Cystic partially differentiated nephroblastoma: A rare differentiated variant of Wilm's tumour

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4-year-old male presented with abdominal lump noted since last 1 month. There was absence of history of jaundice and urinary or bowel complaints. On physical examination, there was a large ballotable intra-abdominal lump measuring $15 \times 10$ cm, involving right hypochondriac, lumbar, and umbilical region. Serum biochemical tests, including renal function tests, were normal.

Ultrasound study of the abdomen revealed a large multiloculated cystic lesion involving most of the right kidney. Cysts were separated by thin septae and no solid area was noted. Hence, a clinicoradiologic diagnosis of multicystic nephroma (MCN) was made and right nephrectomy was performed.

We received a specimen of right kidney with attached perinephric fat measuring $17 \times 10 \times 5$ cm. On cut section, the kidney was replaced by multiple cysts measuring 0.5–2.5 cm in diameter. The cysts were filled with pale yellow serous fluid and possessed smooth walls with thin septae 0.2 cm thick. No proliferating solid nodular or papillary area could be identified. Normal kidney tissue was seen compressed at one pole (Figure 1).

Multiple sections were examined and revealed numerous cysts lined by flat and cuboidal to hobnail epithelium. The cyst walls were composed of delicate fibrous tissue containing foci of blastemal element. Focal areas of abortive tubules were also seen (Figure 2). No features of anaplasia or heterologous stromal differentiation could be seen.

Based on the gross and microscopic findings, a final diagnosis of cystic partially differentiated nephroblastoma (CPDN) was rendered. The child is well after 4 months of follow-up without additional chemotherapy.

CPDN has been described as a rare differentiated variant of
Wilm’s tumor (WT).\(^1\) CPDN usually presents as abdominal mass in the pediatric age group, mostly before 2 years of age and is more common in males.\(^1\) Radiology (CT scan and MRI) is helpful in detecting the cystic character of the lesion. Absence of any solid area favors the diagnosis of CPDN or cystic nephroma over other pediatric cystic kidney lesions. However, detailed and thorough histopathological examination remains the mainstay of diagnosis.\(^2\)

Grossly, CPDN is usually unilateral, multiloculated, and circumscribed lesion. It is composed of multiple cysts of variable size with thin intervening septae without any solid nodular area.\(^2\) Microscopically, the cysts are lined by a single layer of flattened, hobnail, or columnar epithelium. The septae are composed of mesenchymal cells along with focal aggregates of blastemal cells, unlike cystic nephroma. Occasional tubular and glomerular structures in various stages of development may be seen. However, there is absence of anaplasia and mitosis is rare.\(^2\)

The recent discovery of papillonodular type of CPDN has led to modification of its diagnostic criteria, which are:\(^3\)

1. A discrete entirely cystic tumor containing luminal papillonodules in some cases.
2. Septa and the papillonodules, when present, are the only solid portions of the tumor and contain blastemal cells admixed with their normal and aberrant derivatives.
3. The tumor without and with papillonodules is classified as a conventional and papillonodular type of CPDN, respectively.

CPDN is recognized as a tumor with low but definite malignant potential; it has a favorable outcome (100% survival) as compared with classical WT. CPDN can be cured by nephron-sparing surgery or partial nephrectomy. However, it has a potential for local recurrence. Hence, noninvasive postoperative follow-up is recommended to document recurrences.\(^1\)

The clinicoradiological differential diagnoses include other pediatric cystic renal lesions, such as cystic mesoblastic nephroma, cystic renal dysplasia, cystic nephroma, and WT with cystic change. These can be differentiated from CPDN microscopically, and hence pathologic examination of the resected specimen is mandatory.\(^2\) Mixed epithelial and stromal tumor of kidney, a recently described cystic lesion, composed of spindle cells resembling ovarian stroma also comes in differential diagnosis. The presence of cellular stroma and absence of blastemal component differentiates it from CPDN.\(^4\)

CPDN, a rare differentiated variant of WT with potential for local recurrence, comes in differential diagnosis of pediatric cystic renal lesions. It needs to be differentiated from benign lesions such as cystic nephroma and mixed epithelial and stromal tumor of kidney.

References