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Cystic Partially Differentiated Nephroblastoma (CPDN): A Rare Variant of Wilms' Tumor

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Abstract **Cystic partially differentiated nephroblastoma (CPDN) is a rare variant of Wilms' tumor (nephroblastoma). It is an unusual neoplasm with a well-differentiated cystic lesion of the kidney. We herein report a case of a 4-month old boy presented with abdominal distension and left flank mass. Abdominal radiograms showed soft tissue haziness in the left side of the abdomen with no calcification. Abdominal ultrasonograms revealed a large multiseptated cystic mass involving most of the left kidney. CT scan of the abdomen showed a 6 × 7 cm hypodensed mass with well-defined septa of the left kidney. There was no extension of the tumor to the renal capsule and the lymph nodes showed reactive changes. Left nephrectomy was performed. The pathological diagnosis was cystic partially differentiated nephroblastoma (CPDN). Post-operative course was uneventful and the patient has been doing well since.**

Introduction

Wilms' tumor is the most frequent tumor of the kidney in infants and children. It is characterized by a solid mass composed of triphasic embryonal neoplasm. In the entity of Wilms' tumor, if the lesion is composed of purely cystic masses and characterized by multiple thin-wall separation, the differential diagnosis includes cystic nephroma (CN), cystic partially differentiated nephroblastoma (CPDN) and cystic nephroblastoma (cystic Wilms' tumor). They are distinguishable by pathohistological characteristic.

We have one-case experience of a rare variant of Wilms' tumor. We herein report this case for further study.

Case report

A 4-month-old boy presented with abdominal distension due to a left flank mass noticed by his parents 1 week earlier. There was no history of bowel or urinary problems. On physical examination, there was intra-abdominal mass measuring about 10 cm. in diameter occupying left hypochondriac, lumbar and umbilical regions. The mass was hard consistency, smooth surface and positive bimanual palpation. Serum biochemical tests, urinary analysis, renal and liver function tests were within normal limits.

Abdominal radiograms showed soft tissue haziness in the left side of the abdomen with no calcification. Bowel gas was being displaced to the right (Figure1).

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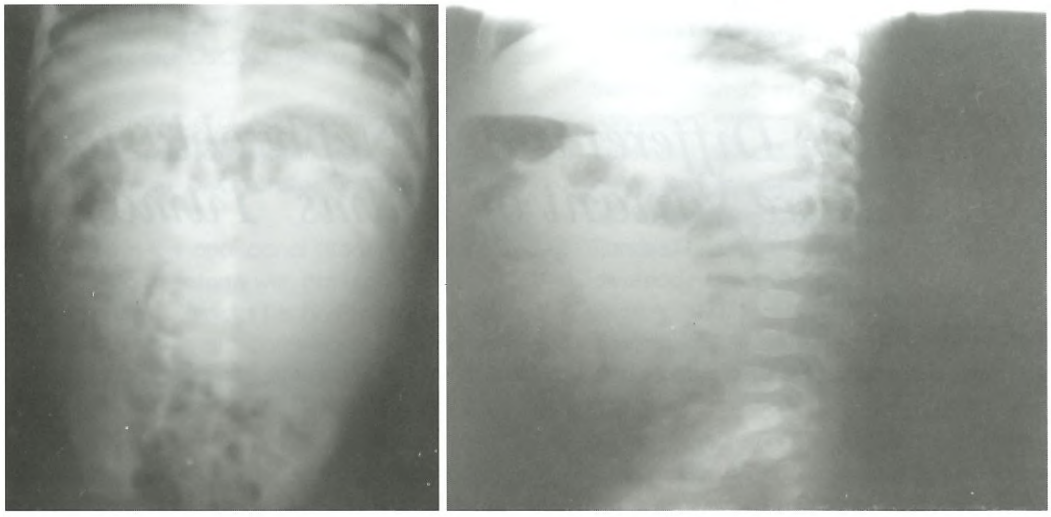

Figure 1 Abdominal radiograms showed soft tissue haziness in the left side of abdomen, no calcification, and bowel gas being displaced to the right

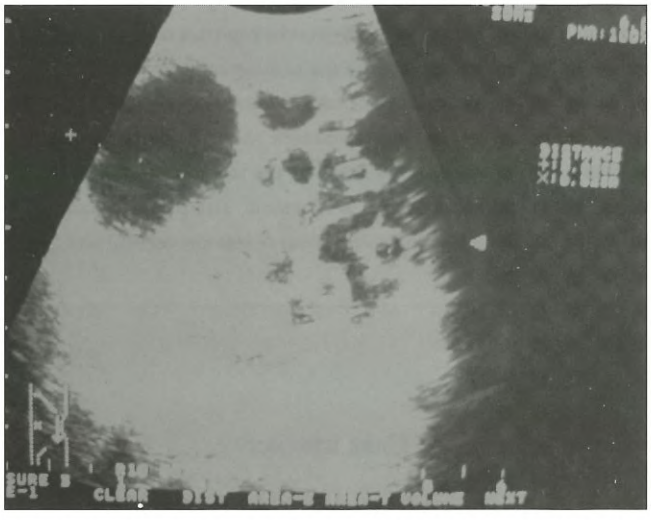

Figure 2 Abdominal ultrasonogram showed a large multiseptated cystic mass involving most of the left kidney.

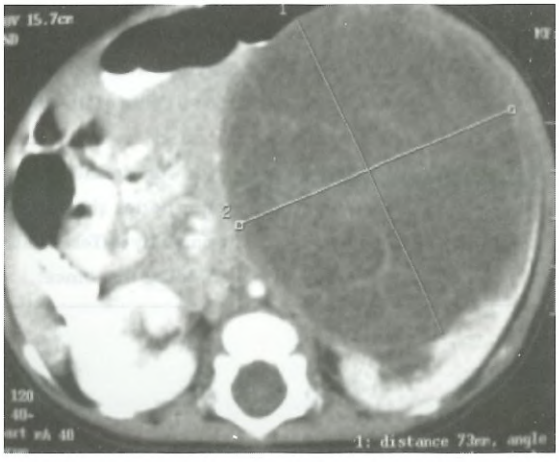

Figure 3 CT scan of the abdomen showed a 6 × 7 cm hypodense mass with well-defined septa of the left kidney.

Chest film was normal.

Abdominal ultrasonograms revealed a large multiseptated cystic mass involving most of the left kidney (Figure 2). CT scan of the abdomen showed a 6×7 cm hypodense mass with well-defined septa of the left kidney (Figure 3). The pre-operative diagnosis was left renal tumor, most likely Wilms' tumor.

At laparotomy, the tumor was seen to be at the lower pole of the left kidney (Figure 4). The ureter and the renal calyceal system were not dilated. There was no thrombus in the renal vein. The liver and the right kidney were normal. Left nephrectomy was performed.

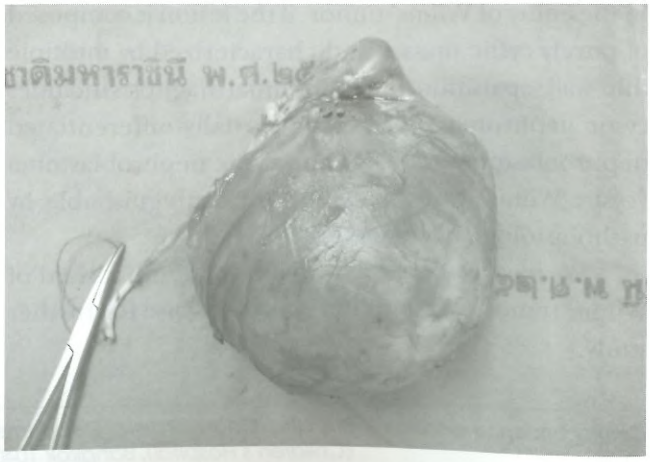

Figure 4 The cystic tumor measuring 9.5×7.5×7.3 cm at the lower pole of the left kidney

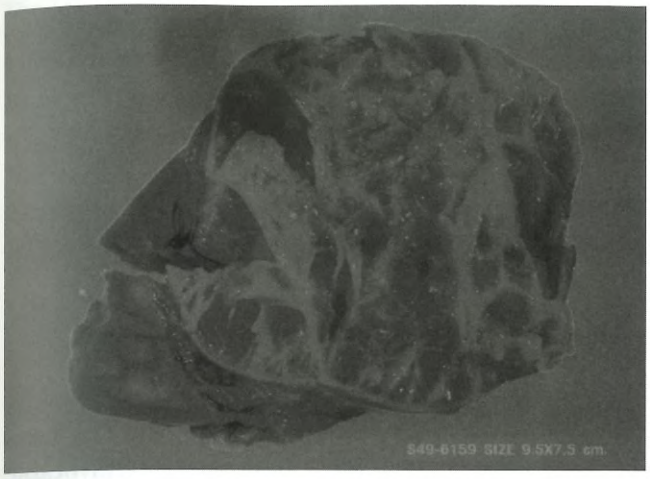

Figure 5 The cut surface showed no solid areas

The specimen was a cystic tumor measuring 9.5 × 7.5 × 7.3 cm, the cut surface showed no solid areas (Figure 5).

Microscopically, the tumor was composed of multiple cysts lined by cuboidal, flattened and hobnail cells (Figure 6). The septa were composed of predominantly fibrous tissue and molded the cystic spaces (Figure 7).

Small foci of blastema and abortive tubules that lacked the proliferative activity present in the septa were noted (Figure 8, 9).

There were no extension of tumor to the renal capsule and the lymph nodes showed reactive changes. The pathological diagnosis was cystic partially

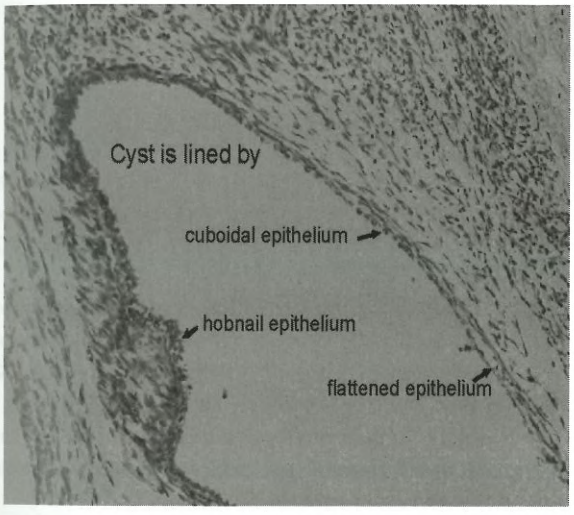

Figure 6 The cyst lined by cuboidal, flattened and hobnail cells.

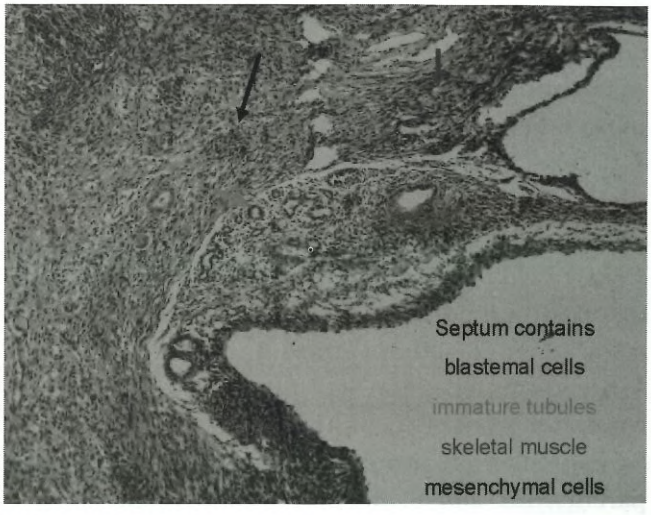

Figure 8 Small foci of blastema and abortive tubules lacking the proliferative activity present in the septa.

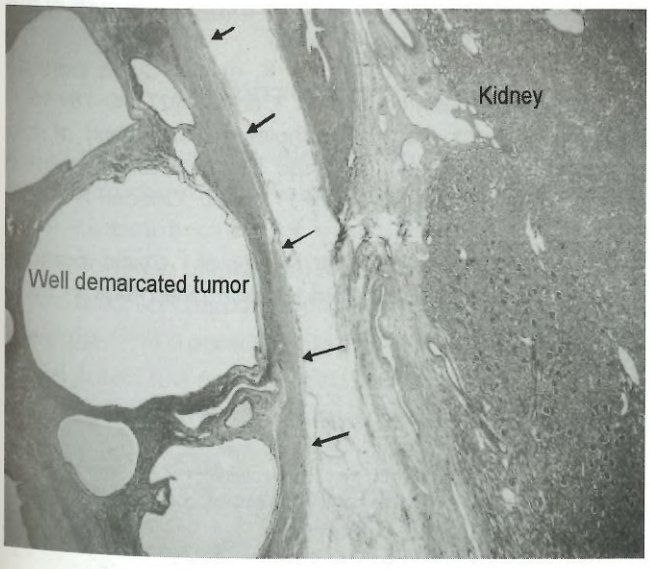

Figure 7 The well-circumscribed cystic tumor

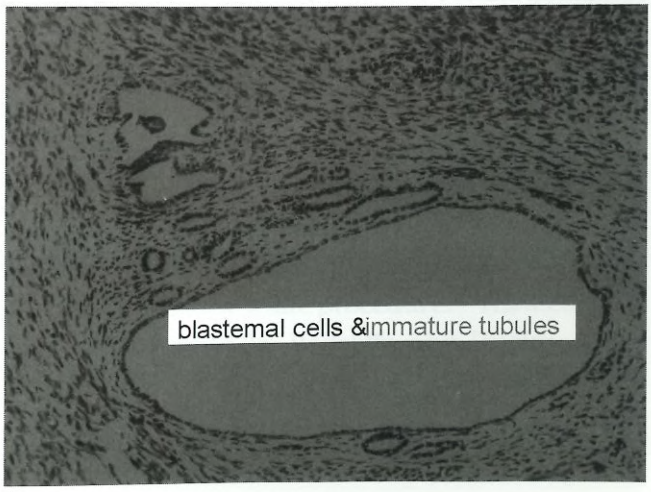

Figure 9 The blastemal cells and immature tubules

differentiated nephroblastoma (CPDN). Post-operative course was uneventful. The oncologists suggested no further chemotherapy. The patient has been doing well since.

Discussion

CPDN is a favorable cystic variant of Wilms' tumor (nephroblastoma) with unique pathological characteristics. It makes up less than 1% of all Wilms' tumor patients (NWTSG-3, -4, and -5).[1] There are two peak-age distributions; at 4 months to 2 years and at middle age.[2] The tumors are more common in boys than girls (Male:Female = 2:1).[1,3] The clinical presentations include asymptomatic mass with pain or hematuria occasionally. Radiological studies show cystic lesion. Histopathologically, it is a mass surrounded by fibrous pseudocapsule 5 to 10 cm. in size without hemorrhage, necrosis or calcification.

Several overlapping terms have been used to describe this group of tumors such as cystic Wilms' tumor, cystic nephroma (CN) and multilocular cyst (MLC).[2,4] In 1951, Powell et al.[5] established 8 diagnostic criteria of multilocular cyst which included 1. unilateral involvement 2. solitary lesion 3. multilocular lesion 4. non-communication with the renal pelvis 5. non-communication of cysts with each other 6. loculi lined by epithelium 7. intralocular septa devoid of renal parenchyma and 8. normal residual renal tissue if present.

In 1989 Joshi et al.[2,6,7] described diagnostic criteria for CPDN and recommended using the term cystic nephroma (CN) instead of MLC when the septa contained mature tubular structures. The presence of blastemal cells or poorly differentiated stromal or epithelial elements should exclude the diagnosis of MLC or CN.

The diagnostic criteria by Joshi et al.[2,6] for CPDN include: the tumor composed entirely of their septa; discrete well demarcated mass; septa being sole solid component and conform to outlines of cysts without expansile nodules; cysts being lined by flattened, cuboidal or hobnail epithelium; and septa containing blastema and/or embryonal stroma or epithelium element.

CPDN is curable by nephrectomy alone.[4,8] However, incompletely excised or ruptured tumor can recur. No case of metastasis has been reported. Regular follow-up is recommended.

Conclusion

CPDN is a rare variant of Wilms' tumor but a curable malignant neoplasm. It is at the hyperfavorable end of Wilms' tumor spectrum and curable by simple nephrectomy.

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