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Case report

# Cystic nephroma: A multicystic renal neoplasm

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ABSTRACT

A 42-year-old female presented with a complaint of abdominal pain for 3 months. Deep palpation revealed tenderness in the left lumbar region. Ultrasonography and contrast-enhanced computed tomography revealed a multiloculated cyst in the left kidney. The results of guided fine needle aspiration cytology were inconclusive. On exploration, the renal lump was found to be a smooth, well-circumscribed swelling protruding from the anterior surface of the left kidney, with no extrarenal infiltration. Left nephrectomy was performed. The cut surface of the resected specimen showed a well-circumscribed tumor containing large cystic spaces within the renal tissue. Histopathological examination revealed cystic nephroma, which is a rare neoplasm of the kidney. The postoperative course was uneventful. The patient was discharged on the 5th postoperative day. Now, at 1-year follow-up, the patient is well without any signs of recurrence on imaging.



## 1. Introduction

Cystic nephroma (CN) is a rare kidney lesion that clinically presents with loin pain or hematuria. Investigations typically reveal a well-circumscribed, multiloculated mass in the kidney, and excision is generally curative. Although the lesion is benign, the suspicion of malignancy dictates nephrectomy/nephron-sparing surgery. Here, we present the case of a 42-year-old female with CN that was surgically excised.

## 2. Case report

A 42-year-old female presented with a complaint of pain in the left upper quadrant of the abdomen for 3 months. Deep palpation revealed moderate tenderness in the left lumbar region.

Ultrasonography revealed a multiloculated cyst in the left kidney, whereas the results of guided fine needle aspiration cytology were inconclusive (predominantly blood). Contrast-enhanced computed tomography of the abdomen (Figs. 1 and 2) revealed a multiloculated and well-defined cyst in the left kidney, which was interpolar in position with otherwise normal function and no evidence of direct spread or lymphadenopathy. On exploration, the renal lump was found to be a smooth, well-circumscribed swelling protruding from the anterior surface of the left kidney, with no extrarenal infiltration (Figs. 3 and 4). Left nephrectomy was performed. The cut surface (Fig. 5) of the resected specimen revealed a well-circumscribed tumor containing large cystic spaces within the renal tissue.

Histopathological examination showed cysts of various sizes that were lined by either single layers of cuboidal cells or hobnail columnar cells, the absence of clear cells with primitive tubules in certain areas, and compressed nephrons, indicating a diagnosis of CN (Fig. 6).

The postoperative course was uneventful. The patient was discharged on the 5th postoperative day. She was on a 3-monthly review, with CT scan every 6 months for the 1st year. Now, at 1 year follow-up, the patient is well without any signs of recurrence on imaging.

## 3. Discussion

CN is a rare kidney neoplasm, and only approximately 200 cases have been reported to-date. The lesion is associated with a bimodal age distribution, with males being affected between 3 months and 2 years of age, and females being affected after 30 years of age.[1]

Microscopically, CN is characterized by well-demarcated, multiple, noncommunicating, fluid-filled cysts lined by cuboidal or flattened epithelial cells and separated by septae. Blastemal cells

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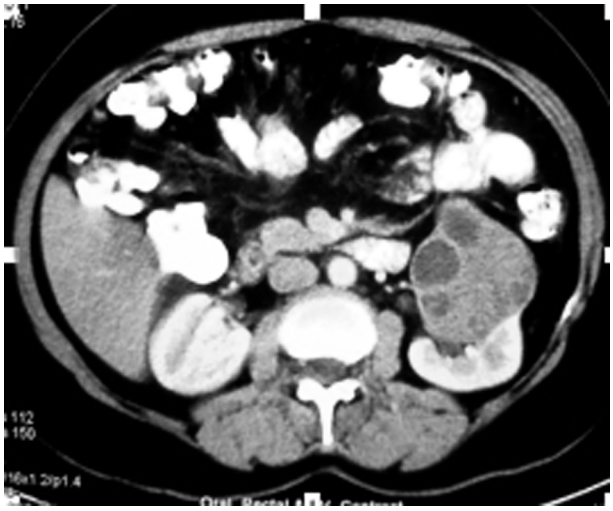

**Fig. 1.** Axial computed tomography showing cystic change in the left kidney.

are generally absent. The embryological origin is from a segmental form of renal dysplasia that is related to polycystic kidney disease or maldevelopment of the ureteric bud. CN is also considered to be a neoplasm occurring at the benign end of the wide spectrum of renal tumors, with a cystic variant of Wilms tumor at the other extreme and cystic partially differentiated nephroblastoma in the center.[2] Cystic mixed epithelial and stromal tumor of the kidney (MESTK) is sometimes referred to as a CN. Any blastemal content, however, classifies this lesion as malignant and excludes it from the category of CN lesions.[3] Solid variants of MESTKs, which often contain female genital tissue or intestinal tissue, are separately classified. Stromal elements may contain spindle cells and stain positive for muscle markers (desmin and smooth muscle actin) and HMB-45, which is an antibody to a glycoprotein in pre-melanosomes that reacts with melanoma cells, junctional nevus

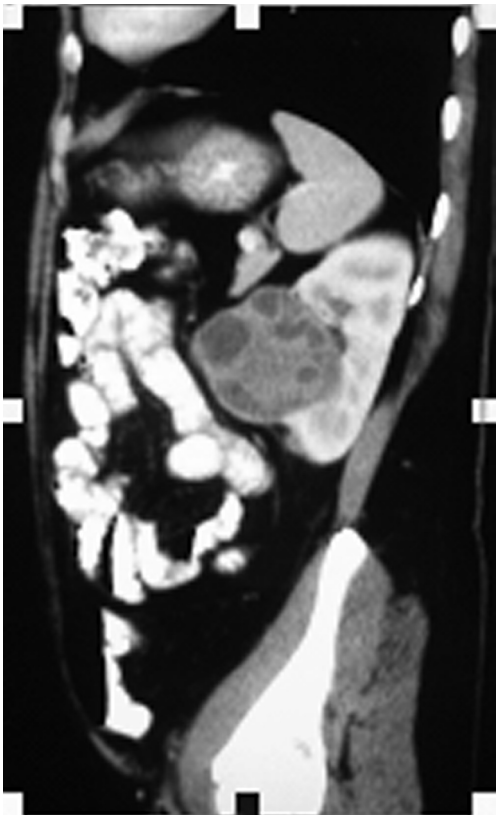

**Fig. 2.** Parasagittal reconstruction on computed tomography.

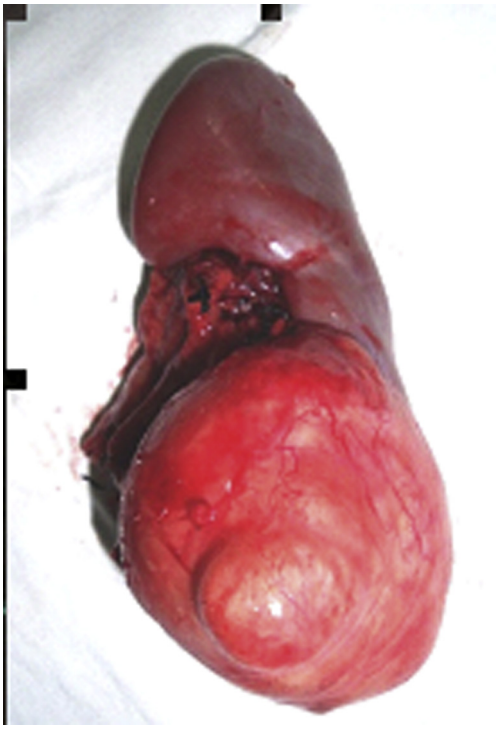

**Fig. 3.** Anterior view of the tumor.

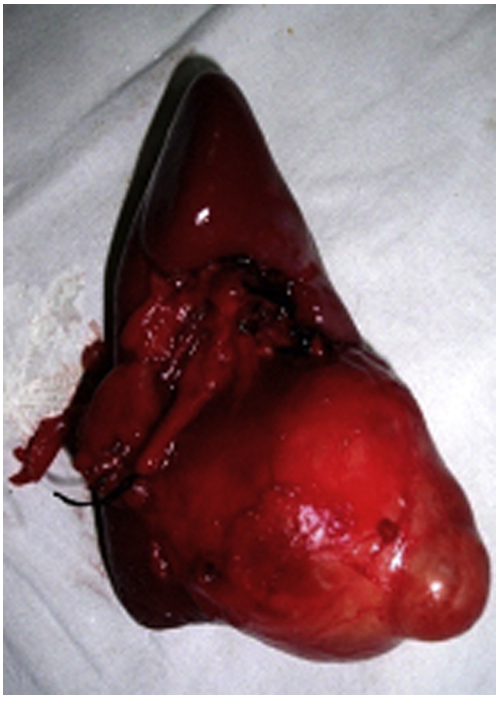

**Fig. 4.** Relationship of the tumor to the hilum.

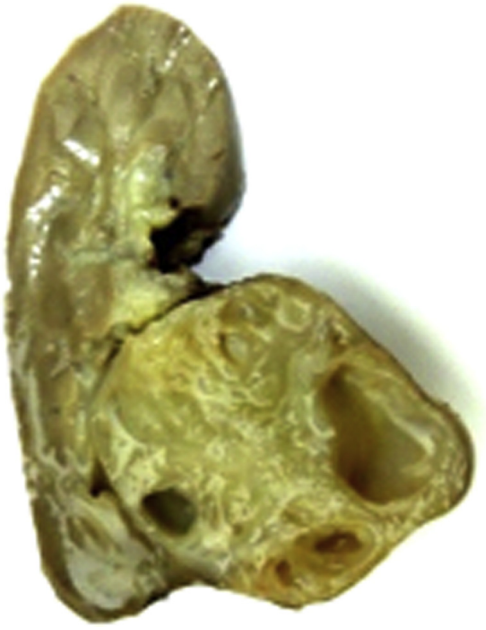

**Fig. 5.** Cut surface showing cysts.

cells, and fetal melanocytes. It is nonreactive with almost all non-melanoma human malignancies, with the exception of rare tumors showing evidence of melanogenesis, such as pigmented schwannoma, clear cell sarcoma, or tumors associated with the tuberous sclerosis complex, such as angiomyolipoma and lymphangiomyoma. CD34, which is broadly expressed in mesenchymal neoplasms, is absent, indicating a benign nature.[4–6] CN and MESTK share similar clinical, pathological, immunohistochemical, and genomic characteristics.[7] The amount of stromal elements distinguishes the two. A unifying term, renal epithelial and stromal tumor, has also been proposed.[8] Genetically, constitutional DICER1 haploid insufficiency predisposes individuals to a broad range of tumors, including CN. Families suspected to have DICER1 syndrome should undergo mutation testing.[9]

The differential diagnosis includes multilocular cystic renal cell carcinoma (RCC); multicystic kidney; segmental cystic disease; other cystic RCCs, including cystic necrosis (pseudocystic necrotic carcinoma); cystic hamartoma of the renal pelvis; clear cell sarcoma; and a hydatid cyst.[10–13] The clinical features in adults include flank pain, hematuria, hypertension, and urinary tract infection.[14] Our patient experienced mild pain and tenderness, with just a palpable lump in the upper left quadrant of the abdomen. Ultrasonography and contrast-enhanced computed tomography are performed for diagnosis, and the latter clearly reveals clear encapsulated cysts with contrast uptake by the intervening tissue,[15] similar to our finding. Fine needle aspiration cytology is generally inconclusive because of the paucity of tissue and remains controversial.[12,16,17] Treatment is mainly surgical, particularly in view of the symptoms and the inability to exclude malignancy. Nephron-sparing surgery can be attempted if the mass measures <4 cm and is unilateral, unifocal, and localized, and when follow-up with imaging is possible.[18] Although CN is benign, cystic RCC from multilocular CN has been reported following incomplete excision/partial nephrectomy; therefore, complete histological evaluation of biopsy samples is necessary.

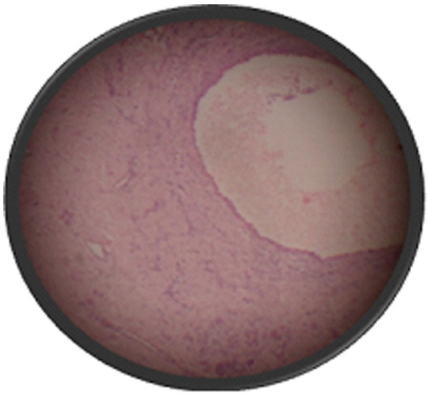

**Fig. 6.** Histopathological appearance showing the cyst.

In conclusion, CN is a rare kidney neoplasm with a good prognosis after surgical treatment. It is difficult to exclude malignancy, and a final diagnosis is made by histopathology. Nephrectomy/nephron-sparing excision with clear margins is recommended.

This report emphasized that CN is of neoplastic rather than developmental or hamartomatous origin. Cystic RCC can arise from multilocular CN, highlighting the need for complete excision and thorough histological examination to rule out coincidental RCC. Accumulation of several similar cases is necessary to assess whether cystic multilocular nephroma is a predisposing factor for renal malignancy.

## Conflicts of interest

The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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