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Cystic Partially Differentiated Nephroblastoma, A Hyperfavourable Variant of Nephroblatoma: A Case Report with Review of the Literature

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Authors' contributions

This work was carried out in collaboration between all authors. Author BAO designed the study, wrote the protocol and wrote the first draft of the manuscript. Authors WTY and CTS managed the literature searches. Authors EIO, BE and DJO performed the surgery. Authors OS, ITA and HM reported the USG and authors CTS, EOU and IVU reported the histological slides. Authors MOO and PA proof read the final manuscript. All authors read and approved the final manuscript.

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ABSTRACT

Cystic partially differentiated nephroblastoma is a rare variant of Wilm's tumor and it represents the hyperfavourable end of the Wilm's spectrum affecting young children.
We described the first documented case from the northern part of Nigeria. The index patient presented with left flank mass. He had unilateral nephrectomy and surgical pathology established its diagnosis.
It is a lower risk but malignant tumor that comes as the differential diagnosis of pediatric cystic renal lesions and need to be differentiated from benign lesions such as cystic nephroma and mixed epithelial and stroma tumor of kidney. It should always be considered as a differential diagnosis of childhood abdominal mass with appropriate investigations and management since it is curable by nephrectomy alone.

Keywords: Cystic partially differentiated nephroblastoma; Wilm's tumor; cystic nephroma.

1. INTRODUCTION

Cystic partially differentiated nephroblatoma (CPDN), an entity in the spectrum of infantile neoplasm with the term CPDN aptly denoting its characteristic gross and microscopic feature is a relatively rare tumor of the kidney usually affecting infants [1] and a variant of nephroblastoma [2]. CPDM is also part of the spectrum of cystic renal tumors of childhood which also include cystic nephroma (CN) and Wilm's tumor with multicystic spaces [3,4]. We are aware of only one documented case from southern part of Nigeria and none from northern Nigeria [5].

CPDN usually follows a benign clinical course and surgery is curative in almost all patients [2,6,7]. Thus the need to differentiate it from other subtypes previously mentioned and from high risk nephroblastoma because of the therapeutic implications. This will ensure that the patients with CPDM are spared from the administration of adjuvant therapy in all clinical situations where it may be unnecessary and potentially harmful [8,9]. Diagnosis has to be made on the basis of histology, as radiological findings are usually inconclusive [4].

We report a case of a 20 month old male-child, who first presented as a 12 month old infant with an 8 month history of left sided abdominal mass, then defaulted and represented 8 months thereafter and underwent radical nephrectomy. Surgical pathology revealed a cystic partially differentiated nephroblastoma.

2. CASE REPORT

A 20 month old male child presented at age 12 months with an 8 months history of left sided abdominal mass. He defaulted to follow up and represented 8 months thereafter with the swelling increasing progressively to its current size. He had no history of abdominal pain, hematuria or symptoms of bowel or urinary obstruction.

Physical examination revealed a chronically ill looking male toddler in no obvious distress. He was afebrile, pale but non-cyanotic and non-icteric with no pedal edema or peripheral lymphadenopathy. Patient weighed 12 kg.

The abdomen was asymmetrically distended with left flank fullness. There was a huge intra-abdominal mass measuring 22 x 20 cm extending from the left subcostal margin to the pelvic brim and crossing the midline. The mass was not tender, oval, firm with smooth surface. It wasn't expansible or pulsatile with no bruit heard over it. Other physical examination findings were essentially normal.

A working diagnosis of left nephroblatoma was made. Contrast enhanced CT scan of abdomen revealed a huge well defined thick walled multi-septated hypodense mildly enhancing mass lesion with calcifications seen at the upper pole of the left kidney and distortion of the renal outline (Fig. 1). The mass measured 18.3 x 17.4 cm. Contralateral kidney was normal.

He was transfused with 15 mls/kg of packed red cells and started on neoadjuvant chemotherapy. A radical nephrectomy was performed with intraoperative findings of 20 cm x 20 cm well encapsulated oval left renal mass (Fig. 2). Apparently normal remnant of the renal tissue was seen at the inferior aspect of the mass. Both hilar and para-aortic lymph nodes were enlarged. Postoperatively, he had one dose of adjuvant chemotherapy which was well tolerated.

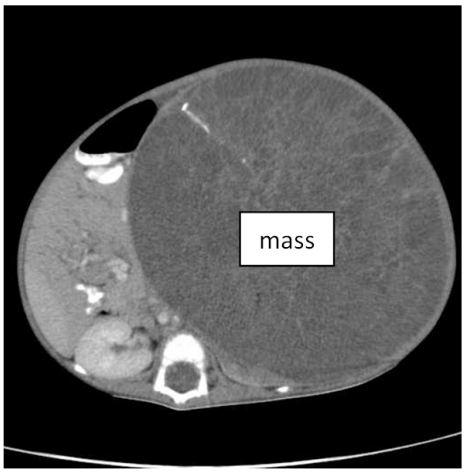

Fig. 1. Axial contrast CT at the level of the kidney showing a huge thin walled cystic mass on the left. There is displacement of the bowel loops to the contralateral side

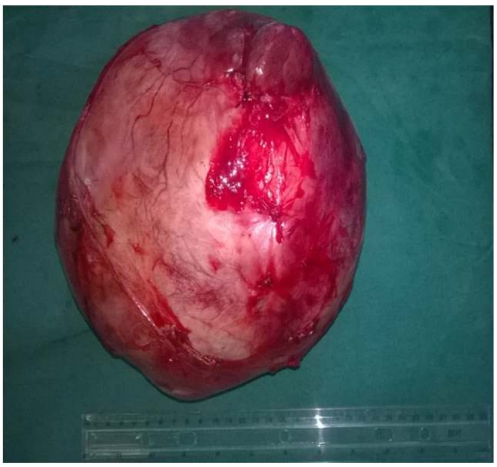

Fig. 2. Well encapsulated mass removed at surgery

Gross examination of the received specimen showed an oval cystic mass weighing 6 kg and measuring 24 cm x 19 cm x 15 cm. Cut sections through the mass revealed a well encapsulated honey-comb surface with multiple micro and macro non communicating cysts measuring 0.4-2.0 cm in diameter and filled with serous fluid (Fig. 3). Remnant of normal kidney tissue was seen at the lower pole.

Histology showed well circumscribed mass with multiple cysts lined by simple epithelium with flattened and cuboidal to hobnail morphology. Blastemal cells are seen within the wall of one cyst and others contain abortive tubules. Elsewhere, areas of focal hemorrhagic necrosis are seen. Based on this and the results of immunohistochemistry study done, the diagnosis of cystic partially differentiated nephroblastoma was made (Fig. 4A, B, C and D).

Fig. 3. Honey-comb surface with multiple micro and macro non communicating cyst

3. DISCUSSION

CPDN is considered as a low risk malignant tumor and sub classified as category multilocular cystic renal tumors. It is a multilocular cystic variant of Wilm's tumor composed entirely of cysts lined by flattened to cuboidal epithelium and separated by delicate septa with the septa stroma often contain foci of blastemal, primitive or immature epithelium and or immature appearing stroma cells which distinguished CPDN from cystic nephroma [2,6,7,10].

The origin of cystic renal tumor has been of much debate in the past but now considered to be neoplastic [11]. The relationship between cystic nephroma, cystic partially differentiated nephroblastoma and Wilm's tumor with multicystic areas remains uncertain. Joshi et al [12] in 1977 differentiated CPDN from cystic nephroma on the presence and absence of blastemal element within the septa respectively. In 1989, Joshi and Beckwith [13] proposed modified terminology and diagnostic criteria to help differentiate cystic renal nephroma from CPDN and other cystic renal tumors such as Wilm's tumor with cyst formation (Table 1). This was refined in 1998 by Eble and Bonsib [2] (Table 2).

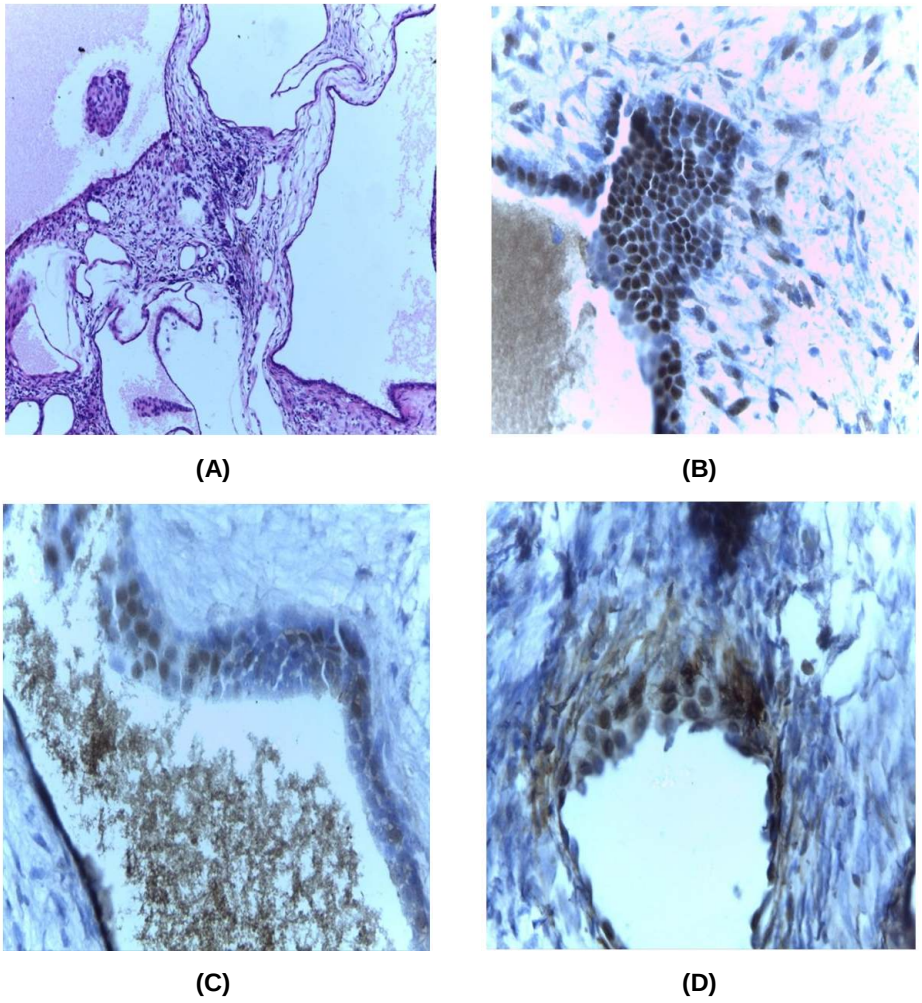

Fig. 4. A composite panel of four pictures showing the histological features of CPDN: A) Histology showed well circumscribed mass with multiple cysts lined by simple epithelium with flattened and cuboidal to hobnail morphology (H&EX40), B, C, D (PRX20, CD10X40, ERX40)

Table 1. Diagnostic criteria for CDPN

Joshi and Beckwith [13]
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 blastemal ± -embroynal stroma or epithelium elements.

Table 2. Diagnostic criteria for CDPN

Eble and Bonsib [2]
1. Pediatric patient, exceptional above age 2 years.
2. Expansile mass surrounded by fibrous pseudo-capsule. Interiorentirely 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 blastemal ± - embroynal stroma or epithelial elements.

CPDN usually present in children between the ages of 4 months to 24 months [2,6] and most of them are males with predominance of male to female ratio of 2:1. The index patient, a 20 months old male falls within the peak incidence of occurrence.

He presented with history of progressive painless abdominal mass which is the usual presentation [2,6,7]. A left flank mass was found on clinical examination. CT image revealed a huge well defined walled multiseptated mass lesion. CT image features emphasizes the multicystic architecture [14]. MR imaging is rarely indicated and it usually shows hypotense signal on T1 and hyperintence signal on T2 [11,15]. It is important to note that radiological findings is retained only on the basis of histology [4,16,17].

CPDN appears grossly as a unilateral, multi-loculated and circumscribed lesion composed of multiple cysts of variable sizes with intervening septae without any solid areas. Microscopically, the cysts are lined by a single layer of flattened, hobnail or columnar epithelium. The stroma between the cysts usually has a fibroblastic, nondescript nature sometimes immunoreactive for hormones [16] along with focal aggregates of blastemal cells, unlike cystic nephroma. Occasionally, tubular and glomerular structures in various stages of development may be seen. However, there is absence of metaplasia and mitosis is rare [14]. Our index case fulfilled these criteria.

For CPDN, a single nephrectomy followed by regular follow up with non-invasive technique seems the best option for treatment [15,18]. The follow up is advised to detect recurrences. This patient was managed using international society of pediatric oncology (SIUP) trials published in 2002 [19] which recommend pre-operative chemotherapy. The main advantage of this treatment protocol is that it significantly reduces the likelihood of tumor spillage, during nephrectomy [20].

An area of hemorrhagic necrosis was found on histology. In Wilm's tumor, extensive necrosis is known to occur following chemotherapy [5]. A diagnosis of a completely necrotic nephroblastoma is made when following pre nephrectomy chemotherapy, no viable tumor is left [9].

CPDN has generally benign outcome with potential for local recurrence, hence need for non-invasive prospective follow up is recommend to document recurrences.

4. CONCLUSION

CPDN, multilocular cystic renal tumor treated by nephrectomy with an excellent progress is a rare differentiated variant of Wilm's tumor with potential for local recurrence. It is considered to be part of the spectrum of nephroblastoma. It should always be seen as differential diagnosis of childhood abdominal tumors for proper diagnosis since it is a curable disease.

CONSENT

Consent was taken from the patient for the publication.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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