

# Cystic partially differentiated nephroblastoma in an infant: A case report

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## Abstract

Cystic Partially Differentiated Nephroblastoma, a variant of the Multilocular Cystic Renal Tumour, is considered as the bridge between Cystic Nephroma and Cystic Wilm's Tumour. It usually occurs in children below the age of two years with predominance in male. Histopathology is considered as diagnostic modality. Neoadjuvant chemotherapy and adjuvant radiotherapy may benefit the patient in selected cases. Here, a five-month-old boy who presented with painless gradually increasing right sided abdominal mass is reported. Imaging and fine needle aspiration cytology findings were inconclusive to reach the diagnosis. The histopathology after a radical nephrectomy concluded the diagnosis of Cystic Partially Differentiated Nephroblastoma.

**Key words:** Infant; Nephroma; Wilm's tumour.

## INTRODUCTION

Cystic Partially Differentiated Nephroblastoma (CPDN) is a variant of a rare form of renal tumour called Multilocular Cystic Renal Tumour. It is considered as the transition stage between Cystic Nephroma (Benign) and Cystic Wilm's Tumour (Malignant).<sup>1,2</sup> Therefore, a case of five months old boy, who was brought with gradually increasing abdominal mass. Clinical assessment, imaging, and needle aspiration cytology were inconclusive to

reach the diagnosis. Histopathology after nephrectomy confirmed the case. Although such cases have been addressed in the literature, the authors did not find any cases reported from Nepal.

## CASE REPORT

A five months old boy was brought with complaints of gradually increasing right upper quadrant mass for one month with whitish discharge in urine. The infant had no fever, haematuria, weight loss, or painful micturition. Abdominal examination showed large 10x10 cm dimension firm, non-tender, ballotable mass in the right hypochondriac region that was extending to right lumbar region. His haemoglobin level was 10.2 gm/dl with normal kidney function test. Urine analysis also showed normal findings.

Ultrasonography of abdomen showed complex cystic lesion (98X88 mm) with multiple internal septations in right loin and non-visualisation of right kidney (Figure 1). Sonological differential diagnoses were exophytic hydatid cyst of liver and complex renal cyst (? Renal Mass). Computerised Tomography (CT) Scan of abdomen revealed large well-defined round to oval cystic lesion of size 8.7 X 8.6 X 8.0 cm with multiple thin internal septations (maximum thickness 2.0 mm) was noted from right kidney causing compression of its parenchyma. There was a mild enhancement of the septae on post contrast study. Superiorly, it was abutting the inferior

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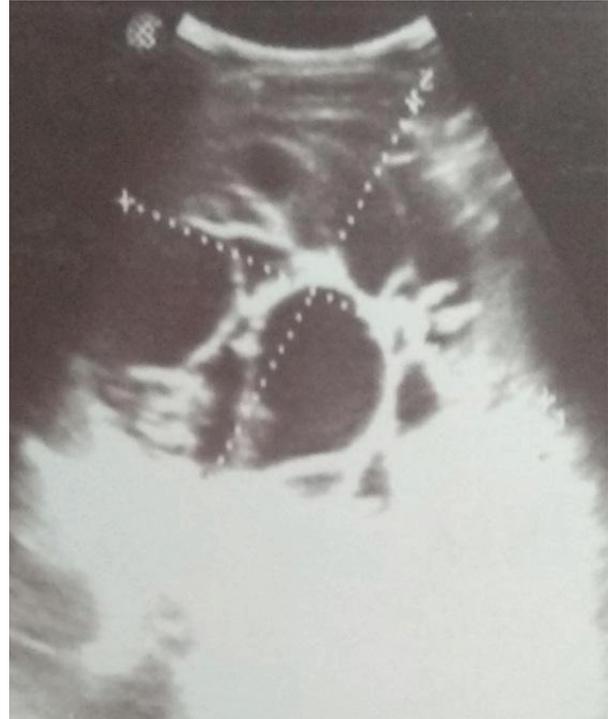


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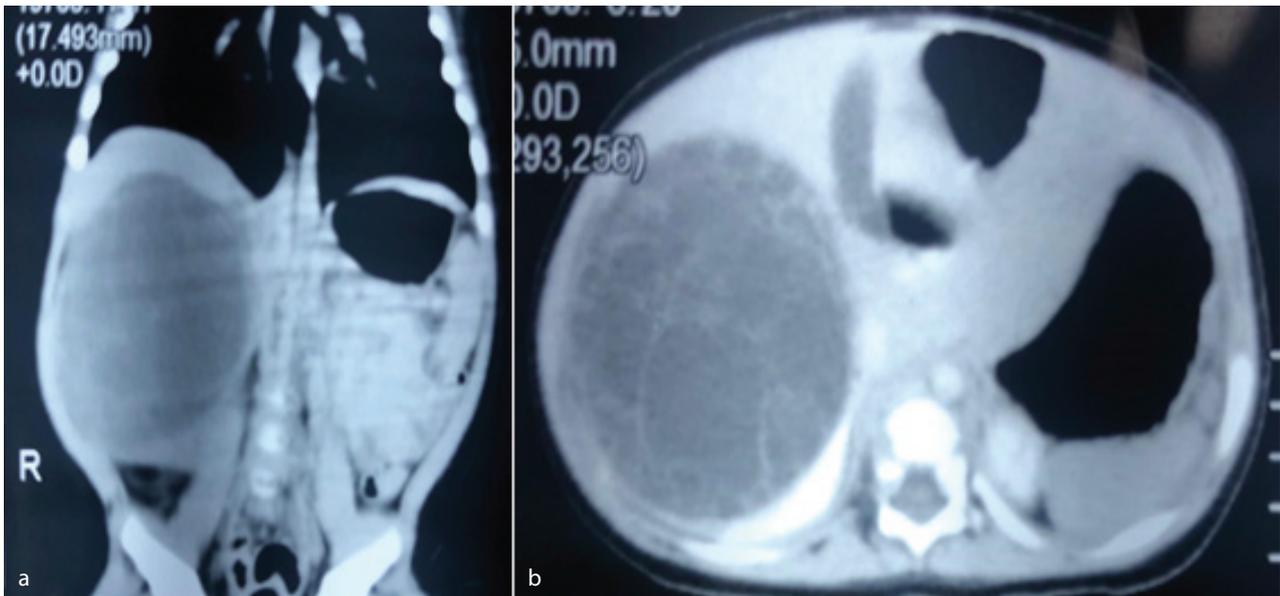
surface of liver and laterally abutting the abdominal wall with main abdominal wall with maintained intervening fat plane. Pelvicalyceal system was not dilated. There was no evidence of excretion of contrast within the loculi. No solid component and calcifications were noted. The features were suggestive of Multilocular Cystic Nephroma with differential diagnosis of Partially Differentiated Cystic Nephroblastoma (Figures 2a, 2b).

Ultrasonographic guided Fine Needle Aspiration from the above mentioned lesion showed features suggestive of Small Round Cell Tumour. Ultrasonographic guided Tru-cut biopsy could not be done because the lesion was multicystic. Therefore, he was planned for right sided radical nephrectomy. Intraoperatively, there was multicystic mass measuring 8X8 cm<sup>2</sup> from upper pole of right kidney with well-maintained fat planes and mass not invading renal vein, inferior venacava, and adrenal gland. Later, the gross description on histopathology showed most of the right kidney specimen involved with cystic areas. There were thin walled septa containing clear to mucinous material in the cystic cavity. Solid area, necrotic areas, or haemorrhage were not seen. Normal renal parenchyma was pushed to periphery by the cystic lesion. Microscopic description showed sections from kidney composed of entirely of cysts separated by thin septa. Cysts were lined by flattened to cuboidal cells. There was variable cellularity of the septa as well as in the underlying stroma. There were round to oval tumour cells with scant cytoplasm blastemal in appearance. Focally, septa were protruding into the

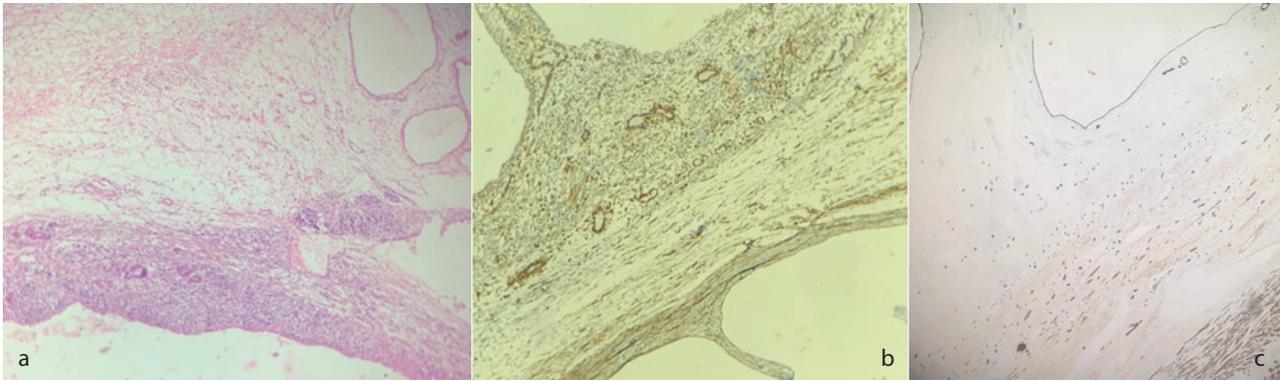
epithelium. Immunohistochemistry done with Vimectin and Pancyceratin were positive for both. The features were conclusive for Cystic Partially Differentiated Nephroblastoma with ureteric margin, renal hilum, a perirenal sinus, and lymphnodes free of the tumour (Figure 3a, 3b, 3c).



**Figure 1:** Sonological finding of complex cystic lesion with multiple internal septations in right kidney



**Figures 2a, 2b:** CT finding of large well-defined round to oval cystic lesion with multiple thin internal septations from right kidney. There is mild enhancement on septae on post contrast study



**Figure 3:** (3a) Hematoxylin and Eosin stain picture showing cystic area and blastemal cells, (3b) Vimectin stain showing the same findings, (3c) Pancytokeratin stain showing the similar findings

## DISCUSSION

Cystic Nephroma (CN), CPDN and Multicystic Wilm's Tumour are the three variants of Multilocular Cystic Renal Tumour (MRCT). CPDN is considered as intermediate from between CN (benign) and Cystic Wilm's Tumour (malignant).<sup>1</sup> It accounts for less than 1% of all Wilms' tumour cases. Commonly, they occur before the age of two years with male predominance (Male:Female = 2:1).<sup>3</sup> Female may show biphasic age distribution like 20 years and fifth-sixth decades of life.<sup>4</sup> Childhood CPDN is uncommon renal tumour. Bilateral CPDN have been reported.<sup>5</sup>

Gradually increasing painless abdominal mass and haematuria are the common presenting features.<sup>6</sup> The variants of CPDN are clinically and radiologically indistinguishable.<sup>4,2</sup> Histology distinguishes the variants and is of therapeutic importance.<sup>2</sup> Lesion containing variable size cysts is well circumscribed from the remaining kidney parenchyma by a fibrous pseudocapsule grossly. CPDN is characterised by presence of blastemal components within the thin septa. The cysts are lined by flattened, cuboidal or hobnail or lack lining epithelium. Skeletal muscle and myxoid mesenchyme are present in the septa of most tumours. Cartilage and fat are present occasionally.<sup>1,7</sup> Cystic Wilm's Tumour has expansile nodule with solid masses of nephroblastomatous component, whereas, CN is characterised by presence of discrete mass with intervening fibrous solid septa.<sup>8</sup> Simply, the presence of blastemal cells or poorly differentiated stromal or epithelial elements should exclude the diagnosis of multilocular cyst or CN.<sup>9</sup>

Other differential diagnoses are segmental multicystic dysplastic kidney, localised cystic kidney disease, polycystic kidneys, and renal hydatid. Polycystic kidneys are diffusely involving bilateral kidney with cysts in other organs.<sup>7</sup> Multicystic dysplastic kidneys have no contrast excretion.<sup>2</sup> Hydatid cysts are characterised by presence of daughter cysts within the mother cyst.<sup>4</sup>

CPDN is believed to be a pathologic derivative of the metanephric blastema. It is not related to renal dysplasia. It is considered as high risk malignant neoplasm due to aggressive appearance of its cell.<sup>9</sup> Stage I CPDN is appropriately managed nephrectomy without need of adjuvants therapy. Nephrectomy alone has been found curative in case of tumor localised within the kidney.<sup>1</sup> Therefore, a case of CPDN is accurately diagnosed and is not managed like a case of high risk nephroblastoma.<sup>1-3</sup> It is necessary to distinguish CPDN from rest of cystic renal lesions for therapeutic purpose. Regular follow-up is recommended after surgery.<sup>7</sup>

## CONCLUSION

CPDN is a rare variant of MRCT. It is not possible to distinguish it from CN and Multicystic Wilm's Tumour clinically and radiologically. Histopathology distinguishes it from rest of the mimicking renal lesions. Stage I CPDN is better managed by nephrectomy without any need of adjuvant measures.

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