Case Report

Cystic Partially Differentiated Nephroblastoma : A Rare Case Report

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ABSTRACT

Cystic partially differentiated nephroblastoma (CPDN) is a rare tumour of infancy which is considered to be a low risk malignant neoplasm. It has a favourable outcome as compared to classical Wilms tumor.

A 10 month old male child presented with abdominal lump of two months duration. Ultrasound abdomen revealed a cystic mass involving most of the left kidney. After nephrectomy we received a specimen of kidney measuring 7x6x5 cms. Multiple cystic areas were seen along with areas of hemorrhage and a narrow rim of normal kidney. On microscopy, multiple cysts of varying sizes were seen lined by cuboidal, flattened, eosinophilic and hobnail cells. The septa were relatively thick containing bland spindle cells with few foci of blastemal cells .A diagnosis of cystic partially differentiated nephroblastoma was made.

CPDN is a rare variant of Wilms tumor with a favorable prognosis. Histopathologic examination helps to differentiate it from other cystic lesions of the kidney and is of therapeutic importance.

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Introduction

Cystic partially differentiated nephroblastoma (CPDN) is a rare tumour of infancy which is considered to be a low risk malignant neoplasm. It has a favourable outcome as compared to classical Wilms tumor. Clinicoradiological differential diagnosis include cystic mesoblastic nephroma, cystic renal dysplasia, cystic nephroma (CN) and Wilms tumor with cystic change. Differentiation from these entities is important due to the therapeutic implications and requires histopathological examination.

Case Report

A 10 month old male child presented with abdominal lump of two months duration. There was no history of jaundice, urinary or bowel abnormalities. Serum biochemical tests were normal. Ultrasound abdomen revealed a cystic mass involving most of the left kidney. A nephrectomy was performed and we received a specimen of kidney measuring 7x6x5 cms on gross examination. Multiple cystic areas were seen along with areas of hemorrhage and a narrow rim of normal kidney. The ureter appeared normal. No lymph nodes were received. On microscopic examination, multiple cysts of varying sizes were seen lined by cuboidal, flattened, eosinophilic and hobnail cells (fig 1,2). The septa were relatively thick containing bland spindle cells. Foci of blastemal cells were seen at places in the septa along with abortive tubule formation (Fig 3). Based on the above findings, a diagnosis of cystic partially differentiated nephroblastoma was made.

Discussion

Cystic renal tumors of infancy are uncommon. Together, they form a spectrum with CN at the benign end, CPDN in the intermediate region and Wilm's tumor with multicystic areas at the malignant end [1,2,3]. 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 [4].There are two peak-age distributions; at 4 months to 2 years and at middle age [5].The tumors are more common in boys than girls (Male:Female = 2:1) [4,6].

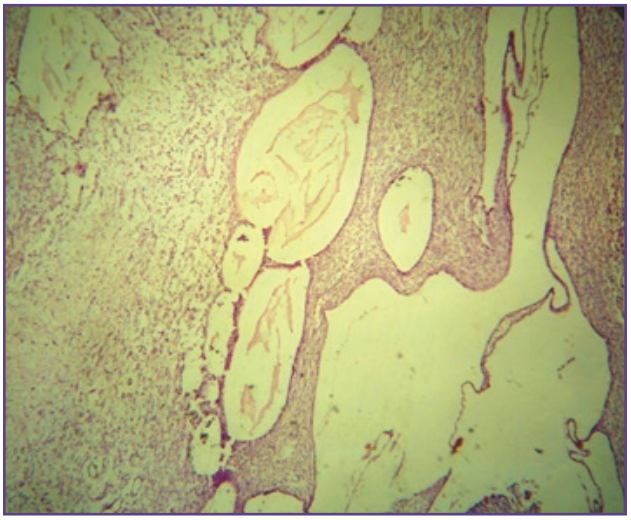

Fig. 1: Numerous cysts lined by hobnail shaped eosinophilic cells; the septa contain cellular fibrous tissue with bland spindle cells (H&E, 40x)

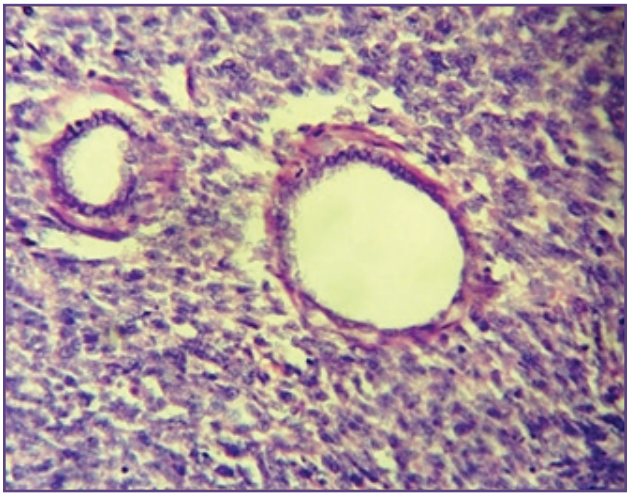

Fig. 2: Cysts lines by hobnail cells (H & E, 100X)

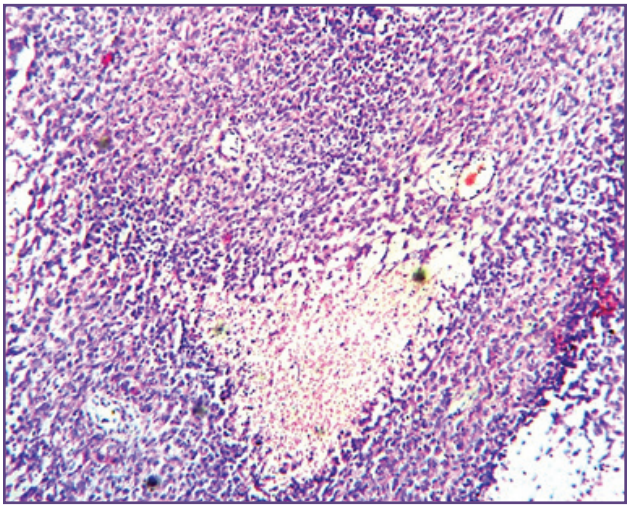

Fig. 3: Section showing foci of blastemal cells (H & E, 40X)

Clinically, patients usually present with non specific symptoms such as a painless abdominal mass [7]. Progressive enlargement, which may be sudden, is reported [7,8]. Hematuria can be seen and is thought to be due to the extension of the tumor into the pelvicalyceal system.

Grossly, tumor is well circumscribed from the remaining kidney by a fibrous pseudocapsule and consists entirely of cysts of variable size. The septa are thin and there are no

expansile nodules to alter the rounded contour of the cysts. The cysts in cystic partially differentiated nephroblastoma are lined with flattened, cuboidal, or hobnail epithelium, or lack lining epithelium [8]. The septa are variably cellular and contain undifferentiated and differentiated mesenchyme, blastema, and nephroblastomatous epithelial elements [8]. Skeletal muscle and myxoid mesenchyme are present in the septa of most tumours. Cartilage and fat are present occasionally .[5,9]

In 1977, Joshi and Banerjee [8] distinguished CPDN from CN depending on the presence and absence of blastemal element within the septa, respectively. Other authors propose that CN is a CPDN in which the blastemal elements maturated or that Wilm's tumor with multicystic areas is simply a precursor for CPDN. [5]

The presence of blastemal cells or poorly differentiated stromal or epithelial elements should exclude the diagnosis of Multilocular cyst or CN. On gross examination, Wilm's tumor with multicystic areas contains expansile nodules. In 1989 Joshi et al.[5,8,9] described diagnostic criteria for CPDN and recommended using the term CN instead of multilocular cyst (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.[5,8] 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.

It is important to distinguish CPDN from the other cystic renal tumors especially Wilm's tumor with multicystic areas since the latter should receive adjuvant chemotherapy. On the other hand, CPDN is curable by nephrectomy alone. [10] [11] However, a regular follow up is recommended to document any recurrence.

Conclusion

CPDN is a rare variant of Wilms tumor with a favorable prognosis. Histopathologic examination helps to differentiate it from other cystic lesions of the kidney and is of therapeutic importance.

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Competing Interests

None declared

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