A rare pediatric renal tumor: Cystic partially differentiated nephroblastoma

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Abstract

The histological features of cystic partially differentiated nephroblastoma (CPDN) require a thorough sampling of the surgical specimen and the application of immunohistochemical techniques to confirm the diagnosis.

KEYWORDS

cystic partially differentiated, kidney, nephroblastoma, pathology, pediatric renal tumors

1 | CLINICAL IMAGE

Cystic partially differentiated nephroblastoma (CPDN) occurs more commonly in males under 2 years of age with a unilateral cystic renal mass.^{1,2} Only histopathological examination of the surgical specimen establishes with certainty the diagnosis. Despite its usual benign course, CPDN may show aggressive behavior and has a tendency for recurrence following surgery because of the presence of blastemal cells in septa.¹

Previously healthy 4-month-old boy underwent radical left nephrectomy for suspected congenital mesoblastic nephroma. Macroscopically, the total renal parenchyma was practically occupied by a welldemarcated mass that was entirely multicystic with no solid or expansile areas. Cysts were separated by thin septa and contained serous fluid (Figure 1A). Microscopically, the tumor was characterized by multiple cystic areas, lined by cuboidal epithelium, without significant atypia. These cysts were separated by loose stroma and contained blastema and epithelium element such as immature tubules (Figure 1B,C). Immunohistochemical study showed that immature epithelium structures between cysts were positive for WT1 (Figure 1D). Based on the above features, diagnosis of CPDN was given.

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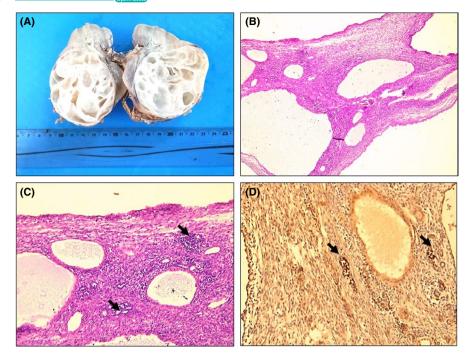


FIGURE 1 (A) Gross appearance of the specimen showing total renal parenchyma practically occupied by a well-demarcated mass measuring $6.5 \times 5 \times 3.5$ cm. This mass is multicystic with no solid expansile areas, and the cysts are separated by thin septa. (B) Histological findings showing multiple cystic areas separated by loose stroma with no expansile or solid foci (HE ×40). (C) The cysts are lined by cuboidal or hobnailing epithelium, without significant atypia. The intervening stroma appeared primitive and oedematous, containing blastema and epithelium element such as immature tubules and compressed glomeruloid structures (black arrows) (HE ×100). (D) Immunohistochemistry, immature tubules showing intense positivity for WT1 (black arrows)

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CONFLICT OF INTEREST None declared.

AUTHOR CONTRIBUTIONS

Seifeddine Ben Hammouda prepared the manuscript. Nouha Ben Abdeljelil guided the author in writing the manuscript and proofread the final manuscript.

ETHICAL APPROVAL

All procedures performed were in accordance with the ethical standards. The examination was made in accordance with the approved principles.

CONSENT

Published with written consent of the patient.

DATA AVAILABILITY STATEMENT

In accordance with the DFG Guidelines on the Handling of Research Data, we will make all data available upon request.

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