Pediatric Urinary System Neoplasms
An Overview and Update

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KEYWORDS

• Pediatric • Neoplasms • Urinary system

KEY POINTS

- Pediatric urinary system neoplasms may be stratified by the age of the child. In the neonatal period, the most common tumor is mesoblastic nephroma; in the first decade, Wilms tumor; and in the second decade, Wilms tumor and renal cell carcinoma occur with equal frequency.
- Wilms tumor is by far the most common renal malignancy of childhood, but many of the neoplasms formerly termed *variant Wilms* represent distinct entities.
- Pediatric and adult renal cell carcinoma differ substantially in their subtype, behavior, and association with cancer syndromes.
- Common mimics of pediatric urinary system neoplasms include focal infection, localized cystic renal disease, abscess, and renal infarction.
- Evolving treatment paradigms emphasize nephron- or bladder-conserving surgery, neoadjuvant chemotherapy, and possibly radiotherapy. With these changes, the role of the imaging shifts from presurgical planning to diagnosis that directs management before tissue confirmation.

INTRODUCTION

The classification and therapies of pediatric urinary system neoplasms are constantly evolving. Whereas previously many renal neoplasms were considered variants of Wilms tumor, increasingly sophisticated histopathologic investigation has refined and improved our understanding of these varied renal neoplasms. The clinical presentation of malignant renal masses is often nonspecific, overlapping with benign or even non-neoplastic causes. Because biopsy of renal lesions is rarely undertaken in the pediatric population due to of the risk of upstaging a malignancy, the presurgical distinction of a malignant versus benign lesion is of critical importance. Additionally, given the increasing use of neoadjuvant chemotherapy as advocated by the International Society of Pediatric Oncology, diagnostic radiology is called to make presumptive diagnoses that guide treatment.

The goal of this article is to provide an up-to-date review on neoplasms involving the urinary system, including the kidneys and bladder, as well as their mimics, with an emphasis on characteristic imaging findings and differential diagnostic considerations. Such improved understanding has a great potential to reach a timely and accurate diagnosis that, in turn, can lead to optimal pediatric patient care.

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SPECTRUM OF RENAL NEOPLASM AND MIMICS

Benign Renal Neoplasms

Congenital mesoblastic nephroma

Congenital mesoblastic nephroma (CMN) is a renal tumor of newborns and infants. Although CMN is the most common renal tumor of the neonatal period, it remains rare overall, accounting for 3% to 6% of pediatric renal neoplasms.[1] Nearly 90% of affected pediatric patients present before 1 year of age,[2] and they may be diagnosed as early as the second trimester.[3] A slight (1.5:1.0) male predominance has been reported.[4] Although CMN is increasingly diagnosed in utero, it most frequently presents as a painless abdominal mass postnatally. There is an increased incidence (71%) of perinatal complications, including preterm delivery, hydrops, and polyhydramnios, with several neonates exhibiting hypertension.[5]

Two subtypes of CMN are currently described, classic and cellular, with substantial histologic, imaging, and prognostic distinctions. On gross inspection, the classic type is solid and unencapsulated, sometimes infiltrating the renal hilum. In contrast to the cellular subtype, the classic type is larger and may demonstrate necrosis, hemorrhage, or cystic changes.[1] Histologically, the classic CMN is composed of interlocking fibroblastic and myofibroblastic cells, whereas the cellular variant shows dense cellular proliferation, reminiscent of round blue cell tumors.[6] Aggressive behavior (5%–10%) occurs exclusively in the cellular variant,[7] potentially with metastases to the lung, liver, bone, or brain. Treatment is complete (or radical) nephrectomy, with chemotherapy reserved for partial nephrectomy.

The imaging features of CMN depend on the subtype. Classic CMNs appear as solid, homogenous soft tissue masses, frequently involving the renal sinus.[8] Ultrasound (US) may demonstrate alternating hyperechoic and hypoechoic rings as a consequence of vascular entrapment,[9] whereas prominent anechoic fluid may be seen in the cellular subtype (**Fig. 1**A). Computed tomography (CT) frequently shows an epicenter in the renal hilum (see **Fig. 1**B). On MR imaging, enhancement is typically peripheral,[10] with diffusion restriction reflecting increased cellularity. Alternatively, the cellular variant may be marked by more central and punctate enhancement, with areas of cystic change and hemorrhage. Locally aggressive features, such as vessel encasement and organ invasion, may be observed.[5] Differential considerations include Wilms tumor, clear cell sarcoma, and rhabdoid tumor, which are discussed in the later sections of this article.

Most recently, cytogenetic analysis has led to a deeper understanding of CMN. It has been shown that the chromosomal abnormalities of the cellular subtype (ETV6-NTRK3 gene fusion) are identical to those in infantile fibrosarcoma, whereas the more benign classic subtype is thought to represent a variant of infantile fibromatosis.[6]

Ossifying renal tumors of infancy

Ossifying renal tumors of infancy (ORTI) are very rare benign neoplasms of infancy and early childhood, with only 17 cases reported in the literature.[11] The tumor presents between 6 days and 30 months of age, mostly in boys (75%),[12] with macroscopic hematuria. Thus far, only one case has presented as a palpable mass.[13]

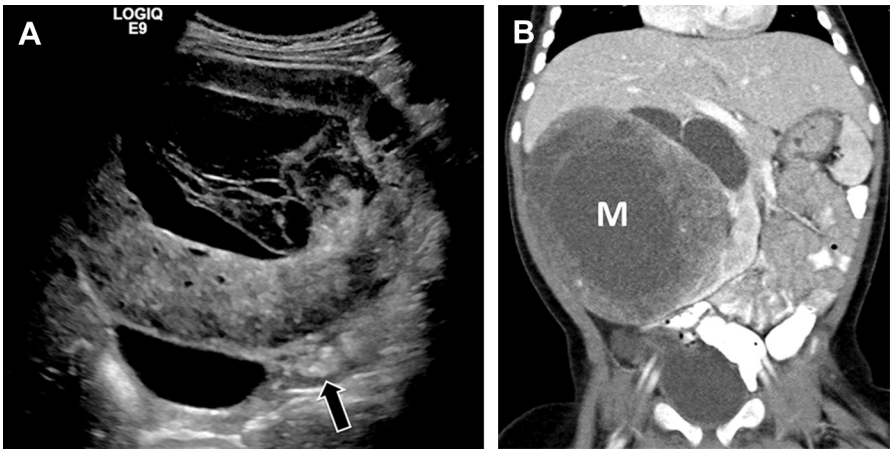

Fig. 1. A 6-month-old boy with mesoblastic nephroma, presenting as a palpable abdominal mass. (*A*) Longitudinal US and (*B*) coronal enhanced soft tissue window CT images show a complex solid and cystic mass (M) originating from the right kidney. The US image additionally shows medullary nephrocalcinosis (*arrow*).