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Cystic Partially Differentiated Nephroblastoma in a Child: A Case Report

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Abstract **Case Report**

Cystic partially differentiated nephroblastoma is a rare cystic variant of Wilms tumor characterized by a multiloculated architecture with cystic septa containing blastemal, epithelial, and stromal components. Its distinction from multilocular cystic nephroma may be challenging, particularly after neoadjuvant chemotherapy, which can modify histological features. We report the case of a 4-year-old girl treated with preoperative chemotherapy according to the SIOP protocol. Nephrectomy revealed a well-circumscribed, encapsulated, multiloculated cystic renal mass measuring 7.5 cm, with an estimated tumor volume reduction of 15 percent. Histological examination showed preserved triphasic differentiation, with predominant mesenchymal stroma, 20 percent epithelial differentiation, and focal blastemal remnants comprising 10 percent of the tumor. Immunohistochemistry demonstrated strong nuclear WT1 expression in blastemal and epithelial components, cytoplasmic vimentin positivity, and nuclear PAX8 expression in epithelial structures. This case highlights the importance of identifying residual triphasic differentiation and using immunohistochemistry to distinguish cystic partially differentiated nephroblastoma from cystic nephroma, thereby ensuring accurate diagnosis and appropriate management

Keywords: Wilms tumor, cystic partially differentiated nephroblastoma, pediatric renal tumor, WT1, PAX8.

INTRODUCTION

Cystic partially differentiated nephroblastoma is an uncommon renal neoplasm in pediatric patients, constituting less than 5 percent of Wilms tumors. Histologically, it is characterized by a multiloculated cystic lesion wherein the septa comprise nephroblastomatous elements, including epithelial, blastemal, and stromal components. According to the SIOP protocol, preoperative chemotherapy can change the shape of tumors a lot. The regression of the blastemal component in cystic tumors may complicate diagnosis and lead to misidentification as benign cystic lesions. A comprehensive histological examination remains essential. We present a case of CPDN in a four-year-old child undergoing neoadjuvant chemotherapy to elucidate the diagnostic characteristics and the function of immunohistochemistry (IHC).

CASE PRESENTATION

A four-year-old girl exhibited a right-sided abdominal mass that was incidentally observed by her parents. Ultrasound and computed tomography of the abdomen showed a multicystic mass in the right kidney that did not involve lymph nodes or spread to other parts of the body. The patient underwent preoperative chemotherapy in accordance with the SIOP protocol. Imaging done after treatment showed that the tumor volume had shrunk by about 15%. The patient then had a right nephrectomy.

Macro-Pathological Findings

The nephrectomy specimen was 10 × 8 × 6 cm and weighed 150 g. The tumor was well-defined and surrounded by a capsule, with a maximum diameter of 7.5 cm. It looked like a multiloculated cystic mass with clear serous fluid when cut. No solid nodules, bleeding,

invasion of the capsule, or growth outside of the kidneys were found.

Histopathological Findings

Microscopic examination revealed a triphasic intrarenal tumor. The cystic septa were mostly made up of mesenchymal tissue that had fibrous and myxoid characteristics. About 20% of the tumor was made up of epithelial cells, which formed tubular and glomeruloid structures. About 10% of the tumor was made up of scattered blastemal foci, which were small round cells with hyperchromatic nuclei. There were changes related to chemotherapy, such as focal necrosis and stromal hyalinization. There was no sign of anaplasia or invasion of blood vessels. There was no tumor at the surgical margins.

Immunohistochemical (IHC) Findings

Immunohistochemical staining revealed strong nuclear WT1 expression in both epithelial and blastemal components. Vimentin was present in mesenchymal and blastemal regions, whereas epithelial components exhibited nuclear positivity for PAX8. These findings validated nephroblastomatous differentiation and corroborated the diagnosis of cystic partially differentiated nephroblastoma.

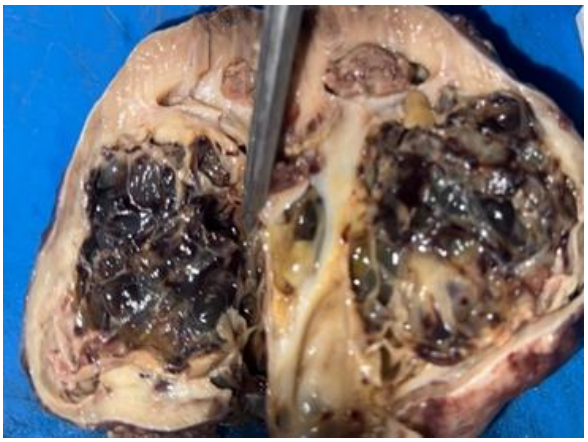

Figure-1: Nephrectomy with multilocular cystic neoplasm

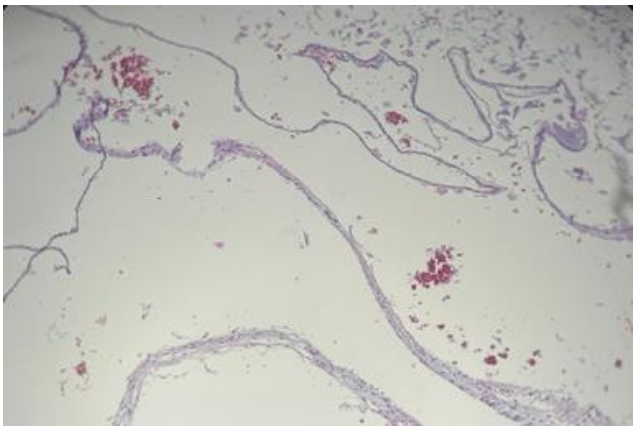

Figure-2: Cystic neoplasm

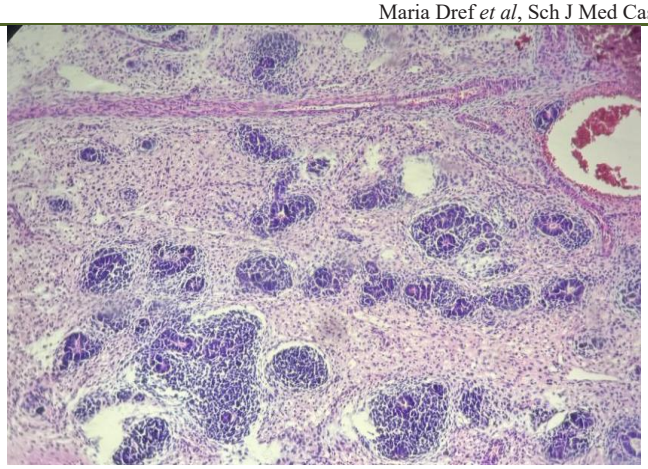

Figure-3: Blastemal component

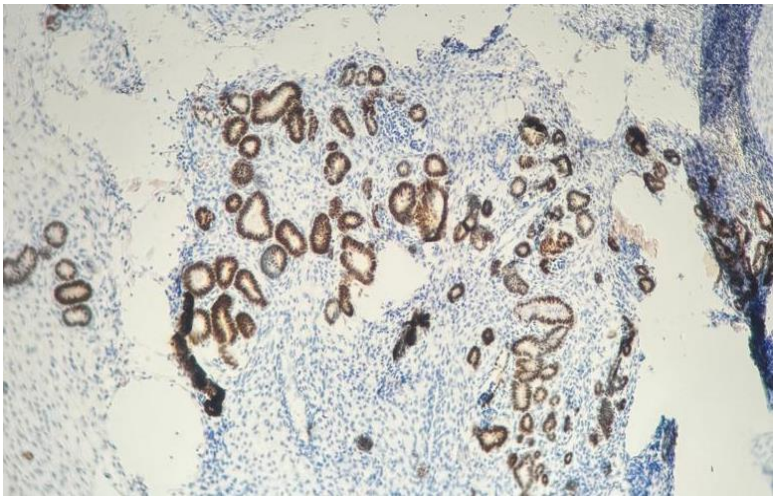

Figure-4: Nuclear expression of PAX8 in the epithelial component

DISCUSSION

Cystic partially differentiated nephroblastoma is situated between classic Wilms tumor and multilocular cystic nephroma. The primary diagnostic difficulty lies in differentiating it from cystic nephroma, especially when blastemal components are scarce or modified by chemotherapy.

In this instance, the existence of residual epithelial and blastemal components within the cystic septa was essential for diagnosis. The immunoprofile that included both WT1 and PAX8 expression gave more evidence that the cells came from the kidneys. The response of tumors to neoadjuvant chemotherapy in CPDN is inconsistent. The prognosis is generally excellent when completely excised and confined to the kidney. But if you classify something wrong, it could lead to the wrong treatment, either too much or not enough.

Awareness of CPDN and meticulous histopathological evaluation are crucial, particularly in the post-chemotherapy context.

CONCLUSION

Cystic partially differentiated nephroblastoma is an uncommon cystic variant of Wilms tumor. An accurate diagnosis depends on finding residual triphasic differentiation and confirming it with immunohistochemistry using WT1 and PAX8. To get the best treatment and follow-up, it's important to correctly identify this entity.

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