AMSER Case of the Month:

35-year-old man with retroperitoneal mass who had been lost to follow up

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Patient Presentation

• HPI:
  • 35 y.o. male presenting with right flank pain.
  • Admits to intermittent symptoms of hot flashes, palpitations, and diaphoresis which occur monthly and last up to 1 hour
  • 15 lbs. weight gain in last 6 months and increased fatigue

• PMH:
  • Essential hypertension
  • Lumbar fusion with chronic pain controlled with opioids
  • Outside medical report of a 3.5 cm retroperitoneal mass in 2015 (images not immediately available)
# ACR Appropriateness Criteria

**American College of Radiology**  
**ACR Appropriateness Criteria**  
**Palpable Abdominal Mass-Suspected Neoplasm**

## Variant 1: Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>CT abdomen with IV contrast</td>
<td>Usually Appropriate</td>
<td>🌟🌟🌟🌟</td>
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<tr>
<td>US abdomen</td>
<td>Usually Appropriate</td>
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<tr>
<td>MRI abdomen without and with IV contrast</td>
<td>May Be Appropriate</td>
<td>🌟🌟</td>
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<tr>
<td>CT abdomen without IV contrast</td>
<td>May Be Appropriate</td>
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</tr>
<tr>
<td>CT abdomen without and with IV contrast</td>
<td>Usually Not Appropriate</td>
<td>🌟🌟🌟🌟</td>
</tr>
<tr>
<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
<td>🌟🌟🌟</td>
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<tr>
<td>Radiography abdomen</td>
<td>Usually Not Appropriate</td>
<td>🌟🌟</td>
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<tr>
<td>Fluoroscopy contrast enema</td>
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<td>🌟🌟</td>
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<tr>
<td>Fluoroscopy upper GI series</td>
<td>Usually Not Appropriate</td>
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<tr>
<td>Fluoroscopy upper GI series with small bowel follow-through</td>
<td>Usually Not Appropriate</td>
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</table>
Findings: (unlabeled)
Findings: (labeled)

- Axial CT (A) showing well-defined, 4 cm x 7 cm, heterogeneously enhancing mass in the portocaval region (red bracket)
- Coronal view (B) showing internal hypodense/necrotic areas (green arrows)
- Mild displacement of inferior vena cava (IVC) (blue arrow) on sagittal view (C)
- Mass is separate from right adrenal gland
Findings (4 years prior)

3.5 cm mass (red bracket) noted
DDx (Based on imaging)

- Paraganglioma (PGL)
- Metastatic Tumor
- Lymphoma
- Neuroendocrine Tumor
- Other Retroperitoneal Tumor (Leiomyosarcoma)
Because of concern for pheochromocytoma, the following labs were drawn:

<table>
<thead>
<tr>
<th></th>
<th>Ref. Range</th>
<th>Date</th>
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<tbody>
<tr>
<td>Epinephrine</td>
<td>Latest Ref Range: 0 - 62 pg/mL</td>
<td>6/10/2019 16:05</td>
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<tr>
<td>Norepinephrine</td>
<td>Latest Ref Range: 0 - 874 pg/mL</td>
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<tr>
<td>Dopamine, Plasma</td>
<td>Latest Ref Range: 0 - 48 pg/mL</td>
<td>1401 (H)</td>
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<th>Ref. Range</th>
<th>Date</th>
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</thead>
<tbody>
<tr>
<td>NORMETANEPIHR., U, 24 H</td>
<td>Latest Ref Range: 82 - 500 ug/24 hr</td>
<td>6/24/2019 08:30</td>
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<tr>
<td>Metanephrine, 24 Hour Urine</td>
<td>Latest Ref Range: 45 - 290 ug/24 hr</td>
<td>1292 (H)</td>
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</tbody>
</table>

Patient was scheduled for surgery
Gross Pathology:

Retroperitoneal mass measuring 4.2 x 5.3 x 7.0 cm
Pathology Findings

- Tumor cells arranged in solid nests with increased number of mitoses (red circle)
- Zellballen pattern
  - Diagnostic for PGL/Pheo
- Tumor cells positive for synaptophysin and chromogranin A
  - * Also S100 positive (not shown)
Pathology Findings:

- “Salt & pepper” nuclei (green arrows) representing granular chromatin
- Some nuclear irregularity (small yellow arrows)
Final Dx:

Extra Adrenal Paraganglioma
Case Discussion

• Paragangliomas are rare tumors (benign) that arise from extra-adrenal chromaffin cells
  • Originate from paraganglia at a number of anatomical sites
    • Head, neck, thorax and abdomen
• Retroperitoneal paraganglioma represents between 21.5 and 87% of all extra-adrenal paragangliomas
• Characterized by secretions of excessive catecholamines
  • Epinephrine
  • Norepinephrine
  • Dopamine
Case Discussion

• Clinical symptoms include
  • Episodic hypertension
  • Tachycardia
  • Diaphoresis
  • Headache

• 40-50% of paragangliomas are non-functional/potentially functional
  • Misdiagnosis prior to removal problematic
    • Possible sudden release of catecholamines
      • Give alpha blocker prior to surgery (phenoxybenzamine, prazosin)
      • Phentolamine intraoperatively for hypertensive crisis
Case Discussion

• Hereditary Syndrome Associations:
  • Multiple Endocrine Neoplasia Type 2 (MEN-2) → RET mutation
  • von Hippel Lindau → VHL mutation
  • Hereditary PGL/Pheochromocytoma Syndromes → SDHx mutation
  • Neurofibromatosis Type 1 → NF1 mutation (Rare)

• Surgery is treatment of choice
  • Usually curative
References

