Pediatric Case Reports

A Case of Bilateral Cystic Partially Differentiated Nephroblastoma vs Cystic Wilms' Tumor: Highlighting a Diagnostic Dilemma

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Cystic partially differentiated nephroblastoma (CPDN) is a rare multicystic renal tumor along the spectrum of cystic nephroma and cystic Wilms' tumor. There have only been two previously reported cases of bilateral CPDN in the literature. We present here a case of bilateral CPDN vs cystic Wilms' tumor treated with neoadjuvant and adjuvant chemotherapy in addition to a bilateral partial nephrectomy. We also review the relevant literature regarding CPDN in an effort to aid in diagnosis and management of these rare cystic renal tumors. UROLOGY 92: 106–109, 2016. © 2016 Elsevier Inc.

Multicystic renal tumors (MCRTs) are a spectrum of infantile tumors including cystic nephroma (CN), cystic partially differentiated nephroblastoma (CPDN), and cystic Wilms' tumor (WT). CN is characterized by multiple septations composed of differentiated tissue without blastemal elements. CPDN is composed of predominantly cystic lesions with blastemal or embryonal elements in the septa. Rather than being uniformly cystic, cystic WT has a solid component and may be associated with necrosis and hemorrhage.[1]

Clinically, CPDN is considered to be a low-risk tumor, and the SIOP Working Classification of Renal Tumors of Childhood recommends that this be treated with surgical resection alone.[2] The pitfall of this approach is that the distinction between CPDN and cystic WT cannot be made reliably with imaging or biopsy alone. Additionally, CPDN is difficult to classify after neoadjuvant chemotherapy as such treatment could affect, involute, or induce maturation of the small solid component that would have otherwise lead to a diagnosis of traditional nephroblastoma. Such nuances make the management of CPDN challenging. Here we present a rare case of bilateral CPDN vs cystic WT and review the literature regarding MCRTs.

CASE PRESENTATION

An 18-month-old male with a family history significant for pancreatic and central nervous system cancer presented to his pediatrician, who noted bilateral abdominal masses on physical examination. Upon referral to Texas Children's Hospital, the child had an elevated blood pressure of 126/76 and an elevated renin activity of 12.2 ng/mL/h. Computed tomography revealed bilateral renal masses concerning for cystic renal tumors (Fig. 1).

Neoadjuvant chemotherapy without biopsy was initiated to potentially reduce the size of the masses prior to resection. The patient was placed on the Children's Oncology Group (COG) research protocol AREN0534 and underwent 6 weeks of vincristine, dactinomycin, and doxorubicin. Follow-up imaging showed a small increase in the size of the masses bilaterally (Fig. 1). Due to the initial poor response to chemotherapy, additional neoadjuvant therapy did not seem like the best clinical option, and the patient underwent bilateral partial nephrectomies.

Bilateral double J stents were first placed and retrograde pyelograms demonstrated right-sided compression from tumor mass effect (Fig. 2). The right kidney was approached and intraoperative ultrasound was used to delineate the surgical margins before and during the resection. The tumor margin was scored using the Bovie cautery. Manual pressure at the base of the kidney was held and the tumor was then excised by enucleation. Collecting system defects were closed, the parenchyma was reapproximated using running 2-0 vicryl, and thombin sealant was applied over the renorrhaphy. The left kidney tumor resection was approached in a similar manner. Postoperative computed tomography showed no residual or metastatic foci. The patient had an unremarkable hospital course and was discharged 5 days later.

Grossly, the right renal mass was a smooth $10.5 \times 10.0 \times 9.7$ cm, 465.2 g multicystic mass with no tumor excrescences. The left multicystic renal mass measured $8.5 \times 8.0 \times 6.2$ cm and weighed 180.6 g. Both masses

Financial Disclosures: *The authors declare that they have no relevant financial interests.*

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Submitted: November 24, 2015, accepted (with revisions): January 19, 2016

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http://dx.doi.org/10.1016/j.urology.2016.01.018
0090-4295

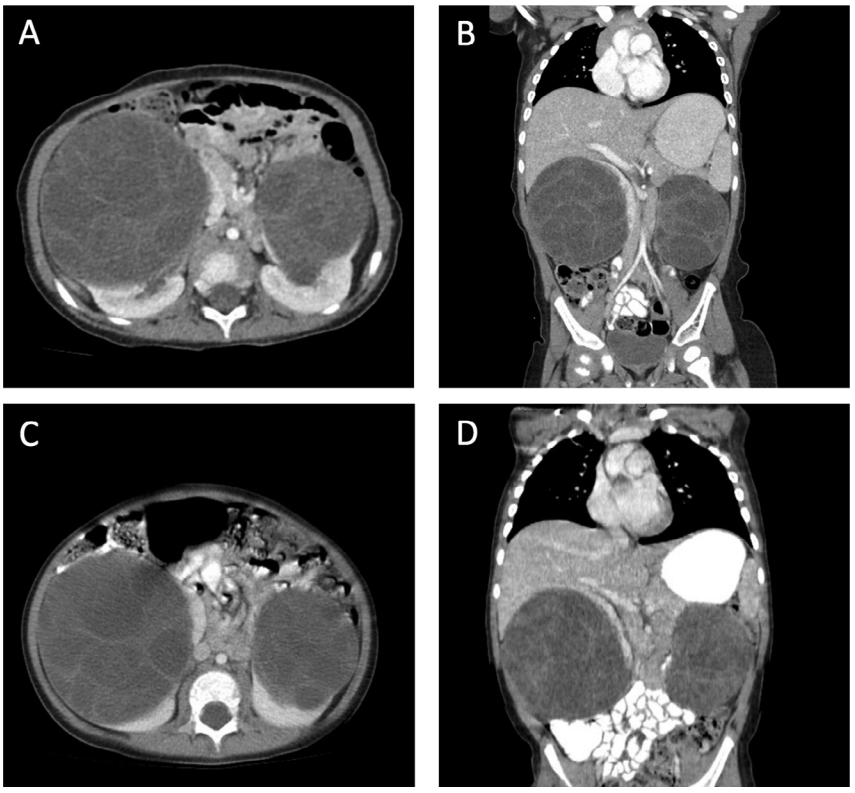

Figure 1. Prechemotherapy **(A)** axial and **(B)** coronal abdominopelvic computed tomography with contrast demonstrates bilateral, well-encapsulated renal neoplasms with multilocular and cystic components and splaying of the renal parenchyma. The right mass measures 8.3 cm × 9.2 cm × 9.3 cm and the left mass measures 6.2 cm × 8.0 cm × 6.3 cm. Postchemotherapy **(C)** axial and **(D)** coronal abdominopelvic computed tomography shows interval growth of the renal masses bilaterally. The right mass measures 9.1 cm × 9.6 cm × 9.6 cm and the left mass measures 6.1 cm × 7.9 cm × 7.9 cm.

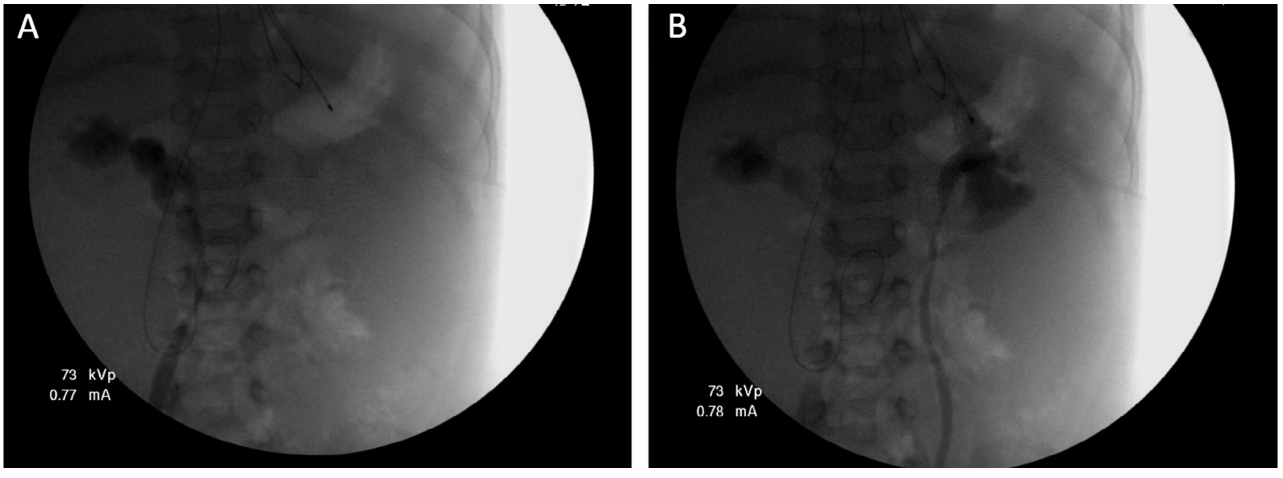

Figure 2. Retrograde intravenous pyelograms demonstrate compression of the **(A)** right renal calyces and pelvis and associated dilation of the proximal ureter. **(B)** Normal anatomy of the left collecting system is better preserved.

demonstrated numerous multiloculated cysts with intervening solid tissue (Fig. 3A). Sections of both masses were reviewed by Texas Children's Hospital pathologists and COG renal pathologists. The tissue sections demonstrated multiloculated cysts lined by low columnar epithelial cells (Fig. 3B). Within the solid tissue were occasional foci of blastemal cells and immature stroma and tubules (Fig. 3B,C). These elements immunoreacted with an antibody to WT1 (Fig. 3D). The majority of the solid tissue was composed of mature stromal cells and tubules. It is likely that the preoperative chemotherapy induced maturation of the nephroblastomatous components. The patient was

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