Hi #MedTwitter

A recent case discussion on #ASPNeph webinar #kidneymass in children intrigued me. Here's what I learned.

#Nephtwitter #tweetorial

Do you know that most of the intra-abdominal masses originate from kidneys in children! (Potisek et al 2017)

Kidney masses

- accounts for 60% of abdominal masses
- mostly benign and urologic in origin (Nelson 2018)

What is the first step when you recognize a kidney mass in children?
It is important to exclude pseudo mass/tumor

Most common causes-

- Congenital anomalies: Prominent renal columns of Bertin, dromedary humps
- Inflammatory: Focal pyelonephritis, abscess
- Vascular: Renal artery aneurysm, AV fistula
- Trauma/Hematoma
Once pseudo masses are ruled out, differentiating benign from malignant is important. Cystic masses—usually benign. Imaging differentiates solid and cystic masses. Some cysts can be malignant. Bosniak Classification is helpful and is reliable in children.

Which of the following has high malignant potential?
Bosniak classification of renal cysts

1. ~0% are malignant
2. ~0% are malignant

2F. ~5% are malignant
3. ~50% are malignant
4. ~100% are malignant

Ans: D. Bosniak 3
How to differentiate if a mass is originating from the kidney or not?

Ans: Claw sign

📍Determines if mass arises from solid structures
📍Refers to sharp angles on the either side of mass, formed by the surrounding normal parenchyma

<table>
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<th>Class</th>
<th>Current Bosniak Classification</th>
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<tr>
<td>I</td>
<td>Hairline-thin wall; water attenuation; no septa, calcifications, or solid components; nonenhancing.</td>
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| II    | Two types:
1. Few thin septa with or without perceived (not measurable) enhancement; fine calcification or a short segment of slightly thickened calcification in the wall or septa  
2. Homogeneously high-attenuating masses ≤ 3 cm that are sharply marginated and do not enhance |
| IIF   | Two types:
1. Minimally thickened or more than a few thin septa with or without perceived (not measurable) enhancement that may have thick or nodular calcification  
2. Intrarenal nonenhancing hyperattenuating renal masses > 3 cm |
| III   | Thickened or irregular walls or septa with measurable enhancement |
| IV    | Soft-tissue components (ie, nodule[s]) with measurable enhancement |

Silverman et al, Radiology, Vol. 292, No.2
What is the most common benign kidney tumor in neonates?

https://radiopaedia.org/articles/claw-sign-mass?lang=us
Mesoblastic Nephroma

- Most common neonatal kidney mass
- M>F
- polyhydramnios, hydrops & preterm birth
- US: the “ring” sign- concentric hyper & hypoechoic rims surrounding tumor
- Path: interlacing fibro/myofibroblastic cell bundles interspersed with collagen & vessels

What is the most common heritable benign tumor?

Ans: Angiomyolipoma

- Rare Benign renal masses
- 80% with syndromes: TS, VHL, SW, NF, ADPKD
- Macroscopic fat in CT/MRI
- Asymptomatic & <4cm → biannual/annual follow-up with US/CT
- Symptomatic or ≥4cm or B/L → surgery or arterial embolization
Other benign tumors are:

- Metanephric stromal tumors
- Metanephric adenoma
- Cystic nephroma
- Ossifying Renal Tumor of Infancy (ORTI)
- Reninoma

A 2.5 years old boy was brought to the pediatrician because mother noticed right sided abdominal distension. He was asymptomatic. What is the most common kidney tumor in the first decade?
Ans: Wilms tumor

- 90% of childhood kidney cancers
- 2-5 yrs of age
- 90% Sporadic
- 10% Hereditary: WAGR (WT1), Denys-Drash (WT1, 11p13), Beckwith Wiedemann (WT2, 11p15), Perlman, & Sotos
- Unilateral - 75-95%
- Symptoms: Abdominal pain, distention, HTN, malaise or hematuria

- Imaging: Large, well defined intrarenal mass with uniform echogenicity in the US (often with necrosis, old hemorrhage, & Calcification ~15%

- Path: Triphasic appearance - stromal, epithelial, & blastemal elements

- Rx: Nephrectomy + adjuvant chemotherapy
Figure 1. Wilms tumor in a 12-week-old infant. (a) Bivalved nephrectomy specimen shows a yellow-tan soft cut surface with small foci of hemorrhage. Arrow = adjacent kidney. (b) Photomicrograph shows the triphasic pattern of Wilms tumor. Nests of small round to ovoid hyperchromatic cells represent the blastemal component (B). Epithelial elements form tubular and glomeruloid elements (arrowheads). Surrounding these components is an immature spindle cell stroma (S). (Hematoxylin-eosin [H-E] stain, original magnification, ×40.) (c) Longitudinal US image shows a large circumscribed tumor of fairly homogeneous echotexture (asterisk), which is slightly hyperechoic compared with the cortex of the adjacent kidney (arrow).

A 14 year old AA boy presents with painless gross hematuria. Which of the following associations should concern you?

<table>
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<th>Stage</th>
<th>Criteria</th>
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| Stage I | Confined to kidney  
Complete excision with renal capsule intact and negative resection margins  
Lymph nodes negative for Wilms tumor spread |
| Stage II| Regional extension beyond kidney capsule, but confined to flank  
May include:  
Tumor penetration through capsule but confined to Gerota’s fascia  
Infiltration into renal vein  
Complete excision with negative resection margins  
Lymph nodes negative for Wilms tumor spread |
| Stage III| Residual tumor, but confined to abdomen  
May include:  
Regional lymph node involvement  
Peritoneal contamination:  
Biopsy  
Pre- or intraoperative tumor rupture  
Tumor growth through peritoneal surface  
Positive resection margins |
| Stage IV| Distant metastases: Lung, liver, bone, brain |
| Stage V | Involvement of bilateral kidneys at diagnosis |

Adapted from Davidoff (2012) [4].

Ans: D - s/o renal medullary carcinoma

- Most common tumor of second decade
- Sickle cell trait or heterozygous sickle cell disease
- Painless hematuria, flank pain & palpable mass
- Overexpression of transcription factor E3 (TFE3): Xp11.2 translocation involves gene fusion

![Image of CT scan showing renal medullary carcinoma](image_url)

**Fig. 1A** — 17-year-old boy with renal medullary carcinoma. CT image shows right upper pole renal medullary carcinoma. Mass (arrow) is infiltrative, and kidney retains its reniform shape.

**Renal Medullary Carcinoma: CT and MRI Features**
Netta M. Blitman, Robert G. Berkenblit, Alla M. Rozenblit, and Terry L. Levin
American Journal of Roentgenology 2005 185:1, 268-272
Malignant tumors are rare in children (6-7% of all childhood tumors)

Other malignant kidney tumors are-
- Clear cell sarcoma
- Nephroblastomatosis
- Rhabdoid tumor

That’s All Folks...
For case-based discussion on this topic logon to @ASPNeph December radiology webinar #FellowFOAMgroup @drM_sudha @RoshanPGeorgeMD @priti899 #pediatricnephrology
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