

CASE REPORT**RENAL CELL CARCINOMA IN CHILDREN: A RARE ENTITY****Jawad Akhtar Gilani, Muhammad Iqbal, M. Rauf Khattak, Safoora Shahid**

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We present a 15-years old young child primarily presented to general surgeon for abdominal mass and gross haematuria. His detailed imaging studies and other relevant investigations revealed that it is a case of Paediatric Renal Cell carcinoma (RCC), which was clinically not associated with any of the known malignancies or syndromes. On detailed histo-pathological examination this was reported as a special type of Pediatric RCC not fitting into clear cell, papillary, chromophobic, oncocytoma or collecting duct RCC.

Keywords: RCC, VHL, IHC

INTRODUCTION

Renal Cell carcinoma (RCC) develops predominantly in individuals with a mean age of 62 years¹ RCC is not a common malignancy in children.² This rare childhood tumor constituting fewer than 0.3% of all tumors and 2–6% of renal neoplasm in childhood and adolescent younger than 15 years.³ RCC is not a single entity but rather a group of tumors originating from kidney epithelium.⁴ There are many types of RCC and the most common type, accounting for 75% of all RCC cases, is clear cell RCC (ccRCC)^{5,6}. The others are papillary (10–15%)^{5,7}, chromophobe (3–5%)^{5,7}, oncocytoma (5–9%)^{5,6} and collecting duct (1%) RCC^{6,7}. Approx 4% of all the RCC are hereditary.⁸ The clinical and pathological features of pediatric RCC are different from those seen in older adolescent and young adults⁹ and it exhibits fundamental differences in biology and behaviour from its adult analogue¹⁰. RCC is universal in children except in association with von Hippel-Lindau (VHL) Syndrome and typically occurs in the 2nd decade.

CASE REPORT

We report a young boy, 15 years old, class VII student, who presented to a General Surgeon with abdominal mass and haematuria of short duration. Abdominal imaging studies revealed upper quadrant mass measuring 3×3×3 Cm related to right kidney. Further diagnostic work-up and histopathology of mass unveiled the diagnosis of RCC localised and early stage disease, i.e., (T_{1a}N₀M₀G₃). The patient was referred to IRNUM for further evaluation and management.

DMSA scan of kidneys was performed at IRNUM that was reported as having reduced cortical tracer uptake by right kidney with central photon deficient area corresponding to the dilated collecting system. The ultrasound examination gave opinion of reduced functioning cortex of right kidney. Post nephrectomy specimen was reported as hemorrhagic tumor nodule in renal pelvis measuring 3×2.5×2.5 Cm³, limited to the kidney. Only Immunohistochemistry (IHC) CD-10 was performed and reported as positive.

Other confirmatory IHC stains are not locally available yet.

Extensive literature review regarding such cases revealed that there occurs translocation in Xp11.2 region involving TFE-3 gene in a good proportion of paediatric RCC, such patients present special subset of papillary RCC. The same was requested by histopathologist which is unfortunately not available in Pakistan yet.

After detailed evaluation of this patient clinically, radiologically, and ophthalmologically, we could not find any pathology either in opposite kidney, adrenal, pancreas, eye and brain or any association with VHL disease, Birtt-Hogg-Syndrome and retinocytoma.

As our patient has an early stage RCC, this young boy needs close follow-up, therefore he will be closely monitored for the issues regarding recurrence, distant metastasis or second tumour in the opposite kidney.

DISCUSSION

This case was discussed in detail with histopathologist and the consensus was that, this special entity of paediatric RCC is not compatible with commonly known types and it does not fall into classical classification of clear cell, chromophobe, papillary or collecting duct. On bi-halving the kidney, the morphological appearance was polyp-like structure with stalk attached to pelvis of right kidney.

The diagnosis of paediatric RCC in this case is based exclusively on histopathology, clinical and special features, because special Immunohistochemistry (IHC) and cytogenetic analysis, which are performed on freshly resected tumour specimen of such cases, are not yet available in Pakistan.

Surgical intervention is similar to adult RCC, though the role of lymph nodes dissection is unclear yet. There are no adequately powered studies to support conclusions about adjuvant or neoadjuvant chemotherapy for children with RCC.¹¹ Similarly there is no clear-cut consensus on the role of targeted therapy as for adults. Role of radiotherapy is debatable in early

stage. However, it can be useful for distant metastasis and pain control.

Scrutiny of last 16-years (1994–2009) paediatric tumour data of IRNUM was performed in detail. Total cases of paediatric tumours were 8,469, out of which paediatric renal tumours were 419 (4.94%), of which 408 (4.8%) were Wilm's tumours and 11 (0.13%) were RCC patients. Majority of these RCC patients showed more or less same clinical presentation. This particular case has same clinical presentation but special pathological features which are not compatible with clear cell, chromophobe, papillary or collecting duct carcinoma.

Low frequency of paediatric RCC reflects the rareness of studies in this age group. RCC in young patients is usually localised at the time of diagnosis and at comparable stage was linked to a better prognosis than similar tumours usually seen in older individuals and present as fewer clear cell carcinoma but more papillary carcinoma.¹⁰

Karakiewicz *et al*¹¹ have demonstrated that age at diagnosis appears to be an independent factor of renal cell carcinoma-specific survival in patients with RCC, confirming findings reported by other investigators^{10,12}. Unlike patients with colon and prostatic cancer, young patients with RCC have better prognosis than old patients. Given the favourable survival data for young patients and less role of adjuvant systemic therapy, the importance of nephron-sparing surgery is increased exclusively in T₁ lesion.¹³ Similarly lymph nodes involvement and distant metastasis is lower in young adults compare to older patients.¹⁴ While treating RCC in children it should also be kept in mind that Neuroepithelial Tumours of Kidney (NETK) are also separate entity which has unique proclivity for youngsters. This is also an aggressive tumour which at presentation shows invasion of renal capsule and metastasis. Primary NETK are characterised by CD-99 (MIC-2) and EWS/FLI-1 or small cell carcinoma and having chromogranin positivity.^{15,16}

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