Radiologic Diagnosis of Wilms Tumor

Adam Friedman
Gillian Lieberman, MD
March 2008
Patient presentation

- Two year old male with abdominal pain, swelling and constipation.
Our patient: RUQ mass on abdominal plain film

Study: Abdominal X-Ray

Findings:

- Dilated large and small bowel, leftward displacement
- Soft tissue density in right upper and lower abdomen obscuring lower liver margin
Our patient: Heterogeneous mass on ultrasound

**Study:** RUQ ultrasound

**Findings:**
- Heterogeneous, septated, soft tissue mass, 9 x 10 x 6cm, with solid and cystic components (anechoic regions with enhanced through-transmission).
- Doppler demonstrates regions of vascular flow in addition to cystic elements.
Differential diagnosis

- Wilms Tumor
- Neuroblastoma
- Hepatoblastoma
- Nephrogenic rests or nephroblastomatosi (multifocal or diffusely bilateral nephroblast remnant)
- Other abdominal soft tissue masses:
  - Rhabdomyosarcoma
  - Lymphoma
  - Renal cell carcinoma (adults)
Our patient: Heterogeneous mass on axial CT

mass

r. kidney

l. kidney
Our patient: Heterogeneous mass on coronal CT

- mass
- spleen
- r. kidney
- l. kidney
Our patient: Summary of findings on CT

Study: Abdominal CT with oral and IV contrast, late arterial phase
Findings:
- 10.5 x 9.3 x 12.2 cm cystic heterogeneous mass within the right hemiabdomen, originating from upper/middle poles of kidney
- Mass-effect shift of liver
- Narrowing and deviation of infrahepatic IVC
- “Claw sign” of engulfment by right kidney
- No evidence of renal vein or IVC thrombosis
- No evidence of pulmonary metastatic disease
Diagnosis: Wilms Tumor

• Wilms Tumor, with cystic elements and favorable histology.
• Stage III due to lymph node metastases and tumor rupture noted intra-operatively, with adherent liver metastases
Radiologic features of Wilms Tumor

- "Claw Sign": Concavity of the renal contour with renal parenchyma cupping the tumor/cyst
- Clawing suggests organ of tumor origin.
- Major differential is neuroblastoma, a neuroendocrine tumor of neural crest origin):
  - Neuroblastoma: Displacement of kidney, adrenal origin, encasement of IVC/aorta, 90% calcifications
  - Wilms: Engulfment of kidney, origin from renal parenchyma, displacement of vessels, 15% calcification
Neuroblastoma vs. Wilms Tumor

**Neuroblastoma**
- Displacement of kidney
- Adrenal origin
- Encasement of IVC/aorta
- 90% calcifications

**Wilms Tumor**
- Enulfment of kidney
- Origin from renal parenchyma
- Displacement of vessels
- 15% calcification
Wilms Tumor: embryology, epidemiology, biology

- Derived from remnant rests of embryonic nephroblastic cells
- Associated with WT1 transcription factor tumor suppressor
- 6% of all childhood cancers, 1:10,000 incidence, with peak incidence between 2-5
- Cure rates ~85%
- Associated syndromes include (WAGR), Beckwith-Wiedemann, Denys-Drash, horseshoe kidney
- 5% bilateral, 5% metastatic (lung, lymph nodes, liver); 6% extend into IVC or RV
- Nephrogenic rests (intralobar or perilobar) increase risk for WT (but only ~1%)
- Histologically: blastemal, stromal, epithelial cells
Wilms Tumor: the role of imaging

- **Ultrasound:** Hypervascular, heterogenous mass
- **CT:** solid, heterogenously enhancing, solitary renal mass (can be bilateral); calcifications in <20%
- **MRI:** Hypointense on T1, hyperintense on T2, with heterogeneous enhancement

- CT used for staging due to superior resolution of lung metastases (vs. bone scanning for neuroblastoma)
- U/S monitoring of high risk (syndromic) patients every 3-4 mo until 7 yo
Wilms Tumor: staging and therapy

• Staging:
  • I - Unilateral, intact renal capsule, total excision
  • II - Regional tumor extension, total excision
  • III - Residual tumor, confined to abdomen
  • IV - Metastases (lung, liver, bone, brain)
  • V - Bilateral

• Treatment:
  • Nephrectomy (if IVC not involved)
  • Chemotherapy (vincristine/actinomycin D, +/- doxorubicin, etoposide, carboplatin)
  • Preoperative chemotherapy in Europe for down-staging, vs. post-op in US.
  • Radiation (<30%)
  • Routine U/S monitoring
Advanced imaging modalities in Wilms Tumor

- 3D MRI tumor perfusion mapping
- DWI/ADC MRI mapping
- FDG-PET/CT
Preoperative 3D mapping of tumor perfusion

- Maximum contrast (absolute) or maximum slope of contrast enhancement during MRI used to correlate areas of necrosis and perfusion of nephroblastomas or neurblastomas for surgical planning.
Apparent diffusion coefficient (ADC) mapping

- Contrast-enhanced T1 MRI vs. ADC map
- Rim-enhancement and interior architecture prominent with ADC map
- ADC mapping may indicate early tumor response, before clear tumor shrinkage (late finding)
FDG-PET/CT

- Companion patient #6 with renal cell carcinoma
- $^{18}$F-fluorodeoxyglucose positron emission tomography (FDG-PET) imaging reveals areas of increased metabolic activity
- Developing utility in Wilms Tumor:
  - Targeting biopsy to most aggressive elements
  - Monitoring treatment response
  - Identification of metastatic lung foci
Summary

• Discussion of 2 y.o. boy with Stage III Wilms Tumor
• Radiologic diagnosis of WT vs. neuroblastoma
• Utility of “claw sign”
• Review of Wilms Tumor embryology, biology
• Advanced radiologic tools for WT diagnosis and tracking
Acknowledgments and References

Acknowledgments:

• Jay Pahade, M.D.
• Gillian Lieberman, M.D.

References:

• Ros, P. (eds), Mortele, K., Pelsser, V., Lee, S. (2007) CT And MRI of the Abdomen And Pelvis: A Teaching File, Lippincott Williams & Wilkins