Pediatric Renal Tumors
Update from COG

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Objectives

- Epidemiology of Pediatric Renal Tumors
- Evidence based approach to Imaging Workup
- Key findings at Imaging
Background

- COG Renal tumor biology, banking, classification study (AREN03B2)
- Any patient (<30 yr) with first time occurrence of renal mass
- >4800 patients
1st 4000 Renal tumors in COG

- Median age 3.2 years (range 1 day-29.7 years)

<table>
<thead>
<tr>
<th>Tumor Type</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wilms tumor- Favorable Histology</td>
<td>75.0</td>
</tr>
<tr>
<td>Wilms tumor- Anaplastic</td>
<td>5.0</td>
</tr>
<tr>
<td>Renal Cell Carcinoma</td>
<td>4.2</td>
</tr>
<tr>
<td>Malignant Rhabdoid</td>
<td>3.7</td>
</tr>
<tr>
<td>Clear Cell Sarcoma</td>
<td>3.3</td>
</tr>
<tr>
<td>Mesoblastic Nephroma</td>
<td>2.2</td>
</tr>
<tr>
<td>Cystic Nephroma</td>
<td>2.0</td>
</tr>
<tr>
<td>Others</td>
<td>4.3</td>
</tr>
</tbody>
</table>
Histology Prevalence in 1st year of life
Histology Prevalence in 1st year of life

Congenital Mesoblastic Nephroma

Classic

Cellular

Pediatr Radiol. 2009 Oct;39(10)
Histology Prevalence Age >10yrs
Renal Cell Carcinoma

- 2\(^{nd}\) most common pediatric renal tumor
- Median age: 12.9 year (1.9-22.1 yr)
- Most common histologic subtype - Translocation morphology
- Median size: 6.4cm (0.8-19.7cm)
- T1 disease (<7cm) - ~40% LN+
Imaging Workup of Renal Mass

- Sonography 1st line of imaging
  - Vascular evaluation
- Required loco-regional Staging
  - Contrast enhanced CT or MRI
- Distant Staging
  - Chest CT
Doppler Sonography after CT/MRI?

- CT has high diagnostic accuracy
  - Detection of tumor thrombus – 83%
  - Delineation of extent of tumor thrombus – 85%
  - Cavoatrial thrombus - CT sensitivity 92%
- Routine Doppler 108/173 cases → 3 cases of thrombus at renal vein level
- Routine Doppler US after CT/MR is NOT required

Ped Blood and Cancer. 2011;58(4)
CT vs MRI for local staging

- CT and MRI have similar diagnostic performance

<table>
<thead>
<tr>
<th>Capsular Penetration</th>
<th>CT</th>
<th>MRI</th>
<th>P-value</th>
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</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>0.69</td>
<td>0.63</td>
<td>0.73</td>
</tr>
<tr>
<td>Specificity</td>
<td>0.87</td>
<td>0.84</td>
<td>1.0</td>
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</table>

<table>
<thead>
<tr>
<th>Lymph Node Metastasis</th>
<th>CT</th>
<th>MRI</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>0.77</td>
<td>0.53</td>
<td>0.22</td>
</tr>
<tr>
<td>Specificity</td>
<td>0.90</td>
<td>0.92</td>
<td>1.0</td>
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</tbody>
</table>

Ped Radiol 2015 Feb 45(2)
Detection of Contralateral Synchronous Lesion(s)

- CT and MRI have similar diagnostic performance

Post Contrast Images KEY

Ped Radiol 2015 Feb 45(2)
CT vs MRI for local staging

- CT and MRI have similar diagnostic performance
Bilateral Tumors/Bilateral Predisposition

- MRI is the preferred imaging modality

Ped Radiol 2015 Feb 45(2)
Nephrogenic Rest vs Wilms Tumor

- Differentiation limited by Imaging and by Biopsy
- Shape: Lenticular, Round
- Attenuation: Homogeneous, Heterogeneous
- Size

Radiology. 1993;188:517-521
Ped Rad. 1998;28:435-443
Diffuse Hyperplastic Perilobar Nephrogenic Rests (DHPLNR)

- Imaging diagnosis - biopsy has limited role
3 year old with renal mass

Cystic Nephroma with Pleuropulmonary Blastoma
DICER-1 germline mutation

- Cystic Nephroma
- Pleuropulmonary blastoma
- Other tumors
  - Ovarian Sertoli-Leydig tumors
  - Multinodular goiter
  - Cervical rhabdomyosarcoma

Key Points

- CT and MRI have similar diagnostic accuracy
  - MRI preferred in children with bilateral tumors or predisposition syndrome
- Routine doppler NOT required after CT/MR
- DHPLNR- “aunt Minnie”
- Renal Cell Carcinoma is the 2\textsuperscript{nd} most common pediatric renal tumor
- Cystic Nephroma with DICER-1 mutation
Thank you!

Questions/Comments
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Acknowledgements
- COG renal tumor committee
- Fred Hoffer, Sabah Servaes, Ethan Smith, Nancy Rosen