



## Cystic Nephroma – An Uncommon Renal Neoplasm: A Case Report

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### Abstract

*Cystic Nephroma is an uncommon neoplasm. It is classified as a part of the mixed epithelial and stromal tumor (MEST) family under the WHO 2016 classification.<sup>1</sup> Here we report a case of cystic nephroma in a 22 year old female who presented with symptoms of flank pain, hematuria and history of recurrent urinary tract infection (UTI).*

**Keywords:** *Cystic nephroma MEST.*

### Introduction

Cystic nephroma is an uncommon benign cystic lesion of the kidney. It was first described by Edmunds in the year 1982 and was termed as cystic adenoma.<sup>2</sup> So far, nearly 200 cases have been reported in literature. The name multicystic nephroma was first proposed in 1951. It was further modified and subcategorized into cystic nephroma and cystic partially differentiated nephroma based on the absence or presence of blastemal component.<sup>3</sup>

In children, males are more commonly affected than females with the male female ratio being 3:1. In adults, the male female ratio is 1:8 showing a female preponderance.<sup>4-7</sup>

Gross similarities with other cystic tumors of the kidney may cause diagnostic difficulty and conflict in the treatment of this lesion.<sup>1</sup>

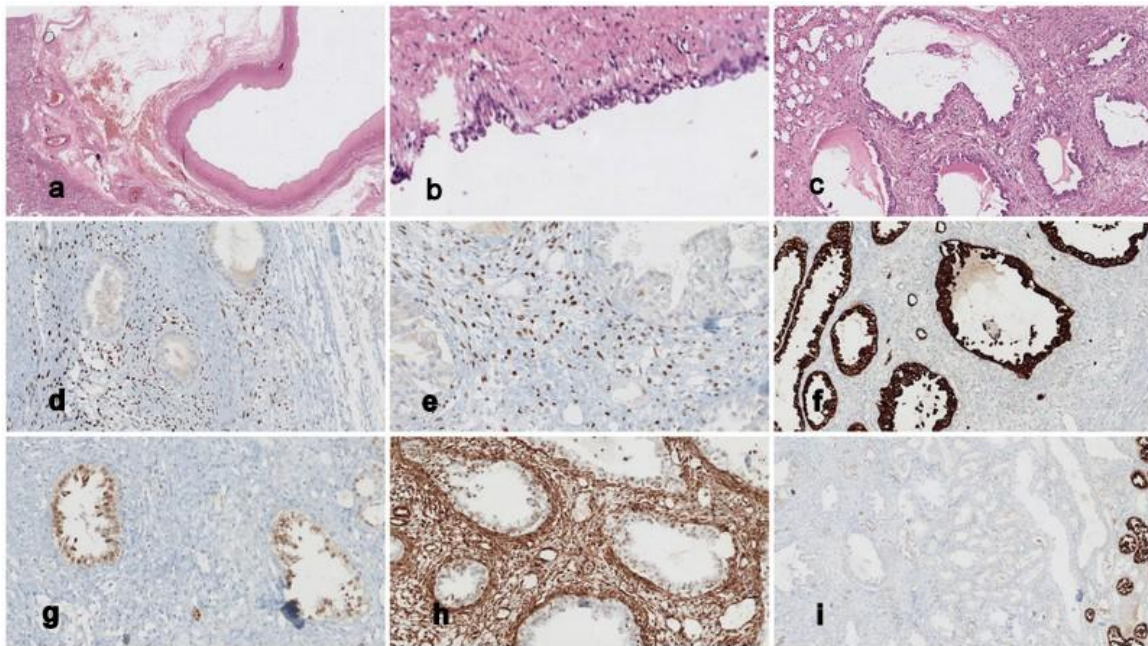
### Case Report

A 22 year old female presented with persistent right flank pain and hematuria. There was a

history of recurrent UTI. Ultrasonography revealed a multicystic mass involving almost the entire renal parenchyma having multiple thick septations within the mass. Radical nephrectomy was carried out. On gross examination, the external surface was unremarkable. Cut surface revealed multiple non communicating cysts filled with serosanguineous fluid. Microscopic examination revealed multiple cysts lined by cuboidal or flattened epithelium and separated by fibroblastic stroma. Immunohistochemical findings revealed that the epithelial cells lining the cyst wall were positive for cytokeratin and PAX 8 whereas the stromal component was positive for Estrogen receptor (ER) and Progesterone receptor (PR). The tumor showed a strong positivity for Smooth muscle actin (SMA). HMB-45 and CD 10 showed negative staining which ruled out the possibility of angiomyolipoma and cystic renal cell carcinoma respectively. Based on the above findings, the final histopathologic diagnosis was cystic nephroma.



**Figure 1:** Gross photograph of nephrectomy specimen showing multiple non communicating cysts filled with serosanguineous fluid.



**Figure 2:**

- Photomicrograph showing cyst wall lined by cuboidal epithelium ( HE X 100X)
- Photomicrograph of the cyst wall lining showing hobnailing ( HE X 400X)
- Photomicrograph showing several small cysts ( HE X 100X)
- Photomicrograph showing nuclear ER positivity of stromal cells (HE X 100 X)
- Photomicrograph showing nuclear PR positivity of stromal cells (HE X 400 X)
- Photomicrograph showing positive staining for cytokeratin in the epithelial component (HE X 100 X)
- Photomicrograph showing negative staining for CD 10. Adjacent normal renal tubules show positive staining (HE X 40 X)
- Photomicrograph showing nuclear PAX 8 positivity in epithelial component (HE X 100 X)
- Photomicrograph showing SMA positivity (HE X 100 X)

## Discussion

Cystic nephroma is a relatively uncommon renal neoplasm with a bimodal age distribution. Approximately two thirds of cases present in children, with male preponderance. In adults, the cases are seen after 30 years of age with a peak in fifth and sixth decade and female preponderance.<sup>8</sup>

In the present case, the patient was a 22 year old female. As per previously published reports, only 5% of cases are seen in the age group of 5 - 30 years which is another interesting feature of the case discussed. The exact etiopathogenesis of this entity is unclear. The etiology has been postulated as both congenital affecting children as well as acquired affecting post menopausal females.<sup>9</sup> It has also been postulated that it could be neoplastic in origin, possibly arising from the ureteral bud.<sup>10</sup>

The pathogenesis of these tumors seem to be based on the influence of hormones since they mostly affect females in the adult age group. This is supported by the presence of estrogen receptor (ER) and progesterone receptor (PR) positivity in the stromal cells.<sup>11</sup> In the present case too, we found ER and PR nuclear positivity in the stromal cells. Also, several investigators have reported the affinity of cyst epithelium for cytokeratins, suggesting an aberrant renal tubular differentiation.<sup>12</sup> In our case too, we found that the cyst epithelium was strongly positive for cytokeratin and PAX 8.

Histologic criteria of cystic nephroma were described in 1989 by Joshi and Beckwith.<sup>13</sup> Accordingly, diagnostic criteria were redefined as follows:

- a) Presence of multiple cysts separated by fibrous septa
- b) Well circumscribed mass distinct from renal parenchyma
- c) Absence of solid component
- d) Presence of cuboidal to flattened epithelium with hob nailing
- e) Septae formed by well differentiated renal tubular or fibrous tissue.

The differential diagnosis include cystic partially differentiated nephroblastoma, multicystic

dysplastic kidney, renal cell carcinoma with necrosis and haemorrhage. Definitive distinction between these entities is very difficult by radiological means. Also, patients usually present with non-specific symptoms like flank pain, hematuria and symptoms of UTI. Therefore, nephrectomy is the treatment of choice and histopathology supported with immunohistochemistry is the ultimate modality for a definitive diagnosis.<sup>14</sup>

## Conclusion

Cystic nephroma is an uncommon lesion and should be considered in the differential diagnosis of malignant renal cysts in both children and adults. Our case and review of literature emphasizes on the importance of IHC to support the histopathologic diagnosis of CN. Further, it highlights the influence of hormones in the pathogenesis of the tumor.

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