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Case Report

Cystic Partially Differentiated Nephroblastoma- A Rare Form of Wilm's

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Abstract

Wilm's tumor is the commonest malignant renal tumor in infants and children. Congenital Cystic Partially Differentiated Nephroblastoma (CPDN) is a rare form of Wilm's tumor. It presents with painless abdominal distention and flank mass. It is considered of low malignancy potential. Complete surgical resection is usually curative. We report a 10 month old boy with a right flank mass. CT scan showed a right well capsulated hypodense cystic mass occupying the whole kidney with septations. Right nephrectomy was done. The pathology revealed (CPDN). Post-operative course was uneventful. The infant is doing well.

Introduction

Wilm's tumor is the commonest renal tumor in children. It includes wide variety of tumors that ranged from benign to malignant and from solid to cystic. The cystic lesions ranged from the Cystic Nephroma (CN) to CPDN. Cystic nephroma is a benign tumor. It consists of cystic lesion with septation and no solid component. The septa contains well differentiated cells with no blastemal or embryonal cells. CPDN which has been designated by various terms like polycystic nephroblastoma, cystic Wilm's tumor, cystic nephroma and differentiated nephroblastoma [1]. These fore mentioned terms should not be used to avoid confusion. Actually the term CPDN has been widely accepted and used. CPDN consists of cystic mass that occupy the whole kidney with variable septations that contains admixture of blastemal cells with variably differentiated epithelial and stromal elements [2]. Subdivision of CPDN into well-differentiated and poorly differentiated subtypes has been suggested. However some authors objected to the inclusion of this subdivision to the CPDN. This tumor is curable by nephrectomy alone. However intraoperative rupture of the tumor or in completed resection leads to recurrence. Regular follow up by noninvasive technique would be advised.

Case Report

11 month old male infant presented with abdominal distention noticed by his mother. He was healthy otherwise, a product of full term vaginal delivery with uneventful neonatal period. He had no history of fever, anorexia, hematuria, changes in bowel or bladder habits. His blood workup was

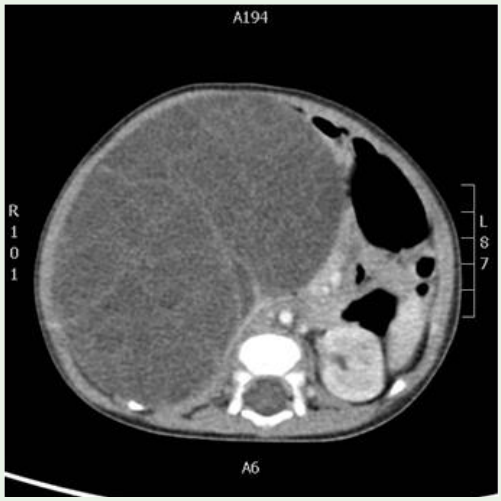

Figure 1: CT scan of the abdomen showing right cystic renal mass completely replacing the kidney with septations.

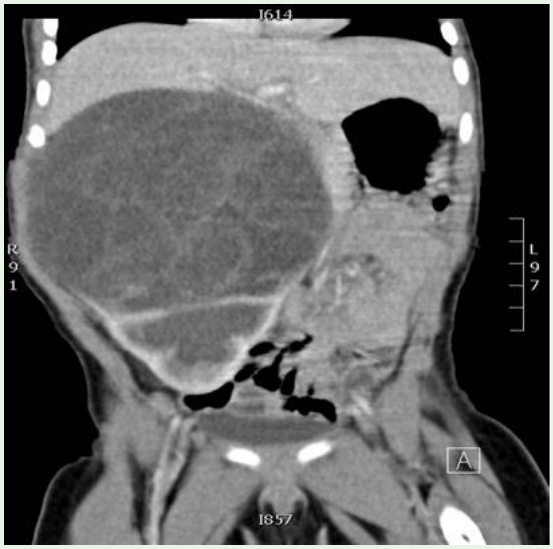

Figure 2: Coronal view of the same patient.

unremarkable. Clinically he looked well with unremarkable systemic examination. The abdomen was distended with ill defined right flank mass occupying the whole right flank. CT scan of his chest abdomen and pelvis showed large cystic mass replacing the whole right kidney with multiple septations. It measured 10 by 13 cm (Figures 1 and 2) no lung metastasis.

Laparotomy and right nehrectomy was done. Intraoperatively the tumor was very well capsulated easy to dissect from surrounding tissues. Hilar, para-aortic and pracaval lymph nodes were sampled for pathology. The surgery went uneventful. Pathology showed typical picture of CPDN. The tumor has been completely excised, no invasion to the renal capsule. Hilar, Para-aortic and paracaval LN were all reactive and free of malignancy.

Post-operative period was smooth and the infant is doing well up to date. His ultrasonography of the abdomen showed no evidence of recurrence.

Discussion

Cystic renal tumors of infancy are uncommon. Together, they form a spectrum with CN at the benign, CPDN in the intermediate region and cystic Wilm's tumor at the malignant end [3]. Numerous terms have been used to describe these cystic renal tumors: benign multilocular cystic nephroma, polycystic well-differentiated Wilm's tumor, differentiated nephroblastoma and CPDN. This had led to confusion and ambiguity. The origin of cystic renal tumors has been debated in the past [4]. Firstly, they have been considered as hamartomatous lesions. Nowadays, these lesions are considered to be of neoplastic nature [6].

In 1977, Joshi and Banerjee distinguished CPDN from CN depending on the presence and absence of blastemal element within the septa respectively [7,8].

CPDN is a multilocular cystic neoplasm of very young children composed of epithelial and stromal elements, along with nephroblastomatous tissssue [9] it is a low risk malignant tumor.

Clinically, patients usually present with painless abdominal mass. Hematuria is uncommon.

Ultrasonography reveals hypoechoic mass with hyperechoic septa. CT scan demonstrates hypodense cystic mass with septations. MR imaging is rarely indicated and adds no much advantages. These radiological findings are only suggestive and the final diagnosis will be based on Histology [9]. Joshi and Beckwith et al described diagnostic criteria for CPDN in 1989 that was refined by Eble and Bonsib in 1998 [2,5].

In summary, CPDN is considered as a low risk malignant tumor. It is treated by nephrectomy alone with excellent prognosis.

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