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**Case Report**

# Cystic nephroma in a 14-year-old-a rare case report

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## ABSTRACT

Cystic nephroma (CN) is an uncommon benign renal tumor with a bimodal age distribution. It often presents with non-specific presenting symptoms. In this study, we report a case of a 14-year-old female patient who presented with a painless abdominal mass, and a left renal cystic lesion was detected on CECT KUB. Left partial nephrectomy was done and a diagnosis of CN made following the histopathological examination of specimen. Owing to its non-specific clinical presentation and radiological findings, it is difficult to distinguish CN from other cystic lesions of kidney like cystic partially differentiated nephroblastoma, tubule-cystic renal cell carcinoma, cystic Wilm's tumor etc., pre-op. This study emphasizes importance of histopathological examination in diagnosis of CN to prevent misdiagnosis and overtreatment. Although it has good prognosis, long term follow-up for local recurrence is recommended.

**Keywords:** CN, Renal tumour, Case report, Histopathology

## INTRODUCTION

Cystic nephroma (CN) is a rare benign cystic renal tumor. It is predominantly encountered in two age groups; adults and the pediatric population, mostly involving infants and young children. According to literature, the first case of CN was reported as cystic adenoma of the kidney by Edmunds in 1892.[1] The important histological clues which aid in the diagnosis of CN include: presence of cysts lined by flat, cuboidal, or hobnail epithelium and septa variably lined by fibrous and/or ovarian-like stroma.[2] Pediatric CN has frequently been associated with mutations in the DICER-1 gene. It often presents with non-specific clinical symptoms like painless abdominal mass and is also sometimes detected incidentally. Pediatric CN is considered to be in the benign end of the spectrum of cystic renal neoplasms, with cystic partially differentiated CN (CPDN) and cystic Wilms tumor occupying the other end of the spectrum.[3]

## CASE REPORT

This was a case of a 14-year-old female, who came to the urology outpatient clinic with complaints of a painless abdominal lump. She had no urinary symptoms or hypertension. There was no family history of renal diseases/ neoplasms as well. On clinical examination, there was a non-tender palpable mass in the left upper abdomen. Her blood investigations and renal profile was unremarkable. CECT KUB was done which revealed a multilocular cystic mass showing a sharp interface with adjacent renal parenchyma, arising from the left kidney (Figure 1). A CT guided biopsy was done. On histopathological examination of the biopsy, the sections studied showed cysts lined by cuboidal epithelium with no evidence of malignancy, following which, a left partial nephrectomy was done for the same. Intraoperatively, multiloculated cysts were noted in the upper pole of the kidney, and pus was drained from the cysts which was sent for culture and sensitivity testing which showed negative results. The specimen was sent to the Pathology department for frozen section and for further histopathological examination.

On gross examination, the partial nephrectomy specimen measured 5.5×3×2.5 cm. External surface was found to be smooth with an intact capsule. The cut section appeared cystic (multiple cysts were seen in the cortex

and medulla), with smallest measuring 0.1 cm in diameter and the largest measuring 0.8 cm in diameter (Figure 2).

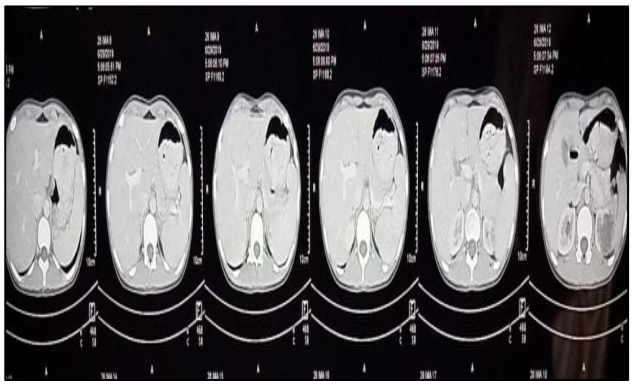

**Figure 1: Multilocular cystic mass in the upper pole of the left kidney.**

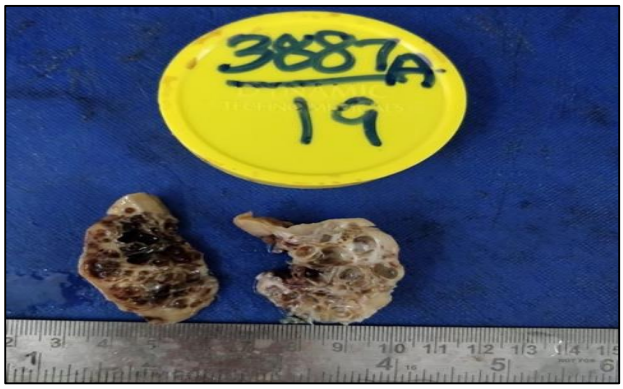


**Figure 2: Gross appearance.**

On microscopy, multiple sections studied showed a rim a of renal parenchyma with multiloculated cysts with septa composed of fibrous tissue (Figure 3). The cysts were lined by cuboidal epithelium (Figure 4) with hob nailing in various cysts (Figure 5). Focal areas in the wall showed structures resembling differentiated renal tubules, inflammatory cells and blood vessels (Figure 6). No blastemal components or glomerular structures or cartilage or skeletal muscle fibers were noted**.** On the basis of these features, a diagnosis of CN was made.

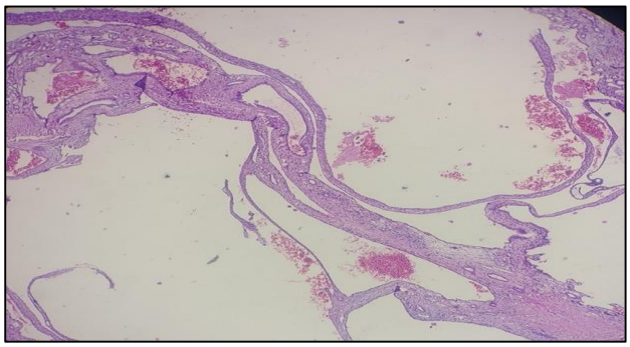

**Figure 3: Multilocular cystic spaces with intervening fibrous septa (20x).**

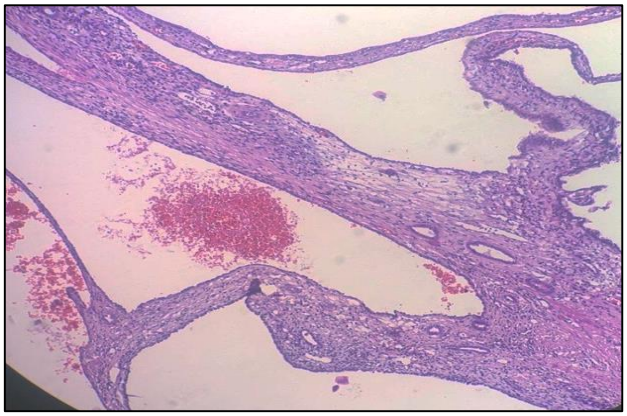

**Figure 4: Cystic spaces lined by cuboidal epithelium with renal tubules in the fibrous septa (40x).**

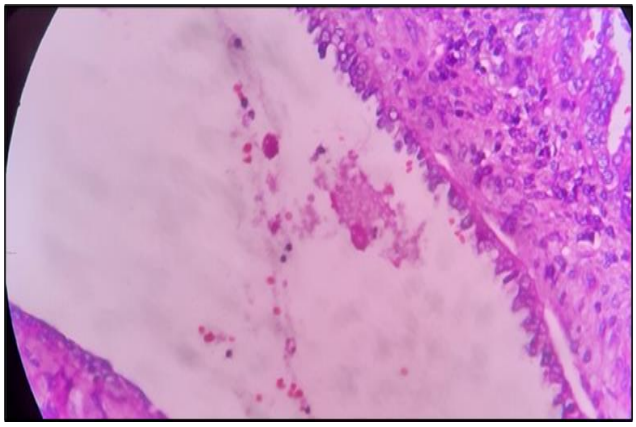

**Figure 6: Hob nailing in focal areas.**

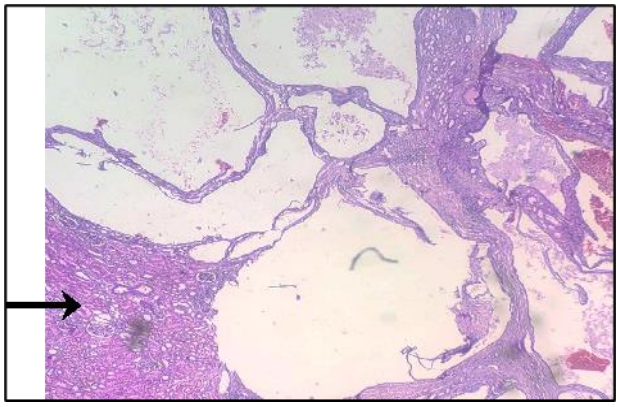

**Figure 5: Mature renal tubules (marked by ➔) in the wall.**

## DISCUSSION

CN is a well circumscribed tumor, which is solely multicystic in nature with a well-defined epithelium and is characterized by the presence of mature nephrogenic components.[3]

As mentioned earlier, CN occurs in 2 distinct age groups. Adult CN is usually predominant in females in the 4$^{th}$-5$^{th}$

decade; whereas, pediatric CN is usually reported in the age group between 3 months to 4 years with a slightly male preponderance.[4] In a study conducted by Cajaiba et al out of 44 cases of pediatric CN studied from 2007-2013, only 2 cases presented beyond 49 months of age (at 12 and 14 years of age).[3] This proves the rarity of the occurrence of CN in the age group of our present case.

The 2016 WHO classification of tumors of the urinary system and male genital organs, considers adult CN to be a part of the mixed epithelial and stromal tumor family of tumors (MEST), because of the similarity in their morphological and immunohistochemical profiles.[5] Pediatric CN was previously considered to be unilateral and sporadic in nature, but many studies in the recent decades have proved that they can be familial in nature and occur bilaterally as well.[6] Clinically, CN presents with symptoms like painless, palpable abdominal mass, abdominal pain, hematuria, etc., and is usually incidentally detected on routine physical examination or in radiological studies.

On ultrasonographic imaging, CN appears as a unilateral encapsulated mass with irregular cysts and septa of variable thicknesses and falls under category III mass in the Bosniak classification of renal cystic masses. On computed tomography, it is seen as a circumscribed and multilocular cystic tumor with several hypo-attenuated components and no contrast excretion into the cystic components.[7]

The histological diagnostic criteria for CN proposed by Eble and Bosnib are: (1) an expansile mass surrounded by a fibrous pseudo capsule whose interior is composed entirely of cysts and septa, with no expansile solid nodules. (2) The cysts should be lined by flattened, cuboidal, or hobnail epithelium. (3) The septa may contain epithelial structures resembling mature renal tubules but should not contain epithelial cells with clear cytoplasm and should be free of skeletal muscle fibers.[8]

Many authors consider pediatric CN to be a part of the spectrum of cystic renal diseases of childhood namely cystic partially differentiated nephroblastoma (CPDN) and cystic Wilms tumor. Several recent studies have documented pediatric CN to be associated with mutations in the DICER-1 gene, and is found to frequently co-exist with familial pleuropulmonary blastoma (PPB).[4]

The diagnosis of CN can conclusively be made only with the help of histopathological examination, since it is radiologically and clinically identical to CPDN and cystic Wilm's tumor. The lack of blastemal/ undifferentiated renal components differentiates CN from other pediatric cystic lesions of the kidney especially CPDN. Cystic Wilm's tumor, which occupies the malignant end of the spectrum of cystic renal diseases, usually has the presence of a solid component associated with necrosis and hemorrhage.[8-10]

Other entities which closely resemble adult CN radiologically are multi-cystic renal cell carcinoma (MCRCC) and tubulocystic renal cell carcinoma. Histopathologically, MCRCC differs from CN by the presence of evident solid areas in cystic mass or extensile nodules of clear cells.[11] Tubulocystic renal cell carcinoma is characterized by the presence of cysts of variable sizes which are lined by a single layer of cuboidal, flattened or hobnail cells with eosinophilic cytoplasm, spherical nuclei and prominent nucleoli. The cysts are separated by thin fibrous septa.[12]

Both adult and pediatric CN exhibit diffuse stromal immunoreactivity for estrogen receptor. The presence of ropy collagen, ovarian type stroma and immunoreactivity to inhibin is more characteristic of adult CN than its pediatric counterpart.[10]

The primary objective of the operative procedure is elimination of all tumor tissue with conservation of the remaining viable renal tissue as much as possible. Enucleation or partial nephrectomy is the treatment of choice in small CNs to preserve functioning renal parenchyma.[12] However, a total nephrectomy will be required in cases of larger CNs that replace most of the renal parenchyma. After the definitive surgical procedure, no further treatment is necessary.[13]

## CONCLUSION

The crucial role of histopathological examination in arriving at the correct diagnosis of CN is indispensable because of the overlapping clinical and radiological features of the various cystic renal lesions. An accurate intraoperative diagnosis of CN by frozen section can aid in considering treatment options like partial nephrectomy or enucleation as the treatment of choice instead of radical procedures. Follow-up of the patients postoperatively is often advocated in view of recurrence, malignant transformation or metastasis.

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## REFERENCES

1. Edmunds W. Cystic adenoma of kidney. Trans Pathol Soc. 1892;43:89-90.
2. Mukhopadhyay S, Valente AL, De la Roza G. Cystic nephroma: a histologic and immunohistochemical study of 10 cases. Arch Pathol Lab Med. 2004;128(12):1404-11.
3. Cajaiba MM, Khanna G, Smith EA, Gellert L, Chi YY, Mullen EA et al. Pediatric cystic nephromas: distinctive features and frequent DICER1 mutations. Human Pathol. 2016;48:81-7.
4. Huang SH, Chen TJ. Cystic nephroma of childhood: a case report and review of the literature. Urolog Sci. 2016;27(3):171-3.

5. Moch H, Cubilla AL, Humphrey PA, Reuter VE, Ulbright TM. The 2016 WHO classification of tumours of the urinary system and male genital organs-part A: renal, penile, and testicular tumours. Europ Urol. 2016;70(1):93-105.
6. A1 Bahubeshi, Bal N, Rio Frio T, Hamel N, Pouchet C, Yilmaz A et al. Germline DICER1 mutations and familial cystic nephroma. J Med Genet. 2010;47:863e6.
7. Granja MF, O'Brien AT, Trujillo S, Mancera J, Aguirre DA. Multilocular cystic nephroma: a systematic literature review of the radiologic and clinical findings. Am J Roentgenol. 2015;205(6):1188-93.
8. Eble JN, Bonsib SM. Extensively cystic renal neoplasms: cystic nephroma, cystic partially differentiated nephroblastoma, multilocular cystic renal cell carcinoma, and cystic hamartoma of renal pelvis. In Seminars in Diagnostic Pathology. 1998;15(1):2-20.
9. Joshi VJ, Beckwith B. Multilocular cyst of the kidney (cystic nephroma) and cystic, partially differentiated nephroblastoma. Cancer. 1989;64:466-79.
10. Li Y, Pawel BR, Hill DA, Epstein JI, Argani P. Pediatric cystic nephroma are morphologically, immunohistochemically, and genetically distinct from adult cystic nephroma. Am J Surgical Pathol. 2017;41(4):472.
11. González-Serrano A, Cortez-Betancourt R, Alías-Melgar A, Botello-Gómez PJ, Ramírez-Garduño E, Trujillo-Vázquez EI et al. Multilocular Cystic Renal Cell Carcinoma or Cystic Nephroma? Case Rep Urol. 2016;2016.
12. Moch H. Cystic renal tumors: new entities and novel concepts. Advances in anatomic pathology. 2010;17(3):209-14.
13. Boybeyi Ö, Karnak İ, Orhan D, Ciftci AO, Tanyel FC, Kale G et al. Cystic nephroma and localized renal cystic disease in children: diagnostic clues and management. J Pediatr Surg. 2008;43(11):1985-9.

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