Imaging of Pediatric Renal Masses

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No Disclosures
Renal Masses in Children

Introduction

Clinically, age is an important discriminator
Pediatric Renal Neoplasms

- Medullary Carcinoma
- Multilocular Cystic Renal Tumor
- Renal Cell
- Lymphoma
- Wilms
- Clear Cell
- Rhabdoid
- Mesoblastic Nephroma

Years:
0  1  5  10  15
Renal Masses in Children

Introduction

- Imaging options
- Pseudomasses
- Predominantly solid masses
- Predominantly cystic masses
- (Some) Multifocal masses
Option: Split Bolus 370 mg/mL
50% dose, 30 sec, (venous)
50% dose, then scan (arterial)
Renal Masses in Children

• Single Mass
  – Pseudomass
  – Solid
  – Cystic

• Multiple Masses
  – Solid
  – Cystic
Renal Masses in Children

• Single Mass
  – Pseudomass
  – Solid
  – Cystic

• Multiple Masses
  – Solid
  – Cystic
Pseudomass

- Prominent pyramids
- Column of Bertin
- Fetal lobulation
- Scarring
Renal Masses in Children

• Single Mass
  – Pseudomass
  – Solid
  – Cystic

• Multiple Masses
  – Solid
  – Cystic
Renal Neoplasms in Children

Relative Percentage

- 87% Wilms
- 5% clear cell
- 2% rhabdoid
- <0.5% renal cell

RCNA 1996;6:1081
## Unique Clinical and Imaging Features of Renal Tumors

<table>
<thead>
<tr>
<th>Renal Mass</th>
<th>Clinical and Imaging Features</th>
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<tbody>
<tr>
<td>Wilms tumor</td>
<td>Large solid mass, often vascular invasion</td>
</tr>
<tr>
<td></td>
<td>Most common solid renal mass of childhood</td>
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<tr>
<td>Nephroblastomatosis</td>
<td>Multiple bilateral subcapsular lesions, associated bilateral solid Wilms tumor</td>
</tr>
<tr>
<td>Renal cell carcinoma</td>
<td>von Hippel-Lindau syndrome</td>
</tr>
<tr>
<td>Mesoblastic nephroma</td>
<td>Most common solid renal mass in newborns and infants</td>
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Unique Clinical and Imaging Features of Renal Tumors

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<tr>
<td>Multilocular cystic</td>
<td>Multicystic mass with little solid tissue</td>
</tr>
<tr>
<td>renal tumor</td>
<td></td>
</tr>
<tr>
<td>Clear cell sarcoma</td>
<td>Associated skeletal metastases</td>
</tr>
<tr>
<td>Rhabdoid tumor</td>
<td>Associated brain malignancies</td>
</tr>
<tr>
<td>Angiomyolipoma</td>
<td>Tuberous sclerosis, neurofibromatosis, von Hippel-Lindau syndrome</td>
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<td>Renal medullary carcinoma</td>
<td>Sickle cell trait or hemoglobin SC disease in adolescents</td>
</tr>
<tr>
<td>Ossifying renal tumor of infancy</td>
<td>Mass with calcification in an infant</td>
</tr>
<tr>
<td>Lymphoma</td>
<td>Appearance highly variable, frequent associated adenopathy</td>
</tr>
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</table>

Wilms Tumor

- Most common solid abdominal mass in a child
- 87% of renal neoplasms in children
- Rare in neonatal period (< 0.16%)
- 80% occur in children <5 years old
- Most sporadic; others assoc chromosome 11
  - WT 1 and WT 2 gene
- 4-13% bilateral (synchronous/metachronous)
- Distant mets most common to lungs > liver
Wilms Tumor

Increased incidence with:

- Beckwith-Wiedemann (4%) (WT 2 related)
- WAGR (WT 1 related)
- Drash (30-40%) (WT 1 related)
- Li Fraumeni
- Trisomy 18, 45X
- Nephroblastomatosis
- Synchronous, metachronous Wilms
- Hemihypertrophy
- Sporadic aniridia (30-50%)
- Other GU: Cryptorchidism, hypospadias, horseshoe kidney
Rhabdoid Tumor

- Unilateral
- Distribution
  - 80% < 2yrs
  - majority in infancy
  - majority in males
  - < 2% ped renal malignancies
- Brain tumors in 10-15%: midline, post fossa, synchronous or metachronous
- Features: subcapsular fluid, lobules separated by necrosis
Clear Cell Sarcoma

- Unilateral
- Distribution
  - 1-4 yrs
  - male predominance
  - < 5% ped renal malignancies
- Painless mass
- Bone mets (also lung, brain, liver, lymph nodes)
- Cannot distinguish from Wilms
Renal Cell Carcinoma

- < 0.5 - 2% in children
- 1:30 compared with Wilms
- 3-5% with hereditary syndromes*
- Very rare <10 yrs
- Never seen in infancy
- r/o VHL
- 2 - 4% of TS Complex*
  - Children and young adults
  - Clear cell and apillary type most common
  - Also hybrid onc/chrmophobe
  - Unclassifiable
- “Generally” smaller than Wilms
- Bleeding: hematuria more common than Wilms

## Hereditary RCC Syndromes

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Gene</th>
<th>Histologic Types of Renal Tumors</th>
<th>Incidence of Renal Cancer and Mean Age at Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>VHL disease</td>
<td><em>VHL</em> 3p25-26</td>
<td>Clear cell RCC</td>
<td>25%-45% 40 y</td>
</tr>
<tr>
<td>Hereditary papillary RCC</td>
<td><em>MET</em> 7q31</td>
<td>Papillary RCC type 1</td>
<td>Unknown &lt; 60 y</td>
</tr>
<tr>
<td>BHD syndrome</td>
<td><em>BHD1</em> 17p11.2</td>
<td>Hybrid oncocytic/chromophobe RCC Oncocytoma Clear cell RCC Papillary RCC</td>
<td>34% 50 y</td>
</tr>
<tr>
<td>HLRCC</td>
<td><em>FH1</em> 1q42-43</td>
<td>Heterogenous, but predominantly papillary RCC type 2-like</td>
<td>2%-21% 46 y</td>
</tr>
<tr>
<td>TSC</td>
<td><em>TSC1/TSC2</em> 9q34/16p13</td>
<td>AML Renal cysts Papillary RCC Clear cell RCC Oncocytoma</td>
<td>2%-4% 30 y</td>
</tr>
<tr>
<td>Hereditary paragangioma-pheochromocytoma syndrome</td>
<td><em>SDHB/SDHC/SDHD</em> 1p36/1q21/11q23</td>
<td>Clear cell RCC Medullary RCC</td>
<td>5%-15% 30 y 10-30 y</td>
</tr>
<tr>
<td>Hereditary sickle cell hemoglobinopathy and medullary RCC</td>
<td>Germline PTEN mutation Cowden syndrome</td>
<td>Clear cell RCC Papillary RCC Chromophobe RCC</td>
<td>34% 40 y</td>
</tr>
<tr>
<td>Hyperparathyroidism-jaw tumor syndrome</td>
<td><em>HRPT2</em> 1q21-32</td>
<td>Mixed epithelial and stromal tumor Papillary RCC Wilms tumor</td>
<td></td>
</tr>
<tr>
<td>BAP1 mutations and familial kidney cancer</td>
<td><em>BAP1</em> 3p21</td>
<td>Clear cell RCC</td>
<td></td>
</tr>
<tr>
<td>Constitutional chromosome 3 translocation RCC</td>
<td>Unknown chromosome 3</td>
<td>Clear cell RCC</td>
<td>Unknown</td>
</tr>
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</table>
Mesoblastic Nephroma

- “Fetal renal hamartoma”
- Most common renal neoplasm 0-6 months
- 90% under 1 yr
- Male predominance
- “Benign” solid tumor. Can occasionally appear cystic
- Surgical resection is usually curative: can be locally aggressive - wide margins.
- Lung mets have been reported
Solid Renal Neoplasms

- Wilms
- Rhabdoid tumor
- Clear cell sarcoma
- Renal cell carcinoma
- Mesoblastic nephroma
- Other
Renal Medullary Carcinoma

- Sickle cell: trait or HbSC, *not* HbSS
- Mean age 20 yrs (as young as 10 yrs)
- Arise from renal pelvis
- Central, expansile mass with caliectasis
Other (Rare) Tumors

- **Ossifying renal tumor of infancy**
  - up to 14 months; benign
  - polypoid extension into collecting system
  - ossification: ddx stone
- **Metanephric adenoma**
  - any age
  - solid
Solid Renal Neoplasms

- Wilms
- Rhabdoid tumor
- Clear cell sarcoma
- Renal cell carcinoma
- Mesoblastic nephroma
- Other: includes non neoplastic masses
Renal Masses in Children

• Single Mass
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Cystic Nephroma of Childhood
Multilocular Cystic Renal Tumor
Multilocular Cystic Nephroma

• Bimodal age and sex distribution
  – Boys 3 mos – 4 yrs
  – Girls 4 – 20, women > 40

• Painless mass

• Well-circumscribed lesion containing variable sized cysts surrounded by fibrous capsule

• Septa can enhance

• No excretion into cysts

• Differentiation from partially diff nephrobalstoma difficult: excise
Lymphatic Malformation
Renal Masses in Children

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Bilateral Multifocal Renal Masses: Solid

- Nephroblastomatosis
- Lymphoma/Leukemia
- Angiomyelipomas
- Metastases
- Pyelonephritis, other infection
- Vasculitis
- Infarcts (assoc. with sickle cell disease)
Nephroblastomatosis

- Metanephric blastema rests persisting > 36 wks
- 1 % normal infants have, give rise to 30-40% of Wilms; present in 99% bilateral Wilms
- Nephrogenic rests may be intralobar, perilobar panlobar (or panlobar = intralobar and perilobar)
- **Intralobar:** sporadic aniridia, DRASH syndrome
  - earlier age of Wilms, metachronous tumors
- **Perilobar:** Beckwith-Weideman, hemihypertrophy, trisomy 18
  - synchronous tumors
Lymphoma

Most common solid multiple renal mass in older children
Pyelonephritis
Angiomyolipoma

- Most common benign renal tumor
- Most sporadic
- Tuberous sclerosis: in 40-80%
- 80% with TS develop by 10 yrs
- Asymptomatic or painful hemorrhage
- DX: fat (eg bright on US)
- Rarely locally aggressive
- Can invade IVC (rare)
- **Lesions > 4cm: intervene** (hemorrhage risk)
Renal Cystic Disease DDX

- Multicystic dysplastic kidney
- Juvenile nephronophthisis/medullary cystic disease
- Autosomal recessive polycystic kidney disease
- Autosomal dominant polycystic kidney disease
- Glomerulocystic disease
- Cystic dysplasias associated with syndromes
- Simple (cortical) cysts
- Consider: obstructive lesions (UPJ, UVJ, PUV)
Renal Masses in Children

**Conclusions**

- Variety of useful modalities
- Technique is important (ie pseudomass)
- Imaging features can helpful in determining etiology
- Clinical history is vital