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# A Rare Case of Renal Cell Carcinoma in a 4 Year Old Child: The Case Report

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***Abstract***
*Renal cell carcinoma is infrequent in pediatric age group. Children with RCC are generally older than those with Wilms tumor. Here we report a 4 year old male patient who came to our outpatient department with chief complaints of hematuria of 4 months duration. Patient had no co-morbidities and clinical examination was normal. USG abdomen and pelvis, CECT abdomen was done and patient was found to have left renal mass. Based on clinical and radiological findings, patient was suspected to have Wilms' tumor. Patient underwent left nephroureterectomy. Intra-operatively tumor of size 3*3cm was noted in the lower pole of left kidney with few enlarged pelvic lymph nodes. Left nephroureterectomy along with pelvic lymph node dissection was done. Pathologically the resected tumor was found to be renal cell carcinoma (stage 1). Hence the patient was not offered any adjuvant therapy.*

***Keywords:*** *Renal cell carcinoma, nephroureterectomy, Wilms' tumor, children*

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## I. Introductions

Renal cell carcinoma (RCC) encompasses a heterogeneous group of cancers derived from renal tubular epithelial cells.Renal cell carcinoma typically presents in the sixth to eighth decade of life with the mean age of presentation being 64 years.RCC is exceedingly rare in the first decade with Wilms' tumor( nephroblstoma) being the most common renal tumor in this age group.(12)

Here, we report a 4 year old male child came to our OPD with chief complaints of hematuria for 4 months duration which was intermittent and progressive in nature. There were no other complaints like fever, mass/ pain abdomen.

## II. Case Report

Examination findings

4 year old male child, presented with the above complaints to the OPD. Patient is conscious, well oriented. Vitals include pulse of 100 bpm, normal in rate, rhythm and volume. BP 90/60 mm of Hg in right brachial artery, supine position. So2 -99% on RA. RR- 28 cpm. General physical examination- no pallor, icterus, cyanosis, generalized lymphadenopathy, clubbing or pedal edema, no neurocutaneous markers noted and no bruit noted over the carotids and abdomen.

Systemic examination

CVS- S1 S2 heard, no added sounds, no murmurs. JVP- normal, no signs of pulmonary arterial hypertension. RS- Trachea central, apex beat palpable( normal), bilateral chest movements symmetrical, bilateral air entry +, no added sounds. PA- soft, non tender, no organomegaly, no bruit. CNS: HMF- normal. No sensorimotor deficits. No signs of meningeal irritation noted.

Radiological evaluation

USG abdomen with renal doppler-

ill defined hyperechoic mass lesion with necrotic components noted in the lower pole of left kidney extending to renal pelvis causing moderate hydronephrosis. Doppler study- No evidence of renal vein/ IVC thrombosis.Probability of WILMS TUMOR of left kidney with moderate hydronephrosis.

CECT Abdomen (KUB)-

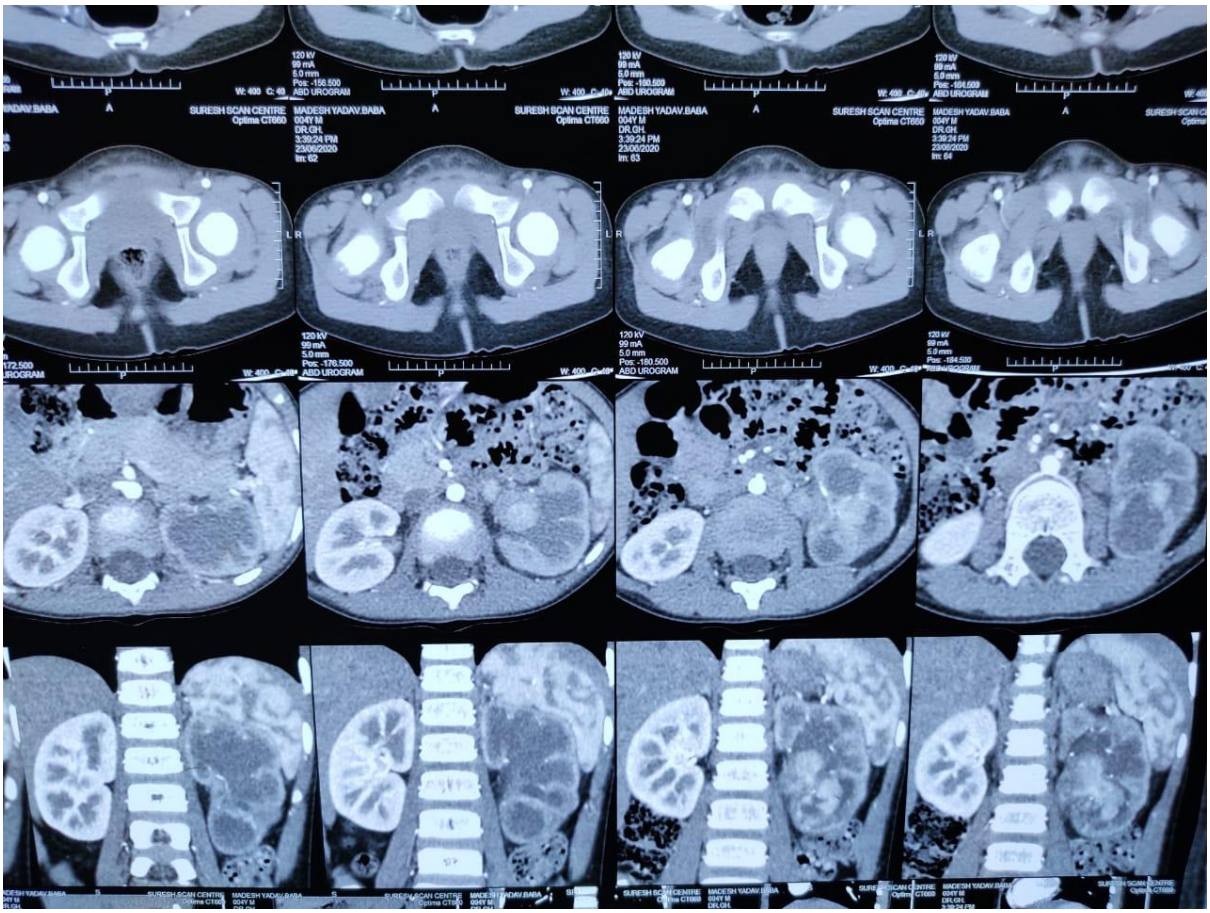

Fig 1: CECT Abdomen and pelvis showing left renal mass

A fairly large moderately enhancing soft-tissue mass ( 3.2* 2.2 cm) noted in the left pelvi-calyceal system, in its middle and lower calyceal groups, occluding the pelvic-ureteric junction with resultant moderate left hydronephrosis, resultant enlarged kidney,thinned-out parenchyma and poorly functioning left kidney , well appreciated on Cturogram. This lesion is confined to the lumen with no extra- luminal infiltration. Discrete lymph nodes are seen in the left renal hilum and para-aortic region. There is no liver metastasis/ ascites or bilateral pleural effusion. Right kidney is normal. Exact etiology is uncertain. Wilms tumor cannot be ruled out.

Blood investigations

Relevant blood investigations were sent

| **CBC** | | **URINE examination** | |
|---|---|---|---|
| **TLC ( cells/ dL)** | 7100 | Sugar | NIL |
| **Hb (g/ dL)** | 9.6 | Albumin | NIL |
| **Platelet count(cells/ dL)** | 237000 | Deposits | 5-6 pus cells |
| **ESR** | 28 | | |
| **RFT** | | | |
| **Urea** | 32 | | |
| **Creatinine** | 0.7 | | |

MANAGEMENT

Based on the clinical examination, radiological findings and considering the age , patient was tentatively diagnosed as Wilms tumor involving the left kidney and hence planned for left nephroureterectomy.

Intraoperatively tumor of size 3* 3 cm was found to arise from the lower calyx of the left kidney. Few enlarged left peri renal lymph nodes were noted. Left nephroureterectomy was performed along with perirenal lymphadenectomy and the specimen was sent for histopathological examination

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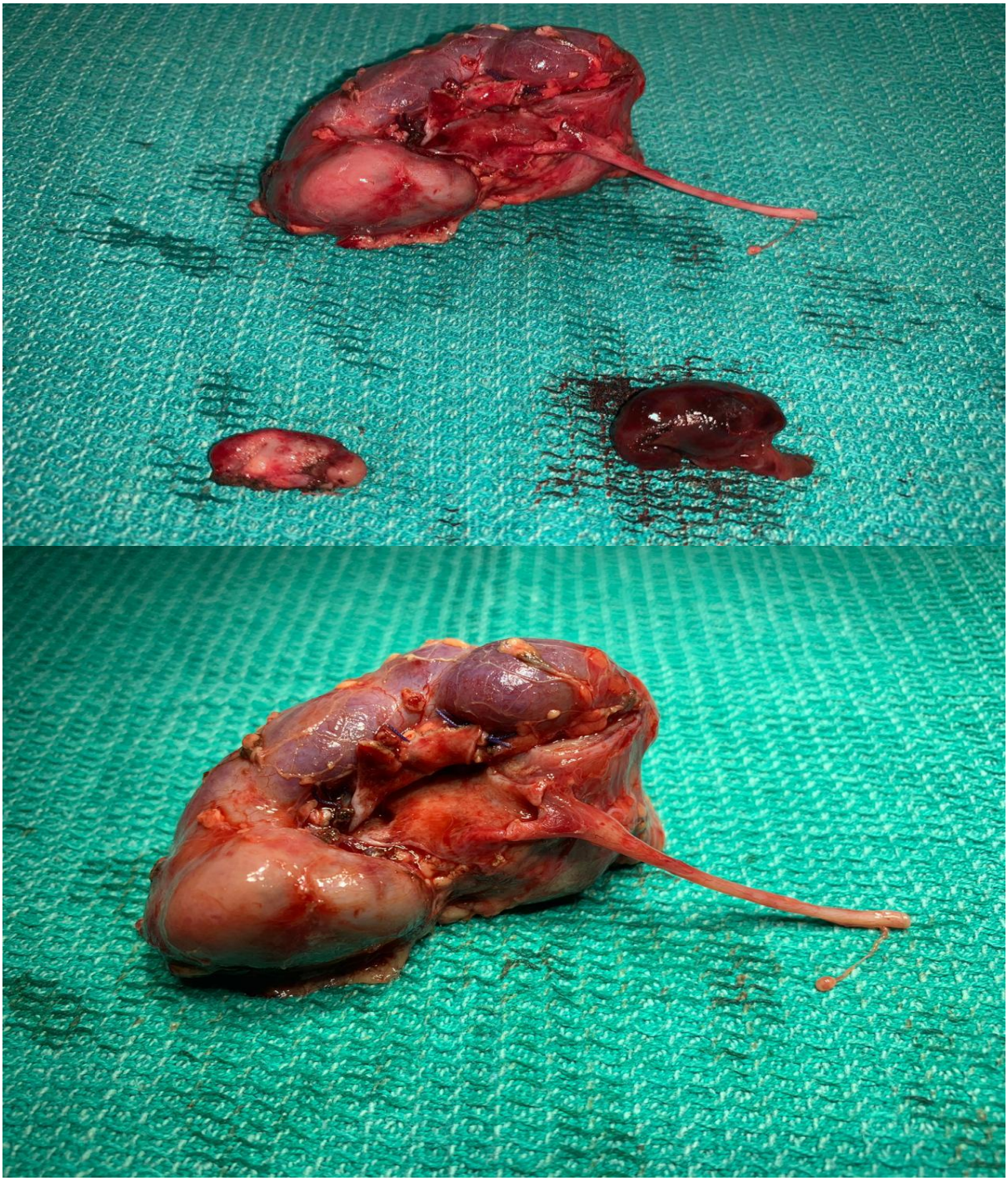

Figure 2 and 3: Resected specimen ( gross)

Histopathological findings

Microscopic examination of the resected specimen shows neoplasm composed of clear cells arranged in lobules sorrounded by thin blood vessels with focal areas of cholestrol cleft of calcification- suggestive of RENAL CELL CARCINOMA ( Clear Cell variant).

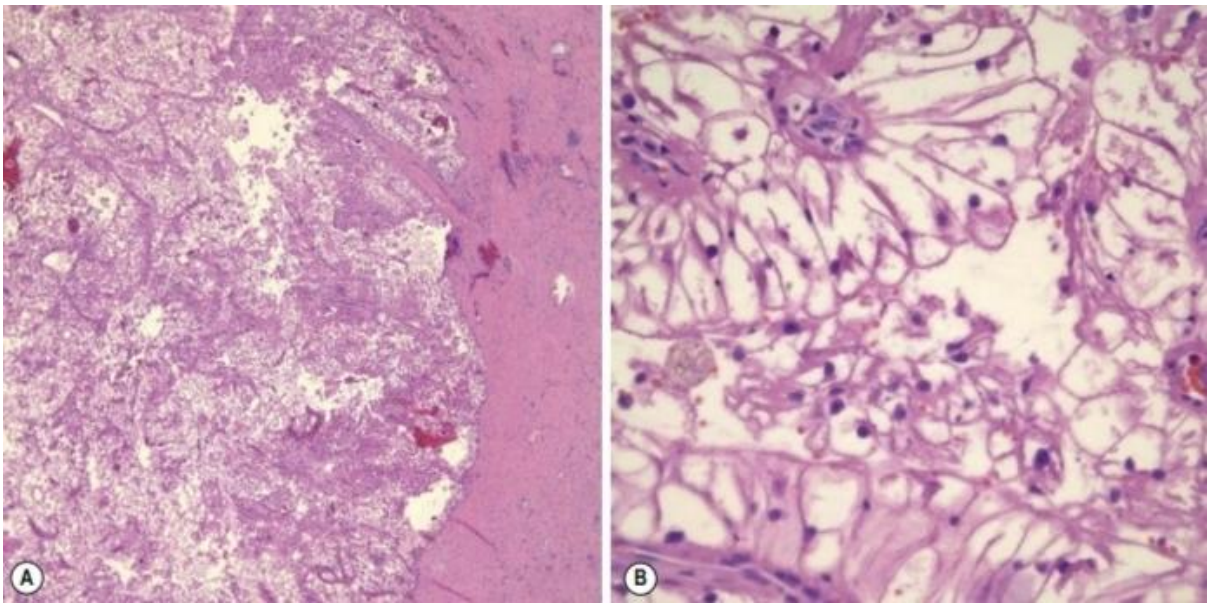


Figure 4a &4b: Microscopic findings of the resected tumor showing clear cells sorrounded by blood vessels

| | |
|---|---|
| **HISTOLOGICAL TYPE** | **CLEAR CELL RCC** |
| **SARCOMATOID/ RHABDOID FEATURES** | NIL |
| **HISTOLOGICAL GRADE** | GRADE 2 (FUHRMAN'S) |
| **NECROSIS** | NIL |
| **LYMHOVASCULAR INVASION** | NIL |
| **FOCALITY** | UNIFOCAL |
| **MARGINS** | FREE OF TUMOR |
| **REGIONAL LYMPH NODE** | FREE OF TUMOR |
| **PATHOLOGICAL STAGE** | Pt1n0 |

Post operative management

Post operative period was uneventful.Lymph nodes were negative, indicating that patient had Stage 1 RCC requiring no further therapy.The boy recovered well during the follow-up period of 18 months.

## III. Discussion

Renal cell carcinoma represents 2% of malignant tumors in adults and is the third most frequent tumor of the urinary tract after bladder and prostate tumors.On the other hand, in the pediatric age group, only 2-3% of malignant renal tumors are proven to be RCC.(2,3). Children with RCC are generally older than those with Wilms tumor (13). According to the survey of Japanese Survey of Paediatric Surgeons, RCC accounted for 1.4% of all renal tumors in children younger than 4 years, 15.2% in children aged 5 to 9 years, and 52.6% in children aged 10 to 15 years (4). Whereas the peak incidence in Wilms tumor occurs around 3 years of age, RCC presents between 9 and 15 years of age.(5,6).

Generally , there is no sex predilection for RCC in children unlike adults, in whom the tumor is predominant among males. The most common form of presentation of RCC in children is macroscopic hematuria or abdominal/ flank pain. Other infrequent symptoms include abdominal mass, anaemia and fever (2,5). It is generally difficult to differentiate it from Wilms' tumor before surgery in pediatric age group. RCC is diagnosed as a solid hyperdense lesion on CT/ USG. Calcification on CT has been reported in 14-28% of RCC cases in children (6,7).

RCC is a malignant tumor arising from the epithelial cells of renal tubules. The 5 subtypes of RCC include- clear cell, papillary, chromophobe, collecting duct type and unclassified variety. Clinical stage at the time of diagnosis is the most important prognostic factor, and the identification of renal vascular invasion doesnot appear to be an adverse predictor. Radical nephrectomy and regional lymphadenectomy have been the primary modality for cure(13).Geller and Dome reported that survival was 60% in children with complete

| STAGE | TNM | SURVIVAL |
|---|---|---|
| I. | T1N0M0 | 92.5% |
| I. | T2N0M0 | 84.5% |
| I. | T1 or T2N1M0<br>T3N0 or N1M0 | 72.7% |
| V. | T4Any NMO<br>Any T Any N M1 | 12.7% |

RCC is remarkably resistant to chemotherapy , preventing cure in most children with metastatic disease.Trials of immunomodulating theraoy with Interferon Alpha and Interleukin-2 have demonstrated efficacy in some studies, but maintainance of a durable cure has been elusive(13). Most recurrences and deaths usually occur within the first 2 years after diagnosis, although late recurrences are frequent. So a strict lon-term follow up is mandatory.

## V. Conclusion

RCC, being exceedingly rare among children in the first decade of life, prompt diagnosis is of utmost importance since its management and therapeutic approach differs from that of Wilms' tumor. Radical nephrectomy along with regional lymohadenectomy remains the gold standard treatment for RCC in this age group. The disease appears to have a less aggressive behaviour in children.The main prognostic factors seem to be staging and complete resection.

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