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RADIOLOGY CASE REPORTS

CASE REPORT

Cystic partially differentiated nephroblastoma: a rare pediatric renal tumor—case report ☆

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ABSTRACT

Cystic partially differentiated nephroblastoma is a rare renal tumor of childhood. It is part of a spectrum of multicystic renal tumors that also includes cystic nephroma and cystic Wilms' tumor. We present a case of cystic partially differentiated nephroblastoma, highlighting the clinical and imaging diagnostic challenge. Although the histological diagnostic criteria for all these 3 entities are well established, they are clinically and radiologically indistinguishable. Cystic partially differentiated nephroblastoma is often observed in male children under 2 years old. Typical clinical presentations include abdominal masses, abdominal pain and/or hematuria. Patients should be treated according to tumor histology and stage.

Introduction

Cystic partially differentiated nephroblastoma (CPDN) is a rare renal tumor of childhood. It is part of a spectrum of multicystic renal tumors that also includes cystic nephroma (CN) and cystic Wilms' tumor (CWT). It has been proposed that CN, CPDN and CWT are different parts of the same disease spectrum, with CN and CWT representing the benign and malignant ends, respectively, and CPDN being the transition between the 2 [1]. This is analogous to the spectrum of differentiation described in the transition of ganglioneuroma to ganglioneuroblastoma and, finally, neuroblastoma. However, to date, the exact nature of the relation between these 3 entities has not been elucidated [1]. Eble and Bonsib even postulated that CN and CPDN in children could be the same entity and that the term
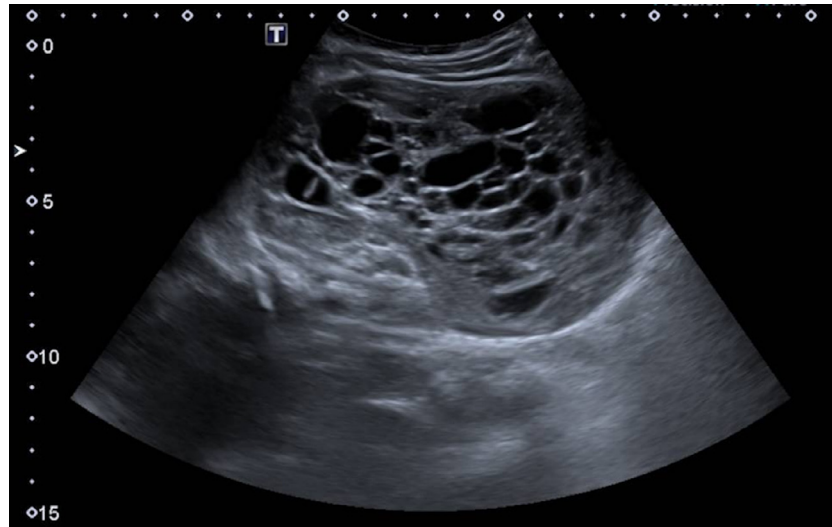
Fig. 1 – Ultrasonography showing multilocular cystic mass with variably sized cystic components and thin echogenic septa located on left renal fossa.

"cystic nephroma" should be reserved for disease that occurs in adults as a neoplasm composed of stroma and epithelium without relationship with Wilms' tumor [2]. CPDN is characterized by multicystic lesions containing blastemal cells with variably differentiated epithelial and stromal elements in the septa [3]. CN contains well-differentiated epithelium without blastemal cells in their septa. CWT in addition to multicystic mass has a solid component that can be associated with hemorrhage and necrosis. There is a close relationship between these entities, where the degree of differentiation determines the type of neoplasm [1]. Although the histological diagnostic criteria of all three are well established, they are clinically and radiologically indistinguishable [4–7]. Treatment may include neoadjuvant chemotherapy, surgery only, surgery and chemotherapy with or without postoperative radiotherapy [4,8].

Case presentation

Previously healthy 34-month-old girl presented abdominal pain and palpable painless abdominal mass on left flank, without fever, prostration, diarrhea or vomiting. At initial evaluation, she was active, hydrated, anicteric, acyanotic, and afebrile. Vital signs were normal.

An abdominal ultrasound was performed and demonstrated a multicystic mass with variably sized cystic components and thin echogenic septa located on left renal fossa (Fig. 1). Laboratory tests showed no abnormalities. She was admitted for diagnostic elucidation. The patient underwent an abdomen and pelvic magnetic imaging resource (MRI) that revealed a large multilocular cystic mass in the left kidney with fluid-equivalent and atypical cysts, without intracystic enhancement and displacing the renal parenchyma medially, measuring 12.5 × 9.9 × 7.5 cm, with estimated volume of 485 cm³. The lesion was characterized by multiple loculations pre-

dominantly hyperintense on T2-weighted sequence and septa of varying thickness (Fig. 2A and B), which were enhanced by intravenous contrast media (Fig. 2C). Hemorrhagic foci were present, especially in the inferior aspect of the mass (Fig. 2D). The hypothesis of Wilms tumor was proposed. Staging Computed Tomography (CT) of the chest showed no secondary lesions.

Four cycles of neoadjuvant intravenous chemotherapy were performed with dactinomycin and vincristine. After that, a restaging MRI scan, performed 38 days after the initial diagnostic imaging, demonstrated increased dimensions of the lesion with an estimated volume of 660 cm³. Therefore, a radical left nephrectomy was performed. During the surgical procedure, moderate amount of free fluid was identified in the abdominal cavity. The oncotic cytology examination of the abdominal fluid showed reactive mesothelial cells; negative for malignant cells. The mass was completely removed along with the perirenal fat, without rupture. Retroperitoneal lymphadenectomy was performed, with removal of hilar, para-aortic and mesenteric lymph nodes. The surgical specimen was referred to anatomopathological examination, which showed a nephrectomy product weighing 900 g, measuring 15.0 × 11.0 × 5.0 cm, accompanied by a 7.5 cm long ureter segment. The histological sections revealed a multicystic lesion, cysts measuring about 2.0 cm in diameter. Microscopy revealed renal parenchyma showing a lesion characterized by cystic areas lined by cuboidal epithelium, without significant atypia and a fusocellular fibrous stroma, without signs of anaplasia (Fig. 3). These characteristics were compatible with CPDN, (low risk after treatment—International Society of Pediatric Surgery/SIOP). The nephrectomy product was margins free.

The patient remains under oncologic follow-up, and no signs of recurrence have been detected. Clinical and laboratory examinations performed so far, have showed no significant abnormalities.
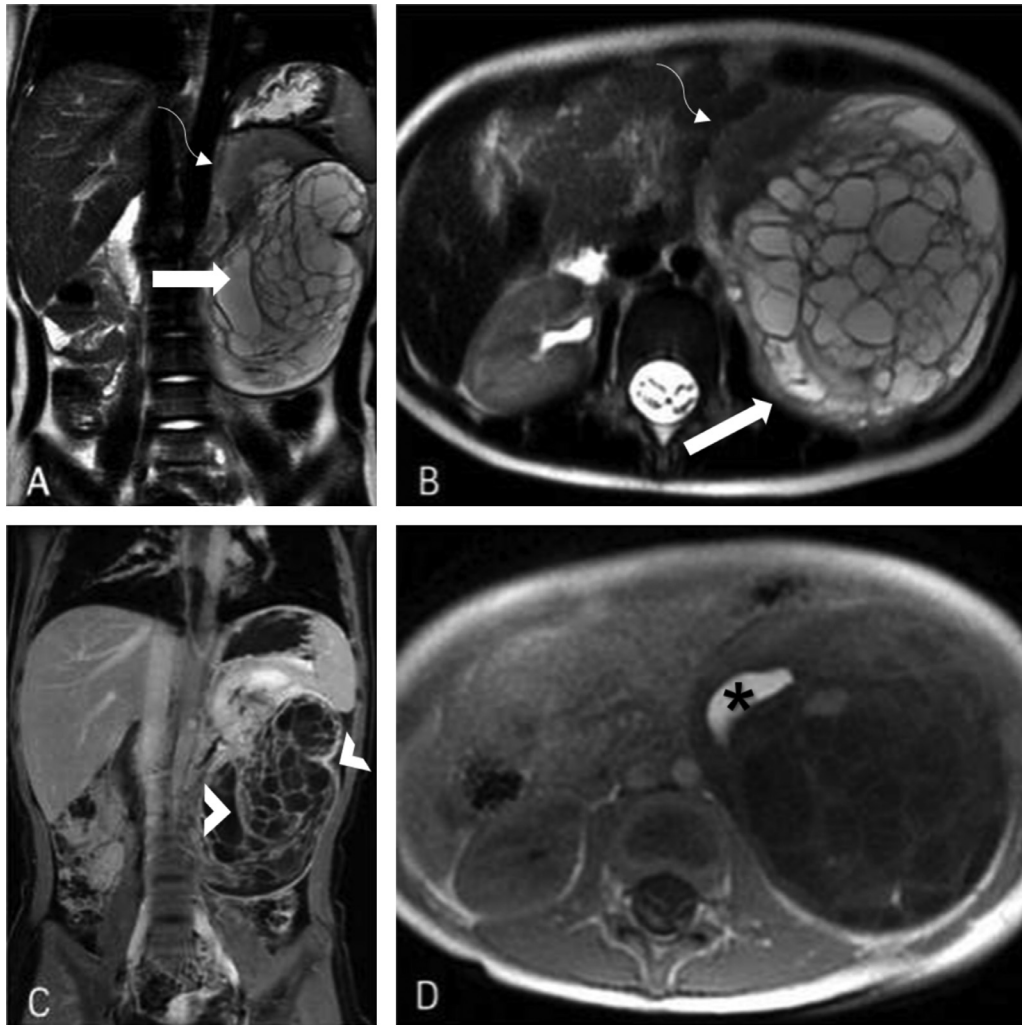
Fig. 2 – Initial diagnostic MRI, before preoperative neoadjuvant chemotherapy, highlighting CPDN main findings. Coronal (A) and axial (B) T2-weighted images show a large multilocular cystic mass (straight arrow), predominantly hyperintense, with septa of varying thickness, displacing the renal parenchyma medially (curved arrow). No solid components were observed. (C) Coronal postgadolinium T1-weitghted fat-saturated image shows septal enhancement (arrowheads). (D) Axial T1-weighted image shows hemorrhagic foci (asterisk) in the inferior aspect of the mass.

Discussion

We present a case of CPDN, highlighting the clinical and imaging diagnostic challenge. Although the histological diagnostic criteria for all these three entities are well established, they are clinically and radiologically indistinguishable [4–7]. Histological criteria for CPDN include the tumor forms a discrete mass, well demarcated from the noncystic renal parenchyma; it is composed entirely of cysts and their thin septa which are the only "solid" portion of the tumor; the cysts are lined by flattened, cuboidal or hobnail epithelium; and the septa contain blastemal cells in any amount, with or without other embryonal stromal or epithelial cell types [3]. CN is composed of multiple septations containing well-differentiated epithelium without blastemal elements. CWT, in addition to the multicystic lesion, has a solid component that may be associated with hemorrhage and necrosis [4,5]. Joshi and Beckwith proposed

diagnostic criteria for the CN, as follows: (1) the lesion is composed entirely of cysts in their septa; (2) it forms a discrete mass, well demarcated from the noncystic renal parenchyma, (3) the septa are the only solid portion of the tumor, conforming to the outlines of the cysts without solid expansile nodules; (4) the cysts are lined by flattened, cuboidal, or hobnail epithelium; and (5) the septa are composed of fibrous tissue in which well-differentiated tubules may be present. The term CPDN was given for those lesions that closely resembled CN but contained immature/blastemal elements in the septa [3]. Both CN and CPDN are devoid of expansive solid nodules, which are characteristic in CWT [5,6,9].

CPDN shows almost no invasion capacity [2,5]. Since CPDN does not recur or metastasize and appears to have a benign course, simple nephrectomy would be recommended [10]. However, 2 cases have been reported in children with intra-abdominal recurrence without metastasis; one case possibly occurred due to incomplete resection of the lesion [3] and the
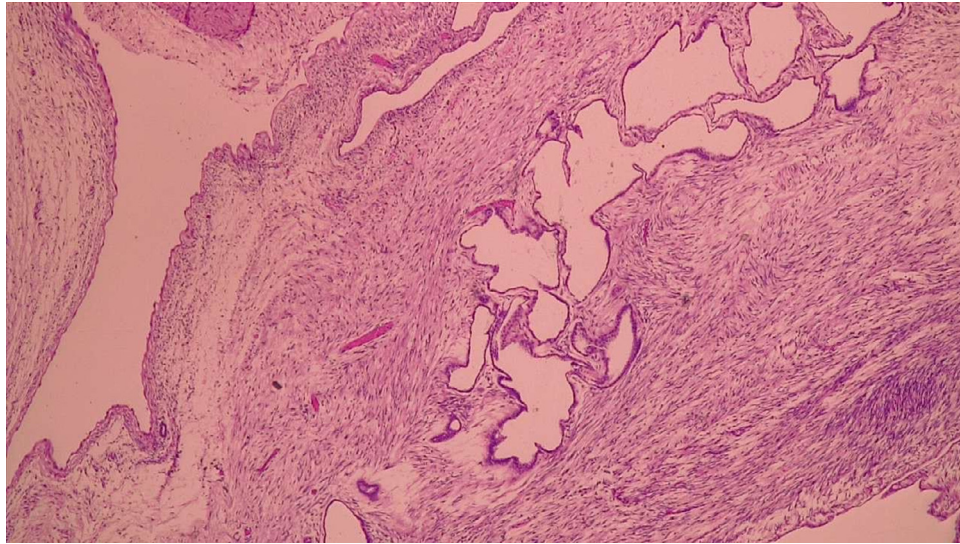
Fig. 3 – Detail of the septal wall lined by simple epithelium and blastomatous stroma, predominantly cellular and fibrous/fusocellular—HE, 200×.

other due to cysts rupture [11]. There is also a case of bilateral CPDN recurring as bilateral CWT [1]. This transformation suggests that there is a close relationship between the three entities [1,3].

CPDN is often observed in male children under 2-year old [1], in disagreement with our case that occurred in a 34-month-old girl. The incidence of CPDN is around 0.5%, according to Société Internationale d'Oncologie Pédiatrique (SIOP) and National Wilms tumor study group [12]. Typical clinical presentations include abdominal masses, abdominal pain and/or hematuria [13], which were observed in the present case, except for hematuria that was absent. Patients should be treated according to tumor histology and stage. Usually, preoperative chemotherapy is also given in the low risk group (stage I) [12]. However, the SIOP Working Classification of Renal Tumors of Childhood recommends primary resection as the treatment of choice in cystic renal tumors with no preoperative chemotherapy [12,14]. Postoperative treatment for high risk tumors will be more aggressive than for the intermediate risk tumors [14]. The dilemma is that the distinction between CN/CPDN and malignant neoplasms, such as CWT, is unreliable through preoperative imaging studies. Wilms' tumor with cystic changes is classified as intermediate risk tumor and therefore should be treated with preoperative chemotherapy, according to SIOP protocols. Graf et al showed that preoperative chemotherapy results in a lower rate of tumor rupture during surgery [15]. Partial nephrectomy is considered safe for CN and CPDN [16–19]. However, there is the potential risk of recurrence in CPDN incomplete excision [2,3]. Due to the impossibility of a definitive preoperative histopathological diagnosis, radical nephrectomy is recommended, limiting partial nephrectomy to special cases, such as those patients with single kidney or bilateral renal tumors [12].

Although preoperative chemotherapy is often performed, it can delay definitive treatment. Our case shows that preoperative chemotherapy did not bring any benefit, on the contrary, an increase in tumor dimensions was observed, corroborating the SIOP recommendation of primary resection as treatment of choice.

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