Diagnosis: Imaging and Clinical Features of Wilms Tumor (including nephroblastomatosis and bilateral Wilms)
Geetika Khanna, MD, MS

1. Which of the following is an indication for pulmonary radiation in children with Wilms tumor?
   A. Lung metastasis at presentation
   B. Lung metastasis at week 6 after chemotherapy
   C. Hilar lymphadenopathy
   D. Mediastinal lymphadenopathy

Correct Answer: B. Lung metastasis at week 6 after chemotherapy

References:

2. Which of the following is the most common site of distant metastasis in Wilms tumor?
   A. Brain
   B. Bone
   C. Lung
   D. Liver

Correct Answer: C. Lung

References:

Surveillance Imaging in Patients Predisposed to Renal Tumors
Lisa J. States, MD

3. Choose the appropriate surveillance protocol for children with predisposition to Wilms tumors:
   A. Ultrasound every six months alternating with MRI
B. Ultrasound with clinical signs or symptoms of tumor such as hypertension, abdominal mass or hematuria
C. MRI every 3 months
D. Renal Ultrasound every 3 months from Birth or diagnosis until 7th birthday

Correct Answer: D. Renal Ultrasound every 3 months from Birth or diagnosis until 7th birthday.

References:

4. The risk of Wilms tumor in BWS and Hemihypertrophy is:
   A. 20%
   B. 4%
   C. 50%
   D. 90%

Correct Answer: B. 4%

References:
Kalish, JM et al., Surveillance Recommendations for Children with Overgrowth Syndromes and Predisposition to Wilms Tumors and Hepatoblastoma. Clin Cancer Res; 23(13); e115-22. 2017

Rare Renal Tumors of Childhood: Imaging and Clinical Features
Meryle Eklund, MD, FAAP

5. Which of the following pediatric renal neoplasms has the highest predilection for lymphatic and osseous metastasis?
   A. Wilms tumor
   B. Clear cell sarcoma
   C. Ossifying renal tumor of infancy
   D. Synovial cell sarcoma

Correct Answer: B. Clear cell sarcoma

Rationale: The most common sites for metastatic disease in clear cell sarcoma are lymph nodes (30%) and bone (13-20%). Osseous metastases may be lytic, sclerotic, or mixed. Options B, C, and D are not correct. Wilms tumor may metastasize to lung (85% of cases) or liver (20%), but rarely metastasizes to bone. Synovial cell sarcoma may present with subcapsular hemorrhage or renal vein invasion, but lymphadenopathy is not a typical feature. Osseous metastasis has not been reported in cases of ossifying renal tumor of infancy, a benign tumor.

References:

6. Regarding congenital mesoblastic nephroma, which of the following features is associated with more aggressive disease?

A. Age < 3 months at presentation
B. Presence of “ring” sign on ultrasound
C. Negative margin at resection
D. Cellular subtype

Correct Answer: D. Cellular subtype

Rationales: Older age at presentation (> 3 months), positive margin at resection, and cellular subtype are more aggressive features of mesoblastic nephroma. The cellular subtype has a chromosomal translocation identical to infantile fibrosarcoma, is sensitive to chemotherapy, and has a good overall survival, but is more aggressive than the classic subtype with the possibility of local organ invasion and metastatic disease. The “ring” sign on ultrasound is more commonly seen with classic subtype.

References: