Case Report (I)

Cystic Partially Differentiated Nephroblastoma

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ABSTRACT:

We describe case records of two children with cystic partially differentiated nephroblastoma (CPDN). Both children underwent unilateral nephrectomy for renal mass. Their metastatic work up was negative. The cases are presented in view of its rarity. Pertinent literature is reviewed.

INTRODUCTION:

Cystic partially differentiated nephroblastoma (CPDN) is a rare primary malignant renal tumor of young children. Incidental detection of asymptomatic unilateral abdominal mass is the usual presentation. Radiological studies are usually non-diagnostic. Macroscopically, the tumor is well encapsulated with multiple cystic spaces separated by the septa. The unique characteristics present in the septa, which are variably cellular and contain differentiated and undifferentiated mesenchyme, blastema and nephroblastomatous epithelial elements. Microscopic study is essential to distinguish CPDN from cystic nephroma and other types of multicystic kidney disease. CPDN is curable by nephrectomy alone. Lesion can recur if ruptured or incompletely excised. In view of rare possibility of recurrence, regular monitoring by non-invasive technique would be advisable[1].

CASE-I :

A five-month-old female child presented with history of left nephrectomy for incidentally detected asymptomatic renal mass. Histopathologic finding revealed CPDN. Her past medical, surgical and family history was non-contributory. Physical examination was normal. Laboratory study was unremarkable. Preoperative intravenous pyelogram revealed cystic lesion in the left kidney with displacement of calyces. Pre-operative CT scan of abdomen (Figure-I) showed 9 x 8.5 cm, lobulated mass with well defined septae located on upper-middle pole of left kidney. Metastatic work up was negative. Surgical findings revealed tumor confined to left kidney with intact capsule and no metastatic foci (Stage-I).

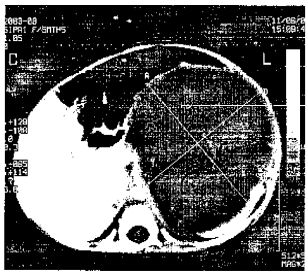

Figure-1: Computed Tomography of the Abdomen showing well-defined cystic lesion involving upper pole of left kidney

CASE II:

A two year old male child presented with history of left nephrectomy for renal mass detected during work up of pyrexia of unknown origin. Histopathologic findings of mass revealed CPDN. His past medical, surgical and family history was non-contributory. His physical examination was

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normal. Laboratory study was within normal limit. Preoperative intravenous pyelogram revealed a cystic lesion in the left kidney with displacement of calyces. Pre-operative CT scans of the abdomen showed 6 x 6.5 cm, lobulated mass with well defined septae located on middle pole of left kidney. Metastatic work up was negative. Peroperatively tumor was confined to left kidney with intact capsule and no metastatic foci.

PATHOLOGIC FINDINGS (Figure 2)

Gross examination revealed well encapsulated tumor. Cut surface of the tumor showed cystic spaces with thin rim of compressed renal parenchyma. Cysts were multiloculated, non-communicating filled with clear pale yellow fluid. Cyst wall was thin. Rest of the renal parenchyma was normal.

Microscopic examination of the tumor in both cases revealed multiple cysts. Cyst wall was lined by flattened, cuboidal epithelium with hobnail cells. The septae formed of fibrous tissue, inflammatory cells and reactive fibroblasts. The septa were variably cellular. The septa were composed of blastema, tubular and mesenchymal component. Blastemal cells have small pleomorphic and hyper chromatic nuclei with typical mitosis and interlacing pattern. Focally stroma was myxoid. Renal capsule, surgical margin, renal vessels and ureteric margin were free of tumor.

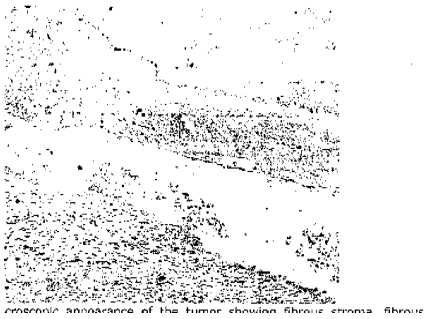

Figure-II: Microscopic appearance of the tumor showing fibrous stroma, fibrous tissue, cystic spaces separated by septae (HPE 10 x)

DISCUSSION

CPDN was first established as clinico-pathologic entity by Brown in 1975. In the past. Multilocular cystic renal tumor has been considered to be lesion of developmental origin[1]. More recently, Joshi and Benerjee[2] proposed a modification emphasising neoplastic rather than the developmental origin of the tumor. They suggested that the term used to denote predominantly cystic lesion without nodular solid region and in which septa contain blastemal or embryonic elements.

This lesion is rare as there are very few cases reported in the literature. Joshi & Benerjee[2] described three cases of CPDN and reviewed 10 cases reported in the literature. Gallo & Penchansky[3] found four such cases (2%) among 165 primary renal tumors in children. Joshi & Beckwith[4] reported 18 cases of CPDN while review of lesions with prominent cystic changes that had been entered into the National Wilm's Tumor Study. There are also isolated case reports available in the literature.

Review of Literature : Usual age of presentation is between 4 months to 24 months. Joshi & Beckwith[4] found only two patients older than 24 months and 65% were diagnosed in the first year of life. Very few cases of CPDN are also reported in the adult[5,6] . It is less frequent in boys than girls. (Male to female ratio 1:1.6) Asymptomatic abdominal mass is usual presentation[7]. Presentation with PUO has not been reported in literature.

Radiologic study in CPDN is usually not diagnostic. On excretory urography the kidney function normally with multilocular cystic intrarenal tumor. It may reveal stretching, displacement, extension of the tumor to the renal pelvis. Ultrasound reveals multiple anechoic spaces separated by hyper echoic septae. Renal origin of mass can be confirmed by identifying normal renal parenchyma around the periphery of well defined mass. CT scan

demonstrates intrarenal mass with well defined margin, multicystic architecture, enhancing septae and herniation into renal collecting system[8].

Histopathologically, the cysts are lined by flattened, cuboidal or hobnail epithelium. The septa of cysts show a mixture of partially differentiated and undifferentiated blastemal or other embryonic tumor. CPDN is prominently cystic lesions lacking nodular soild regions, in which blastemal or other embryonic cells are present in the septa of the cysts[9,10]. Joshi and Beckwith et. al, described diagnostic criteria for CPDN, which was refined by Eble and Bonsib (Table 1).

Table-1 Diagnostic Criteria for CPDN

Joshi and Beckwith (1989)[1,4]
1. Tumor composed entirely of cysts and their septa
2. Discrete well demarcated mass.
3. Septa are sole solid component and conform to outlines of cysts without expansile nodules.
4. Cysts are lined by flattened, cuboidal or hobnail epithelium.
5. Septa contain blastema ± -embroynal stroma or epithelium elements.
Eble and Bonsib (1998)[1]
1. Pediatric patient, exceptional above age 2 years.
2. Expansile mass surrounded by fibrous pseudo capsule. Interior entirely composed of cysts and septa with no expansile solid nodules.
3. Septa may contain flattened, cuboidal or hobnail epithelium
4. Septa may contain epithelial structures resembling mature renal tubule.
5. Septa contain blastema ±- embroynal stroma or epithelial elements.

Common differential diagnosis of CPDN is from cystic nephroma, which is well differentiated and lacks immature blastemal/embryonic cells. Cystic Wilm's tumor has cysts presents as a minor feature accompanying the triphasic stroma. Polycystic nephroblastoma may have extensive cysts but solid nodules of tumors are present. Other renal neoplasm may contain cysts, usually as a minor component; these include renal cell carcinoma, mesoblastic nephroma and clear cell sarcoma[1].

CPDN is curable by nephrectomy alone. However, incompletely excised or ruptured tumor can recur. Joshi and Benerjee reviewed 13 cases; in seven cases simple nephrectomy and in remaining cases nephrectomy with radiation and/or chemotherapy was done. The disease free interval range from 5-72 months without reports of recurrence or metastasis. Blackely - et. al[11], reported that outcome of patients with CPDN is favorable with 100% survival rate and no recurrences for stage 1 tumor treated with complete surgical resection. Patients treated with partial nephrectomy have been reported with successful outcome[12].

CPDN appear to take benign natural course, the lesion may occasionally follow aggressive course; in view of remote possibility of recurrence regular monitoring by non-invasive technique would be advisable[1,3,11].

CONCLUSION

CPDN is rare but curable malignant cystic renal neoplasm of young children; it represents the ''hyperfavourable'' end of Wilm's spectrum. Simple nephrectomy with regular follow up visits may be the management of choice especially in stage I tumor.

REFERENCES

1 *Eble JN and Bonsib SM. Extensively cystic renal neoplasm: Cystic nephroma, Cystic partially differentiated nephroblastoma, Multilocular cystic renal cell carcinoma and Cystic hamartoma of renal pelvis. Seminar in Diagnostic Pathology 1998; 15 (1): 2-20.*

2 *Joshi VV., Benerjee A.K. Yadav K., Pathak I.C. Cystic partially differentiated nephroblastoma-A clinico-pathological entity in the spectrum of infantile renal neoplasia. Cancer 1977; 40:789-795.*

3 Gallo GE, Penchansky L. Cystic Nephroma. Cancer 1977; 39:1322-27

4 Joshi VV., Beckwith JB. Multilocular cyst of the kidney. Cystic nephroma and Cystic partially differentiated nephroblastoma. Terminology and Diagnostic criteria. Cancer 1989; 64: 466-479.

5 Nagao T, Sugano I., Ishida Y et.al. Cystic partially differentiated nephroblastoma in adult: An immunohistochemical, lectin histochemical and ultra structural study. histopathology 1999; 35(1): 65-73 (Abstract)

6 Kumar R, Seth A, Dinda AK. Incidental, cystic partially differentiated nephroblastoma in an adult. J Urol. 2001 165(5) 1622.

7. Graundy PE. Green DM., Coppes MJ et al Renal tumors. In PA Pizzo and Poplock DG. (Editors): Principle and Practice of Pediatric Oncology. Pub. Lippincott William and Wilkins, Philadelphia 2002, 4th edition. 865-895.

8 Madwell JE, hartman DS, Liechtenstein JE Radiologic-Pathologic correlations in cystic disease of the kidney. Radio logic Clinics of North America 1979; 17:261-62.

9 Richard K. Sibley, Louis P. Dehner. Kidney. Pediatric surgical pathology: 2nd Edition. Edited by Louis P. Dehner. Published by Lippincott Williams and Wilkins. Philadelphia-2002.

10. Epstein L, Wackman J and Daughtry J, et al. Multilocular cysts of kidney: A diagnostic dilemma. Urology 1978; 11:573-76.

11. Blakely ML, Shampberger RC, Norkool P.et al. Outcome of children with Cystic partially differentiated nephroblastoma treated with or without chemotherapy. Pediatric Surg. 2003; 38(6): 897-900.

12. Ranjangam K, Narasimhan KL, Trehan A et. al. partial nephrectomy in cystic partially differentiated nephroblastoma. J. Pediatric Surg. 2000; 35(3):510-20 (Abstract)