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# Multilocular Cystic Nephroma: A Case Report and Review of the Literature

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## Abstract

Cystic nephroma is a relative rare benign renal lesion with a bimodal age distribution. Diagnostic peaks occur primarily in the first 2 to 3 years of life, and again in the forty to fifty years. They are usually incidentally found as an asymptomatic abdominal mass. Radiologically, it is difficult to differentiate between cystic nephroma and cystic RCC in adults. The exact diagnosis primarily depends on the histopathologic examination. We present a 28-year-old male patient with a symptomatic, incidentally found left complicated renal cyst. A laparoscopic partial nephrectomy was performed on this patient. Microscopically, the tumor composed of variable-sized cysts lined by a layer of flattened or cuboidal cells that confirmed a diagnosis of cystic nephroma. The literatures were reviewed.

**Keywords:** Cystic nephroma; Benign renal tumor; Multilocular

## Introduction

Cystic nephroma is a rare benign renal tumor and usually present as a multiloculated cystic neoplastic lesion. The age of diagnostic peaks occur primarily in the fourth and fifth decades with a significant (8:1) female prevalence [1]. It is difficult to differentiate between cystic nephroma and other renal tumors, especially cystic renal cell carcinoma (RCC) radiologically. The classical symptoms are palpable abdominal mass, abdominal pain, and gross hematuria, such as other kinds of RCC. However, most of the patients with cystic nephroma are asymptomatic and usually found incidentally. Herein, we present a case of cystic nephroma presenting as a complicated renal cyst. The clinical presentation, radiological findings, and histopathologic results were reported and the literature was reviewed.

## Case Report

A 28-year-old male suffered from abdominal discomfort for one year. There were no vomiting, diarrhea, fever, or gross hematuria. An ultrasonographic examination revealed one 3.8 cm cyst with calcification in November 2015. A computed tomography (CT) was arranged and a multilocular, and hypodense mass arising from the upper and middle pole of the left kidney was found. Thickening of the cyst wall was also noted (Figure 1). A Bosniak class III renal cyst was impressed and laparoscopic left partial nephrectomy was performed for this patient. No postoperative complication was encountered and the patient was discharged uneventfully 4 days after the surgical procedure.

The specimen was measured 2.6 × 2.5 × 2.0 cm in size (Figure 2). The cut surface revealed variable-sized cysts with focal wall thickening with obvious tumorous tissue or necrosis (Figure 3). The resection margin measures 0.4 cm away from the tumor.

Microscopically, the tumor was composed of variable-sized cysts separated by fibrous septa with matured tubules under 40x zoom H&E stain (Figure 4). The wall of the cysts was lined with a layer of flattened or cuboidal cells under 400x zoom H&E stain (Figure 5). A multilocular cystic nephroma was diagnosed. The margin between renal parenchyma (above) and cystic nephroma (bottom) was shown histologically (Figure 6).

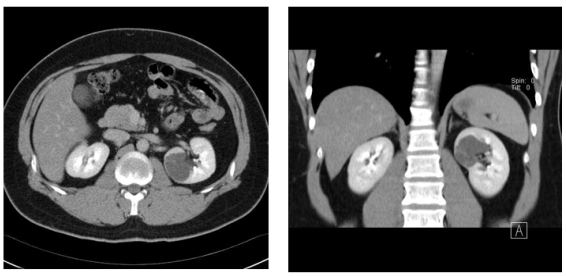

**Figure 1:** Computed tomography with contrast enhanced.

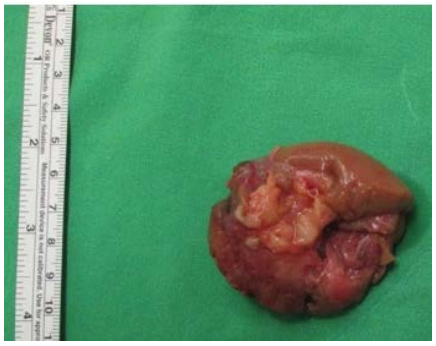

**Figure 2:** Specimen, origin.

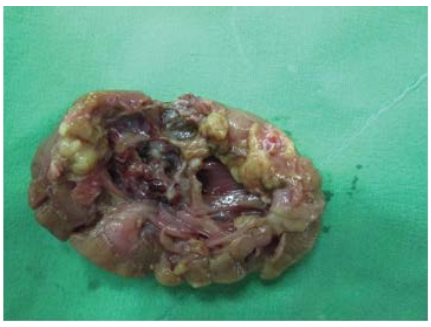

**Figure 3:** Specimen, cut.

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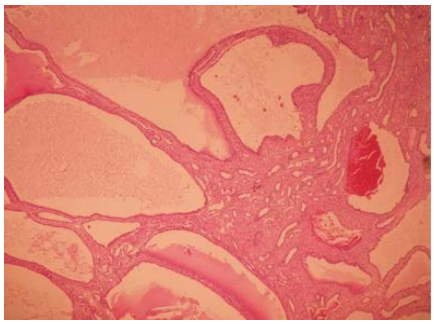

**Figure 4:** Tubules in fibrous septa, (H&E 40x), H&E=Hematoxylin and Eosin.

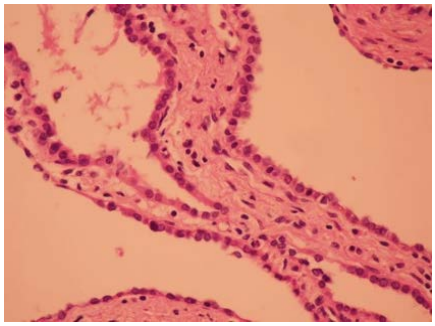

**Figure 5:** Cuboidal cells lining, (H&E 400x), H&E=Hematoxylin and Eosin.

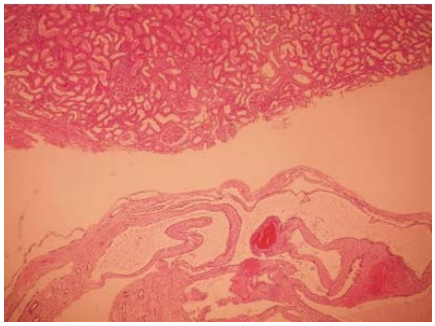

**Figure 6:** Cystic nephroma (below) and margin of renal parenchyma (above).

## Discussion

Cystic nephroma was first reported by Edmunds in 1892 who described the lesion as a "cystic adenoma of the kidney" [2]. Cystic nephroma has a special bimodal age distribution, one in childhood of 2 to 3 years of life, predominantly in boys, and the other in the 40 to 50 years of life. The disease is predominantly found in adult female with an 8-time prevalence compared to male.

Interestingly, this phenomenon is not observed in younger patients [1]. The prevalence is still difficult to determine due to rarity. The diagnostic criteria of histology were modified by Joshi and Beckwith in 1989 [3]. Microscopically, the cysts are formed by a layer of flattened or cuboidal cells. The septa are composed of fibrous tissue with or without mature tubules. The residual renal tissue should be essentially normal without malignant potential.

Similar to other renal tumors, classical signs can include palpable abdominal mass, abdominal pain, and gross hematuria. However, most of cystic nephroma were found incidentally and asymptomatically [4]. Several familial cystic nephroma have been reported linked to sarcoma and clear cell carcinoma with limited evidence [5].

Usual ultrasonographic findings are multiple hypoechoic spaces separated by thin septa. CT scan, on the other hand, usually reveals a multilocular cystic tumor with curvilinear calcifications. Herniation into the renal collecting system with septal enhancement is commonly seen.

Occasionally, all or part of the tumor may appear solid or soft tissue-like because of the aggregation of several small-sized cysts [6]. Unfortunately, it is pretty difficult to differentiate between cystic nephroma, cystic RCC in adults and Wilms tumor in children radiologically even on CT scan [7].

Grossly, cystic nephroma is a well-circumscribed tumor with a smooth surface. The cut surface reveals variable-sized cysts separated by thin septa. The cysts contain mostly clear to yellowish fluid but occasionally dark-colored fluid because of blood clots retention. They may be herniated into the renal collecting system but do not communicate with renal pelvis. That is why only parts of patients have hematuria microscopically or macroscopically. Bilateral cystic nephroma and recurrence after excision was rarely reported. Only several case reports in childhood co-existing with nephroblastoma [8].

The differentiation between benign renal cysts and cystic RCC remains difficult in image or histology. When a complicated renal cyst is identified, it is crucial to evaluate the cystic wall, such as thickness, calcifications and the density of fluid. Bosniak developed a useful classification system for identifying these cystic lesions [9]. Category III lesions hardly differed from malignant neoplasms. However, Category IV lesions are almost certain as cystic RCCs that require surgical intervention.

Traditionally, treatment for any solid renal mass or multilocular cystic lesion was nephrectomy. Nowadays, partial nephrectomy has become the initial standard option for small renal mass [10]. If the lesion is localized enough, excision of the lesion or partial nephrectomy can be considered, even for large tumor in children [11].

## Conclusion

In conclusion, cystic nephroma is a rare, benign renal tumor, and it is difficult to differentiate from cystic RCC. Neither clinical signs nor radiological findings can obtain a pre-operative diagnosis of cystic adenoma. Surgical intervention and histopathologic examination are necessary for the final diagnosis. Noninvasively radiological follow-up is recommended after complete resection.

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