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Case Report

# Cystic Nephroma and its Varied Management Scenarios: A Report of two Cases

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**Abstract**

Cystic nephroma is a rare benign cystic neoplasm of the kidney. The preoperative diagnosis with its malignant counterparts cystic partially differentiated nephroblastoma or cystic Wilms' tumor is not easy but is important when one is considering for nephron-sparing surgery.

**Keywords:** *Cystic nephroma, cystic tumors of the kidney, nephron-sparing surgery*

## Introduction

Cystic nephroma (CN) also called multilocular CN comprises a rare multicystic benign renal mass without any solid components. It is important to differentiate it from its malignant cousins such as cystic partially differentiated nephroblastoma (CPDN) or cystic Wilms' tumor (CWT), as CN is at the benign end of the spectrum and does not require radical management.

We report our experience of two cases of CN managed with different strategies. The purpose of presenting is to make pediatric surgeons aware of its clinical, radiological, and histopathological characteristics and also to highlight the available options for surgical resection.

## Case Reports

### Case I

A 10-month-old healthy female infant of 8 kg presented to us with complaints of an abdominal lump noticed by parents 15 days back. On abdominal examination, there was a large firm, well-defined mass, involving almost whole of abdomen.

Contrast-enhanced computed tomography (CECT) abdomen [Figure 1a] revealed a large well-defined thin-walled cystic mass of size 14 cm × 13 cm × 11 cm with hypoattenuating contents and multiple thin enhancing internal septations. The mass was involving the upper two-third of kidney and extending up to the renal hilum but sparing the renal vasculature. The rest of the renal parenchyma was unremarkable. There was no invasion of adjacent structures, and the contralateral kidney was normal. Furthermore, there was no evidence of lymphadenopathy. These findings were highly suggestive of benign cystic renal mass, and so without any further investigations, following the National Wilms' Tumor Study group (NWTS) protocol, upfront surgery was planned. The infant was taken up for laparotomy with a plan of frozen section and nephron-sparing surgery (NSS). On laparotomy, there was a large well-capsulated mass of about 15 cm × 12 cm × 10 cm, arising from the upper pole of the left kidney involving almost 90% of the left kidney. The mass was extending into the hilum without any definite surgical plane near hilum. Considering the large

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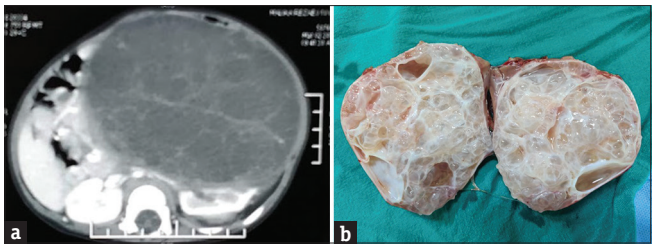

**Figure 1:** (a) Contrast-enhanced computed tomography and (b) gross specimen showing well-capsulated multicystic mass arising from left upper pole of kidney without any solid component

size of mass with extension into the hilum, left radical nephroureterectomy along with hilar and para-aortic lymph node sampling was done. The cut section of specimen showed multiple cysts [Figure 1b], which on histology was lined by flattened epithelium separated by fibrous septa, consistent with CN. The postoperative period was uneventful, and the child is asymptomatic on follow-up of 2 years.

### Case II

A 4-year-old healthy male child presented with complaints of an abdominal lump noticed by parents 1 month back, which was progressively increasing in size. The child was evaluated, and a CECT abdomen [Figure 2a] was suggestive of a right renal multicystic mass of approximately 10 cm × 8 cm × 8 cm. The appearance of mass was very similar as described in the previous case, suggestive of benign cystic tumor, most likely CN. It was localized to the upper pole of the right kidney, with well-defined margins and compressing the lower pole. The mass was separate from the renal vessels and adjacent viscera with no lymphadenopathy. The vascular supply of the lower pole was intact and was separate from the mass on the arterial phase. On laparotomy [Figure 2b], the mass was localized to the upper pole of the right kidney. Upper pole vessels were ligated, and mass was enucleated, preserving the lower one-third of the kidney. The frozen section of the mass was suggestive of benign lesion. The lower pole was repaired and fixed in the retroperitoneum. Histopathology confirmed the diagnosis of CN. The postoperative period was uneventful, and the child is on follow-up for the past 9 months.

## Discussion

CN is a rare benign multicystic tumor of the kidney seen in infants/young children and adult females. Because of its similarities with other cystic renal tumors, a lot of controversies exist regarding the terminology, classification, and natural history. It forms the benign end of the spectrum of cystic tumors of the kidney, the others being CPDN and CWT, which has malignant components. Most of

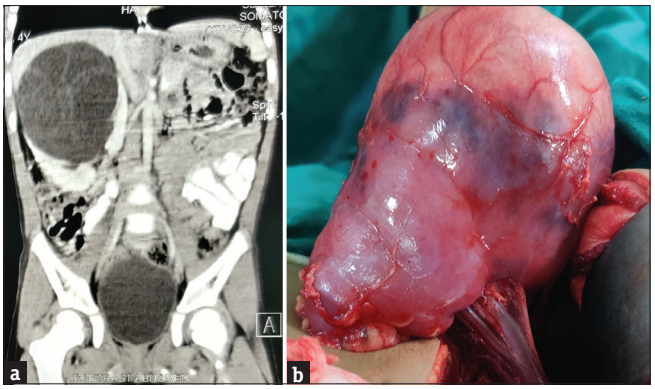

**Figure 2:** (a) Contrast-enhanced computed tomography showing right upper pole multicystic mass with spared lower pole and its vessels and (b) intraoperative picture with upper pole mass and spared lower one-third renal parenchyma

the cases recorded are younger than 4 years of age, with a male predominance of 2:1. CPDN and CN comprise 0.5% of all pediatric renal tumors registered in the International Society of Pediatric Oncology (SIOP))/German society of Paediatric Oncology and Hematology.[1,2]

CN is usually a unilateral and sporadic tumor. The exact pathogenesis is not known. However, there are reports of bilateral involvement and familial cases, which are usually associated with a mutation in the DICER-1 gene. Some reports also categorize pediatric CN as a distinct entity as it is associated with a mutation in the DICER-1 gene, which is also associated with pleuropulmonary blastoma.[3]

In children, presentation is similar to that of Wilms' tumor with an abdominal mass noticed incidentally as was seen in both our cases, however, can also present with abdominal pain, hematuria, or hypertension.[4,5]

Because of lack of any characteristic findings on imaging preoperative diagnosis is always challenging and can be fallacious.[4] Main limitation is that imaging cannot distinguish between malignant and benign cysts.[4] Ultrasound findings correspond to a category III mass in Bosniak classification of renal cystic masses with malignant potential of 60%. As seen in both of our cases, on CECT, these masse were well-circumscribed and multiloculated with enhancing septa in between. The absence of a solid component can only help in differentiating CN and CPDN from CWT.[6]

The goal of the operative procedure is to excise all tumor tissue with maximum preservation of normal renal parenchyma. Enucleation or partial nephrectomy should be attempted in small, localized CNs to preserve functioning renal parenchyma. However, a total nephrectomy is indicated when CN replaces most of the kidney. No further treatment is required after the definitive surgical

procedure.[2,5,7] The SIOP protocol also recommends CN and CPDN be treated primarily by surgery, and CWT by a combination of neoadjuvant chemotherapy and surgery.[2,4] There has been no ambiguity in the management of CN, as upfront surgery has been recommended by both NWTS and SIOP. Kurian *et al*.,[4] in his experience of nine cases of pediatric multiloculated cystic tumors, highlighted the diagnostic and therapeutic dilemma associated with these tumors and recommended a similar approach of upfront surgery, although at the same time, the author has raised the concern for tumor spillage. In a review of six pediatric patients with cystic renal neoplasms, van den Hoek *et al*.[8] concluded that, in cystic neoplasms without solid components, surgery should be considered first, and whenever feasible, enucleation should be done with no further therapy.

In the first reported case, although we planned for NSS, it was not feasible as it was a large mass involving almost the whole of kidney and extending up to the hilum. While in the second case, the tumor was localized to the upper pole of the right kidney, and so enucleation could be done preserving the normal renal parenchyma. However, the tumor was sent for a frozen section to confirm its benign nature as it is important to differentiate it from the malignant cystic renal tumors. The differential diagnosis with CPDN can be made intraoperatively by frozen sections to find out if the septa contain any blastemal elements.[9] A complete enucleation without spillage[4] is mandatory as recurrence has been reported in the literature, and this has been attributed to an incomplete enucleation.[10]

Histopathology in both of our cases showed circumscribed, encapsulated mass containing multiple noncommunicating fluid-filled cysts that were lined by flattened cuboidal epithelium separated by fibrous septa without any blastemal elements. The absence of blastemal cells and poorly differentiated stromal and/or epithelial elements confirmed the diagnosis of CN.

The revised criteria for the diagnosis of CN are a discrete mass composed entirely of cysts; the cysts are lined by flattened, cuboidal, or hobnail epithelium. The septa are composed of fibrous tissue, in which well-differentiated tubules may be present. A similar definition has also been used in SIOP.[2] However, CPDN, which has low malignant potential has blastemal elements, nephroblastomatous epithelial elements, and undifferentiated or differentiated mesenchyme.[1] WT is the malignant end of the spectrum and is characterized by triphasic elements characterized by blastemal, stromal, and epithelial elements.

To conclude, CN is a rare benign renal tumor, and its surgical management can be variable depending on its size and location. Preoperative imaging may not be useful to differentiate it from other cystic renal tumors, and the only definite way is to differentiate it by histopathology. We recommend that surgery should be the first line of treatment, and every attempt should be made to perform NSS after ruling out malignancy with the help of intraoperative frozen section biopsy.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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## Conflicts of interest

There are no conflicts of interest.

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