Wilms Tumor - Multifocal
Joseph Junewick, MD FACR
11/25/2012

History
4 year old male with abdominal pain.

Discussion
Bilateral and multifocal Wilms tumors may have a genetic predisposition. At least 10 genes have been associated with Wilms tumor. WT1 gene is located on the short arm of chromosome 11 (11p13). WT1 encodes a zinc finger transcription factor that is critical to normal development of the kidneys and gonads but also acts as a tumor suppressor. The WT1 protein likely affects cell growth, differentiation, and/or apoptosis. Germline mutations in WT1 have been found in about 2% of phenotypically normal children with Wilms tumor. Germline WT1 mutations in children with Wilms tumor does not necessarily confer a poor prognosis. The offspring of those with germline mutation in WT1 may also be at increased risk of developing Wilms tumor. WT1 mutation is more common in those children with Wilms tumor and one of the following: WAGR syndrome, Denys-Drash syndrome, or sporadic aniridia. Genitourinary anomalies, including hypospadias and cryptorchidism. Bilateral Wilms tumor. Unilateral Wilms tumor with nephrogenic rests in the contralateral kidney.

Findings
CT-Axial and multiplanar reformatted images of the abdomen show multiple left intrarenal masses of varying sizes with perirenal rupture and/or hemorrhage. No renal vein invasion, adenopathy or distant metastatic disease.

Reference
Ruteshouser EC, Robinson SM, Huff V. Wilms tumor genetics: Mutations in WT1, WTX, and CTNNB1 account for only about one-third of tumors. Genes Chromosomes and Cancer (2008); 47(6):461-470.
Disclaimer
This teaching site is partially funded by an educational grant from GE Healthcare and Advanced Radiology Services, PC. The material on this site is independently controlled by Advanced Radiology Services, PC, and GE Healthcare and Spectrum Health have no influence over the content of this site.

Content Download Agreement
The cases and images on this website are owned by Spectrum Health. Permission is granted (for nonprofit educational purposes) to download and print materials to distribute for the purpose of facilitating the education of health professionals. The authors retain all rights to the material and users are requested to acknowledge the source of the material.

Site Disclaimer
This site is developed to reach healthcare professionals and medical students. Nothing this site should be considered medical advice.

Only your own doctor can help you make decisions about your medical care. If you have a specific medical question or are seeking medical care, please contact your physician.

The information in this website is provided for general medical education purposes only and is not meant to substitute for the independent medical judgment of a physician relative to diagnostic and treatment options of a specific medical condition.

The viewpoints expressed in these cases are those of the authors. They do not represent an endorsement. In no event will Advanced Radiology Associates, PC, Spectrum Health Hospitals (Helen Devos Children's Hospital) or GE Healthcare be liable for any decision made or action taken in reliance upon the information provided through this website.