversion of cholesterol to pregnenolone and the conversion of 11-deoxycortisol to cortisol (11B-hydroxylation). It produces selective adrenocortical necrosis in both the adrenal tumor and metastases
Mitotane

- Side effects are major and frequent, including:
  - CNS disturbance (vertigo, somnolence, ataxia)
  - Liver Toxicity
  - Renal Toxicity
  - Nausea, Vomiting
  - Diarrhea
  - Rash
Mitotane

Table 5. Effect of mitotane treatment on adrenal cortical cancer.

<table>
<thead>
<tr>
<th>Study</th>
<th>Year</th>
<th>Institution</th>
<th>No.</th>
<th>Result/conclusion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kasperlik-Zaluska [72]</td>
<td>1995</td>
<td>Poland</td>
<td>36</td>
<td>Suggested benefit of adjuvant mitotane</td>
</tr>
<tr>
<td>Haak [59]</td>
<td>1994</td>
<td>Netherlands</td>
<td>62</td>
<td>Response rate 21% (6/29) in setting of measurable disease</td>
</tr>
<tr>
<td>Vassilopoulou-Sellin [73]</td>
<td>1993</td>
<td>MDACC</td>
<td>13</td>
<td>No effect on survival</td>
</tr>
<tr>
<td>Pommier [26]</td>
<td>1992</td>
<td>MSKCC</td>
<td>29</td>
<td>PR 24%</td>
</tr>
<tr>
<td>Luton [74]</td>
<td>1990</td>
<td>France</td>
<td>37</td>
<td>PR 22%, no effect on survival</td>
</tr>
<tr>
<td>Venkatesh [75]</td>
<td>1989</td>
<td>MDACC</td>
<td>72</td>
<td>Stable disease or PR 29%</td>
</tr>
<tr>
<td>Karakousis [76]</td>
<td>1985</td>
<td>Roswell Park</td>
<td>10</td>
<td>Stable disease or response 40% (n = 4)</td>
</tr>
<tr>
<td>Van Slooten [77]</td>
<td>1984</td>
<td>Netherlands</td>
<td>34</td>
<td>Serum levels &gt; 14 μg/ml associated with improved survival</td>
</tr>
<tr>
<td>Henley [61]</td>
<td>1983</td>
<td>Mayo Clinic</td>
<td>24</td>
<td>PR 4% (n = 1)</td>
</tr>
</tbody>
</table>

PR: partial response.
Cytotoxics

• Various systemic cytotoxics have been used for advanced disease, usually for those failing mitotane.
• Most studied have been Etoposide, cisplatin, and adriamycin.
• Paclitaxel and Temozolamide have recently demonstrated antitumor activity in vitro
Cytotoxics

- Original studies utilized Cisplatin and Doxorubicin with Cyclophosphamide or 5-FU. RR was 20%
- Cisplatin/ Etoposide reported to have an 11% response rate
# Chemotherapy

## Table 6. Effect of chemotherapy on adrenal cortical cancer.

<table>
<thead>
<tr>
<th>Series</th>
<th>Year</th>
<th>Institution</th>
<th>Regimen</th>
<th>No.</th>
<th>Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>Williamson [83]</td>
<td>2000</td>
<td>SWOG</td>
<td>ECM&lt;sup&gt;a&lt;/sup&gt;</td>
<td>45</td>
<td>PR 5</td>
</tr>
<tr>
<td>Abraham [84]</td>
<td>1999</td>
<td>NCI</td>
<td>MEDV</td>
<td>28</td>
<td>CR 1, PR 4</td>
</tr>
<tr>
<td>Berruti [85]</td>
<td>1998</td>
<td>Italy</td>
<td>MEDP</td>
<td>28</td>
<td>CR 2, PR 13</td>
</tr>
<tr>
<td>Zidan [86]</td>
<td>1996</td>
<td>Israel</td>
<td>EP&lt;sup&gt;b&lt;/sup&gt;</td>
<td>1</td>
<td>PR 1</td>
</tr>
<tr>
<td>Bukowski [87]</td>
<td>1993</td>
<td>SWOG</td>
<td>MP</td>
<td>37</td>
<td>PR 11</td>
</tr>
<tr>
<td>Berruti [88]</td>
<td>1992</td>
<td>Italy</td>
<td>EDP</td>
<td>2</td>
<td>PR 2</td>
</tr>
<tr>
<td>Schlumberger [89]</td>
<td>1991</td>
<td>France</td>
<td>DP5-FU</td>
<td>13</td>
<td>CR 1, PR 2</td>
</tr>
<tr>
<td>Hesketh [90]</td>
<td>1987</td>
<td>Boston University</td>
<td>EPB</td>
<td>4</td>
<td>CR 1, PR 1</td>
</tr>
<tr>
<td>Johnson [91]</td>
<td>1986</td>
<td>Vanderbilt</td>
<td>EC</td>
<td>2</td>
<td>PR 2</td>
</tr>
</tbody>
</table>

<sup>a</sup>Mitotane given only after disease progression on EC and only to patients who had not received mitotane previously.

<sup>b</sup>Mitotane failure.

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5-FU: 5-fluorouracil; B: bleomycin; CR: complete response; D: doxorubicin; E: etoposide; M: mitotane; NCI: National Cancer Institute; P: cisplatin; PR: partial response; SWOG: Southwest Oncology Group; V: vincristine.
**Localized ACC** (stage I-II-III)

- **Surgery**
  - Final diagnosis/staging: pathology
  - **Complete resection R0**
    - Low-intermediate risk*
    - Follow-up or ADIUV0 trial
    - Follow-up
    - No recurrence
    - Progression
  - **Incomplete resection R1, R2, Rx**
    - High risk**

**Metastatic ACC** (stage IV)

- **Initial diagnosis**
  - Endocrine work-up
  - Chest/abdominal CT scan
  - 18FDG PET

- **Options**
  - Debulking Surgery
  - Radiotherapy
  - Loco-regional approaches + mitotane

- **Mitotane monotherapy**
- Loco-regional approaches

- **Follow-up every 2 months**
- **Stable disease/response**
  - Continue mitotane
  - Options:
    - Reconsider surgery
    - Radiotherapy
    - Loco-regional approaches

- **Follow-up**
  - **Stable disease**
  - Progression
  - **1st line chemotherapy (EDP)**

- **2nd line chemotherapy**

*Low-intermediate risk*: stage I-II, Ki 67 ≤10%

**High risk**: stage III, Ki 67 >10%
Recurrence

• Recurrences that are amenable to re-operation may be resected for long term survival
• 5 year survivals compare from 57% in those amenable to resection to 0% for those who are not
Recurrence

- MSKCC: 47 patients with recurrent/metastatic disease
- Patients who had a complete second resection had a median survival of 74 months (5-year survival, 57%), whereas those with incomplete second resection had a median survival of 16 months (5-year survival, 0%)
Hospital Course

- Patient has been seen by oncology who has recommended radiation and mitotane therapy
- She met with radiation oncology and will start radiation soon for 6 weeks. She will start on mitotane afterwards
- She is off insulin and Tradjenta. Has been checking her blood sugars and values 69-90. Prior to adrenalectomy, she was requiring low dose insulin therapy for diabetes and was also on blood pressure medication that have been discontinued
- She is only on potassium 40 meq daily. Repeat K levels have been improved and her dose of potassium has been reduced
- She has been taking the hydrocortisone 10 mg in am and 5 mg at pm.
Summary

• Adrenocortical carcinoma is a rare disease that often presents late
• Primary curative therapy is surgical
• No role for adjuvant chemotherapy has been definitively demonstrated to date
• Palliative therapy with mitotane may be useful; its palliative effect may be entirely due to adrenolytic effect
• Re-operation appears to be the only long term curative option in recurrent cases
• Cytotoxic chemotherapy in the advanced/metastatic setting has not been definitively demonstrated to be useful in controlled trials
References

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Sikarin Upala, MD, MS, 2018

Dr. Upala does not have any relevant financial relationships with any commercial interests