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Pediatric Renal Tumors

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Pediatric Renal Tumors

Christopher S. Cooper, MD

INTRODUCTION

Pediatric renal tumors are rare relative to adult tumors, and the most current methods of treatment may not be familiar to many urologists. Active research and advances in treatment of pediatric renal cancers continues to change urologic practice. A basic understanding of the pathology of these tumors, standard evaluation techniques, and risks and benefits of various treatment options helps the urologist to determine the treatment approach that will likely yield the best results for a particular patient.

This manual considers Wilms' tumor as well as other kidney tumors, including mesoblastic nephroma, multicystic dysplastic kidney, and angiomyolipoma. Case-based discussions are used to highlight issues related to pathophysiology, patient presentation and evaluation, treatment options, and patient outcomes.

WILMS' TUMOR

CASE 1 PRESENTATION

Patient 1 is a 2-year-old boy who initially presented to his primary care physician with a history of gross hematuria. He was treated with antibiotics for a presumed urinary tract infection, and the hematuria resolved. A voiding cystourethrogram revealed neither reflux nor bladder or urethral abnormalities.

The patient presents 1 month later with a history of recurrent gross painless hematuria and is referred to a urologist for evaluation. There is no other significant past medical, family, or surgical history. On physical examination, the patient is afebrile with a pulse of 113 bpm and a blood pressure of 127/65 mm Hg. The examination is remarkable for a palpable 5-cm mass in the right upper quadrant of the abdomen. The results of the physical examination are otherwise normal, and no adenopathy is detected.

- What imaging studies are appropriate in the evaluation of patient 1?

- What benign and malignant renal masses should be included in the differential diagnosis of patient 1?

INITIAL EVALUATION

Evaluation of Gross Hematuria in Children

Any patient—child or adult—with a history of hematuria should undergo urinalysis to confirm the presence of erythrocytes in the urine and to exclude infection, proteinuria, or casts suggestive of medical renal disease. However, the imaging evaluation of children differs from that of adults. Because children are much less likely than adults to be affected by neoplasm, intravenous pyelogram (IVP) and cystoscopy—components of the standard evaluation in adults—are not routinely recommended in children.

Causes of hematuria in children include infection, medical renal disease, and congenital anomalies, as well as neoplasm. If the bleeding occurs only as spots in the underwear, the history is consistent with idiopathic anterior urethritis (urethrorrhagia), which is a benign, self-limited process and requires no further evaluation.¹ In a child with gross painless total hematuria, a renal and bladder ultrasound should be obtained. A voiding cystourethrogram also should be obtained to evaluate the bladder and urethra.²

If a solid renal mass is detected on ultrasound, Wilms' tumor is often suspected. Wilms' tumor characteristically demonstrates a heterogeneous echo pattern on ultrasonography and can vary from predominately cystic to solid. Ultrasonography often permits assessment of the renal vein and inferior vena cava for the presence of tumor thrombus. If IVP is performed, an intrarenal mass may be manifested by distortion of the calyceal morphology. The use of IVP for evaluating patients with suspected Wilms' tumor has declined, however, because of the increased availability of ultrasonography, computed tomography (CT), and magnetic resonance imaging (MRI).

MRI accurately evaluates the extent and size of renal tumors, including Wilms' tumor, which often gives variable signal intensities. Areas of hemorrhage show increased signal intensity on T1- and T2-weighted pulse sequences. Regions of necrosis demonstrate a decreased signal on T1- but not on T2-weighted pulse sequences.³