



## Case Report

# A rare presentation of cystic nephroma in a young adult



Amit Sharma\*, Mukund Andankar, Hemant Pathak

Department of Urology, TNMC & BYL Nair Hospital, Mumbai, Maharashtra, India

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### KEYWORDS

Cystic nephroma;  
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**Abstract** Cystic nephroma is a rare benign renal lesion with non-specific presenting symptoms. This tumour has a bimodal age distribution presenting in children between 3 months and 2 years and in adults over 30 years of age. There is male preponderance with a 2:1 male to female ratio in the childhood whereas it is commoner in females in adults. It is uncommon in the age-group of 5 years–30 years. We describe a case of cystic nephroma in a 25-year-old female which was managed by partial nephrectomy.

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## 1. Introduction

Cystic nephroma is an uncommon, benign renal tumour of uncertain aetiology predominantly seen in paediatric population. It was first described by Edmunds in 1892 in an 18-year-old girl [1,2]. Since then, approximately 200 such cases have been reported in the literature [1]. In the past, this tumour has been variously described as multilocular cystic tumour, renal multilocular cyst, multilocular cystic nephroma, renal cystadenoma and partial polycystic kidney [1]. It was considered to be a developmental tumour with a malignant potential [1]. Recently, there has been increased awareness about this tumour [1]. Surgery is the mainstay of

treatment and also helps to exclude malignancy. We describe a young female who presented with flank pain.

## 2. Case report

A 25-year-old female was admitted with complaints of intermittent dull aching left-sided flank pain of 7 days' duration. There was no history suggestive of urinary tract infection (UTI) or stone disease. At admission, she was stable; per abdomen examination revealed a non-tender ballotable well-defined mass of 6 cm × 5 cm of soft to cystic consistency in the left flank region (Fig. 1). Her blood investigations including haemogram, renal and liver function tests and coagulation profile were normal. Urine examination showed occasional red blood cells, few pus cells, 20–25 epithelial cells and no sugar or albumin. An abdominal ultrasound revealed a moderate size (8 cm × 7 cm × 6 cm) cystic lesion abutting lower pole of left kidney suggesting

\* Corresponding author.

E-mail address: [dramiturology@gmail.com](mailto:dramiturology@gmail.com) (A. Sharma).

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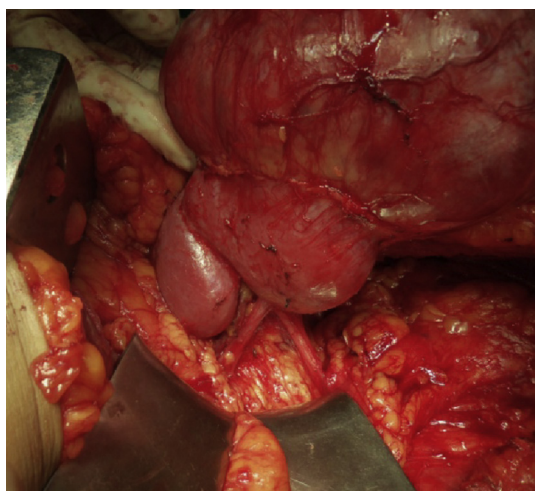


**Figure 1** Computed tomography scan showing a large well defined lesion of the size 9.9 cm × 8.2 cm × 8.1 cm arising from left lower pole of kidney mostly neoplastic in aetiology and normal right kidney.

the possibility of left renal exophytic cystic neoplasm. Contrast enhanced computed tomography (CECT) scan (Fig. 1) showed a large well defined lesion of the size 9.9 cm × 8.2 cm × 8.1 cm arising from left lower pole of kidney mostly neoplastic in aetiology. There was no hydronephrosis or hydroureter on either side. The right kidney was normal. Open left partial nephrectomy was done by left flank approach (Figs. 2–4). Histopathology report revealed variable-sized cysts lined by flattened to cuboidal cells with foci of calcification, inflammatory cells, slings of smooth muscle fibres and tubules with eosinophilic secretions suggesting cystic nephroma. The resection margins were free.

### 3. Discussion

Cystic nephroma comprises of about 1%–2% of all renal tumours [1]. It has a bimodal age distribution with



**Figure 2** Intra-operative image showing the mass arising from lower pole of the left kidney.



**Figure 3** The excised specimen.



**Figure 4** Cut section of the excised specimen.

approximately two-thirds of tumours presenting in childhood, with male predominance and the rest presenting in adults over 30 years of age, with a peak in the fifth and sixth decades of life [3] and female preponderance [1]. Only 5% of this tumour is seen in the age group of 5–30 years [1]. This tumour is not associated with cysts in other organs and is less commonly associated with other congenital anomalies [3].

The aetiology has been postulated as both congenital affecting children as well as acquired affecting post-menopausal females [4]. It has also been proposed to be of neoplastic origin arising from the ureteral bud [1]. Cystic nephroma has now been classified with cystic partially differentiated nephroma (CPDN) as a multilocular cystic renal tumour [3]. Although histologically distinct, they are anatomically and radiologically identical [3]. In 1989, a modified terminology was proposed by Joshi and Beckwith [5] to differentiate cystic nephroma from CPDN and other cystic renal tumours like Wilms' tumour with cyst formation [3]. Steele et al. [6] has observed similarities of the tumours' stroma with ovarian stroma suggesting possible entrapment of Mullerian type tissue into the kidney.

The presentation is usually as a painless abdominal mass in children and with nonspecific symptoms of flank pain,

haematuria, urinary tract infection and hypertension in females [1]. Recently, childhood cystic nephroma has been associated with a germline mutation in the *DICER1* gene [7] and also with risk of developing pleuropulmonary blastoma. Hence, appropriate genetic counselling and testing of *DICER1* mutation may be required. Also, screening should be done for the presence of pulmonary cystic or solid lesions [7]. Similarly, children diagnosed with pleuropulmonary blastoma or with *DICER1* mutation should also be screened with regular renal ultrasounds [7].

Imaging modalities like CECT scan and USG aid in diagnosis and help to differentiate cystic nephroma from other cystic renal masses like Wilms' tumour with cyst formation, clear cell sarcoma, cystic variants of mesoblastic nephroma and cystic renal cell carcinoma [3]. The presence of solid elements in the lesions excludes the diagnosis of cystic nephroma and points towards aggressive neoplasms [3]. Though these imaging modalities demonstrate the multilocular character of the neoplasm, distinct between class II and III cyst using the Bosniak classification is difficult [1]. On CT scan, cystic nephroma appears well-circumscribed, multiseptated cystic lesions with discernible enhancement and a propensity to herniate into the renal pelvis or proximal ureter [1]. Calcification, though rare, can occur at the edge of the herniating portion of the pelvis [3]. Magnetic resonance imaging and renal scintigraphy play a less important role in the diagnosis.

Surgery offers a definitive cure. Multilocularity, absence of communication between the cyst and the renal tissue, cysts filled with clear fluid and no communication between the locules are the chief characteristics of cystic nephroma [1]. Necrosis and haemorrhage are less common. Histologically, cystic nephroma is characterized by the presence of variable-sized cysts lined by flattened to cuboidal cells, foci of calcification, inflammatory cells, ovarian-like stroma, corpora albicans-like structures, slings of smooth muscle fibres and tubules with eosinophilic secretions, as stated in the extensive study by Eble and Bonsib [8]. Though recurrences have been reported, malignant transformation is rare. In childhood, bilaterality and association with *DICER1* mutation have been associated with pulmonary malignancy and should be suspected and managed accordingly [9].

## 4. Conclusion

Cystic nephroma are rare benign renal masses of unknown aetiology and present with nonspecific symptoms. Imaging modalities aid in diagnosis; however, the definite diagnosis is made by histology. Malignant transformation is rare.

## Conflicts of interest

The authors declare no conflict of interest.

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