

## CASE REPORT

# Cystic partially differentiated nephroblastoma: A report of two cases with review of the literature

SarraFOULI,<sup>1</sup>Faten HAMMEDI,<sup>1</sup> Rim HADHRI,<sup>1</sup> LeilaNJIM,<sup>1</sup>Manel NJIMA,<sup>1</sup>Imed KRICHENE,<sup>2</sup> Houda MAHMOUDI,<sup>1</sup>Adnène MOUSSA<sup>1</sup>And Abdelfateh ZAKHAMA<sup>1</sup>

<sup>1</sup>Department of pathology.FattoumaBourguiba Hospital, Monastir, Tunisia

<sup>2</sup>Department of Paediatric Surgery.FattoumaBourguiba Hospital, Monastir, Tunisia

### Abstract

Cystic partially differentiated nephroblastoma is a rare tumor of the kidney usually affecting infants. It should be distinguished from Wilms' tumor with multicystic areas and multilocular cystic nephroma. Multilocular cystic nephroma is a benign tumor whereas Wilms' tumor with multicystic areas is at the malignant end of the range of classification of such tumours. Cystic partially differentiated nephroblastoma may undergo local recurrence but there is no report of metastasis. In this paper we report the cases of two young boys aged fewer than three years presented with painless tumefaction on left side flank. The diagnosis of partially differentiated cystic nephroblastoma was retained on histological evidences.

**Key words:**Cystic partially differentiated nephroblastoma, kidney, prognosis.

### INTRODUCTION

Cystic renal tumors of childhood represent a range of lesions that include cystic nephroma (CN), Cystic partially differentiated nephroblastoma (CPDN), and Wilms' tumor with multicystic areas.<sup>1,2</sup>The distinction between these subtypes is important considering the therapeutic implications. Diagnosis has to be retained on the basis of histology, as radiological findings are usually inconclusive.<sup>2</sup>CPDN is a rare tumor of infancy and is considered as a low-risk malignant tumor. Herein, we report two male infants less than 3 years of age, who underwent nephrectomy in which we have elaborated the diagnosis of CPDN.

### CASE REPORT1

This is a one year-old male infant who presented with a large mass on the left side of his abdomen evolving over a period of several months. He had no history of fever, anorexia and weight loss, or symptoms of urinary tract infection. Laboratory findings were normal. CT tomography of the abdomen confirmed the presence of a multilocular cystic mass that

measured 20 cm across arising from the left kidney. Three retroperitoneal lymphadenopathies were detected.Needle guided aspiration cytology of the mass was non contributory.

The patient underwent left nephrectomy with retroperitoneal lymph node resection.

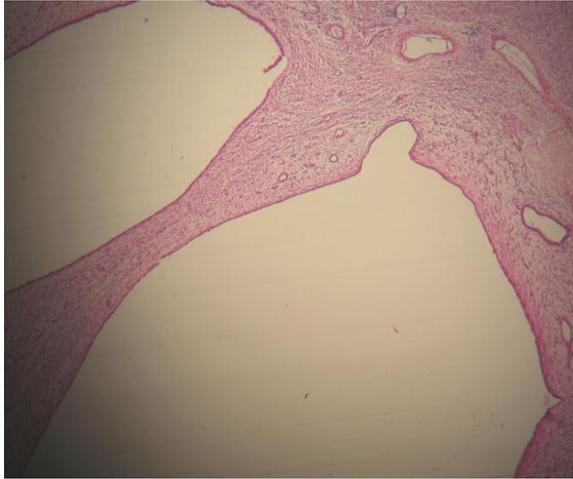
On gross examination, the resected kidney was unrecognizable. We received a lobulated mass that measured 23 x 16 x 16 cm. The cut surface revealed noncommunicatingloculi which contained fluid. The cyst walls were thin, without expansile nodules. This mass was surrounded by a thick fibrous capsule. It compressed laminated residual renal parenchyma (Fig. 1).



**Figure 1** Gross examination of a well-circumscribed mass composed entirely of small and large cysts.

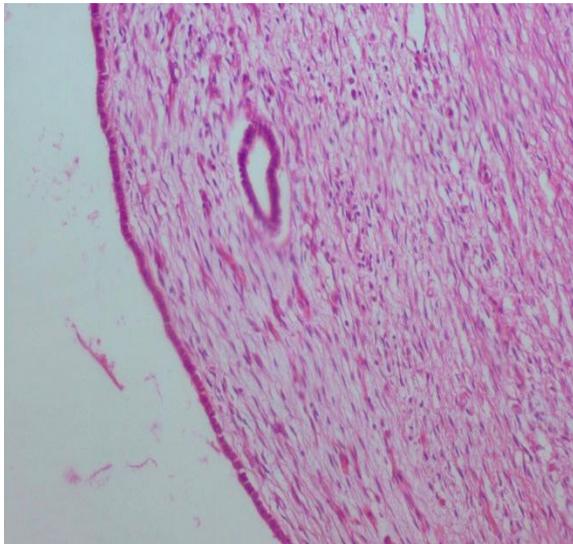
correspondence: FatenHammedi. Department of pathology.FattoumaBourguiba Hospital, Monastir, Tunisia  
E-mail: faten\_hammedi@yahoo.fr

Microscopic findings were consistent with CPDN. The cysts were lined by hobnail shaped eosinophilic cells (Fig. 2).



**Figure 2** Numerous cysts lined by hobnail shaped eosinophilic cells; the septa contain cellular fibrous tissue with bland spindle cells (HEEx40)

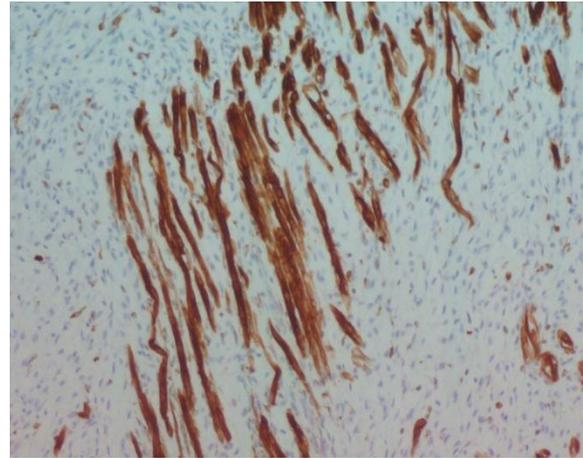
The septa were relatively thick and contained cellular fibrous tissue with bland spindle cells. Focally, were found clusters of rhabdomyoblastic differentiated cells (Fig. 3).



**Figure 3** Focally were found clusters of rhabdomyoblastic differentiated cells (HEEx100)

On one specimen, the stromal septa showed round undifferentiated cells corresponding to the blastemal component. Areas of focal necrosis were present. The adjacent renal parenchyma was normal. There was no microscopic evidence of vascular thrombosis. The lymph node samples showed reactive lymphadenitis with follicular hyperplasia.

Immunohistochemically, rhabdomyoblastic cells were positive for desmin (Fig. 4).



**Figure 4** The rhabdomyoblastic cells were immunoreactive to desmin (IHCx100)

The patient remains alive and free of disease 5 months after treatment.

## CASE REPORT 2

A two-and-a-half-year old male, with no medical history, was presented to the outpatient department with a large tumefaction on his left side of the abdomen. CT tomography of the abdomen demonstrated a roughly 6 x 5 cm multilocular cystic mass arising from the left kidney. These findings evoked nephroblastoma. So the patient received 4 cycles of preoperative chemotherapy according to the SIOP 2001 protocol (the International Society of Paediatric Oncology), using VINCRISTINE and Act-D during four weeks. Then, he subsequently underwent radical nephrectomy.

On gross examination, the kidney presented, on its superior pole, a well defined mass measuring a maximum of 6 cm across. This mass was multiloculated and had an appearance similar to a bunch of grapes.

Histologic examination confirmed the diagnosis of CPDN. The mass was encapsulated. It was composed of multiple variably-sized cysts that contain clear fluid. These cysts were lined by flattened or cuboid cells some of which were oncocytic. The septa contained loose fibrous tissue, admixed with tubules and stubby papillae resembling immature glomeruli. Focally, were seen clusters of small round cells corresponding to the blastemal component. There was neither necrosis nor hemorrhage. The adjacent renal tissue was normal. The vessels were permeable.

The patient remains disease free 32 months after treatment.

## 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.<sup>2,3,4,5</sup> First designed as cystadenoma of the kidney,<sup>6,7</sup> numerous terms have been used to describe these cystic renal tumors: benign multilocular cystic nephroma, polycystic well-differentiated Wilm's tumor, differentiated nephroblastoma, and cystic partially differentiated nephroblastoma. All these designations prove a certain ambiguity, probably due to the controversial pathogenesis of these lesions.<sup>6</sup> The origin of cystic renal tumors has been in the past debated.<sup>6</sup> Firstly, the authors considered them as hamartomatous lesions. Nowadays, these lesions are considered to be of neoplastic nature.<sup>6</sup>

In 1977, Joshi and Banerjee<sup>8</sup> 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 matured or that Wilm's tumor with multicystic areas is simply a precursor for CPDN.<sup>9</sup> Several other pathogenic theories have been proposed in literature. Nevertheless, the relationship between CN, CPDN and Wilm's tumor with multicystic areas remains uncertain.

CPDN is a multilocular cystic neoplasm of very young children, composed of epithelial and stromal elements, along with nephroblastomatous tissue. It is a low-risk malignant tumor.

Patients with CPDN present generally before one-year of age<sup>6,10,11</sup>, most of them are males with a predominance of 2:1.<sup>6,10</sup>

Clinically, patients usually present with non specific symptoms such as a painless abdominal mass<sup>6</sup>. Progressive enlargement, which may be sudden, is reported.<sup>6, 8</sup> Hematuria can be seen and is thought to be due to the extension of the tumor into the pelvicalyceal system.

Standard radiography shows displacement of bowel and adjacent structures by a soft-tissue flank mass<sup>6</sup>. Ultrasonography is more contributory as it reveals the hypoechoic cysts delineated by hyperechoic septae.<sup>6,13</sup> CT image features emphasizes the multicystic architecture. MR imaging is rarely indicated, and it shows classically hypointense signal on T1 and hyperintense signal on T2.<sup>6,13</sup> However, it is important to note that these radiological findings are suggestive and the final diagnosis is retained on the basis of histology.<sup>2</sup>

CPDN is generally a solitary, well circumscribed multiseptated mass often occupies an entire renal pole. This mass is composed entirely of multiple non-communicating fluid filled loculi surrounded by a

thick fibrous capsule and compressed renal parenchyma. There are no solid nodules. Microscopic examination shows numerous cysts lined by hobnail shaped eosinophilic cells. The septa are variably cellular and contain epithelial structures resembling mature renal tubules, and blastema, with or without embryonal stromal. Most tumors contain skeletal muscle, like our case 1. Occasionally, we found cartilage and fat tissue.

Differential diagnosis arises with CN and Wilm's tumor with multicystic areas. All three entities are very similar in their clinical presentation, their imaging features and in their gross appearance<sup>12</sup>. Consequently, they have always posed diagnostic dilemmas.<sup>2,9</sup>

On gross examination, Wilm's tumor with multicystic areas contains expansile nodules.

The key to diagnosis is histological examination which distinguished CPDN from CN depending on the presence and absence of blastemal element in septa walls, respectively. 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.

However, in CPDN, a simple nephrectomy seems to be the best option for treatment. A regular follow up with ultrasound is recommended.<sup>1</sup>

CPDN has generally benign outcome, however, it is considered theoretically as a low grade malignant tumor, as it contains blastemal elements in the cystic walls. These blastemal elements imply the potential for more aggressive behavior.<sup>6,9</sup>

In 1989, Joshi and Beckwith<sup>9</sup> attempted to subclassify CPDN into two groups in order to predict the aggressiveness of this entity. This subclassification was based on the content of mature septal elements. In grade I the septa contained more than 50% mature elements. In grade II, there were less than 50% mature elements in septa.<sup>13</sup>

Some authors reported recurrence, possibly as complication of incomplete resection.<sup>9</sup> Nevertheless, there is no report of metastasis.<sup>1,6,12</sup>

Pelvic involvement in CPDN is extremely rare, thought ureteric extension has been reported.<sup>1,9</sup> In CPDN, ureteric extension has special significance and implies a regular cystoscopy follow up.<sup>1</sup>

In summary, CPDN is considered as a low-risk malignant tumor and is subclassified as category multilocular cystic renal tumors. It is treated by nephrectomy alone with an excellent prognosis.

## REFERENCES

1. Ravi Kumar V R, Rajamani G, Raman M L. Cystic partially differentiated nephroblastoma with ureteric extension. *Indian J Pathol Microbiol* 2010; 53: 831-2
2. Bindhu J, Imtiaz A, Kumar RV, Thejaswini MDRT. Cystic variant of favorable-histology **Wilms'** tumor

- presenting with osteolytic metastasis to the ribs. *J Postgrad Med* 2010; 56: 28-30
3. Eble JN, Bonsib SM. Extensively cystic renal neoplasms: cystic nephroma, cystic partially differentiated nephroblastoma, multilocular cystic renal cell carcinoma and cystic hamartoma of renal pelvis. *Semin Diagn Pathol* 1998; 15: 2-20.
4. Parikh KS, Shukla NS, Shah MP et al. Cystic partially differentiated nephroblastoma. *Indian J Med Pediatr Oncol* 2004; 25: 34-7
5. Van den Hoek J, De Krijger R, Van de Ven K, Lequin M, van den Heuvel-Eibrink MM. Cystic nephroma, cystic partially differentiated nephroblastoma and cystic wilms tumor in children: a spectrum with therapeutic dilemmas. *Urol Int* 2009; 82: 65-70.
6. Agrons G A, Wagner B J, Davidson A J, Suarez E S. Multilocular cystic renal tumor in children: radiologic-pathologic correlation. *RadioGraphics* 1995; 115: 659-660
7. Edmunds W. Cystic adenoma of kidney. *Trans. Path. Soc* 1892; 43: 89.
8. Joshi VV, Banerjee AK, Yadav K, Pathac IC. Cystic partially differentiated nephroblastoma. *Cancer* 1977; 40: 789-795
9. Joshi VV, Beckwith JB. Multilocular cyst of the kidney (cystic nephroma) and cystic partially differentiated nephroblastoma. Terminology and criteria for diagnosis. *Cancer* 1989; 64: 466-79.
10. Singh S, Gupta R, Khurana N. Cystic partially differentiated nephroblastoma: a rare differentiated variant of Wilm's tumour. *J Postgrad Med* 2006; 52 :45-6.
11. Glick RD, Hicks MJ, Nuchtern JG, Wesson DE, Olutoye OO, Cass DL. Renal tumors in infants less than 6 months of age. *J Pediatr Surg* 2004; 39: 522-5.
12. Puvaneswary M, McIntosh J, Cassey J. Cystic partially differentiated nephroblastoma: *Australas Radiol* 2006; 50: 55-7.
13. Wilkinson C, Palit V, Bardapure M, Thomas J, Browning AJ, Gill K, Biyani CS. Adult multilocular cystic nephroma: Report of six cases with clinical, radio-pathologic correlation and review of literature. *Urol Ann* 2013; 5: 13-7