

Renal cell carcinoma in a duplex kidney in pediatric

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Abstract

Renal cell carcinoma (RCC) in children is relatively uncommon, especially in the absence of syndromes or hereditary diseases. Duplex kidney – as a common congenital anomalies in children – is usually insignificant with no serious impact or known risk of tumor association. Herein, we report a 5-year-old girl who presented with gross hematuria and a right renal mass in duplex system identified on computed tomography. Radical nephrectomy was performed; the patient was diagnosed with Xp11 translocation RCC and no other modalities of treatments were needed over 3-month follow-up.

Keywords: Duplex kidney, pediatric, renal cell carcinoma

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INTRODUCTION

Renal cell carcinoma (RCC) in children is relatively uncommon, accounting approximately 2%–12% of all pediatric renal malignancies.^[1] Unlike in adult, where are most of RCC discovered incidentally; in pediatric RCC, many patients are symptomatic at diagnosis, experiencing of abdominal pain, hematuria, and/or abdominal mass.^[2] RCC in children tend to be more aggressive and advanced compared to adults.^[1] Because of the rarity of this disease, surgery remains the main stay of treatment and results in cure when tumor is completely resected; however, for advanced disease requiring systemic treatment, available data are scarce.^[3]

Duplex kidney is a common congenital renal anomaly described in 1 in 125 people (0.8%) and tend to be more common in females.^[4] There is an increase in ureterorenal pathologies including ectopic ureter,

ureteroceles, vesicoureteral reflux (VUR), and ureteropelvic junction obstruction (UPJO) with unknown risk of renal tumor association.

In the following case report, we reported a child with RCC in duplex system. To our knowledge, this is the only case described in the literatures for a child with RCC in a duplex system. However, Mohan *et al.* reported RCC in a duplex system in an adult patient.^[5]


CASE REPORT

A 5-year-old girl presented with few days history of painless gross hematuria with no other urinary tract symptoms. There was no family history of cancer, and on clinical examination, she was completely well with no dysmorphic features. Contrast-enhanced computed tomography (CT) of the chest and abdomen revealed a well-defined solid heterogeneously enhancing right renal mass measured 3

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cm × 2.8 cm × 2.8 cm arised from the lower moiety of a duplex collecting system [Figures 1 and 2] and slightly prominent hypodense lymph nodes in the retrocaval and aortocaval regions with no lungs lesions. Hence, radical nephrectomy was performed [Figure 3]. Histopathological examination revealed Xp11 translocation RCC Grade 3, T3a, and N1. Repeated CT scan 1 and 3 months postsurgery demonstrated no evidence of local recurrence, residual disease, or distal metastasis. The patient did not need any further management beyond surgery, and she is still on regular follow-up.

DISCUSSION

RCC is a rare malignant renal tumor in children and adolescents younger than 15 years. The clinical features of RCC in children are different from those seen in adults; in children, most cases are symptomatic. Sausville *et al.* found that 88% of patients are symptomatic at initial presentation. The symptoms were mainly abdominal pain and/or hematuria in most of cases where abdominal mass, fever, or weight loss were presented in lower incidence. The median age at presentation with RCC was 12.9 years, with roughly equal male to female distribution,^[2] although female showed higher prevalence in some studies.

Akhavan *et al.* demonstrated that the younger age was associated with higher stage, grade, and larger tumors at presentation.^[1] In our case, the young age of the patient of 5 years was associated with a relatively small tumor but invading lymph nodes in the retrocaval and aortocaval regions.

Nearly 10% of all human beings are born with a congenital anomaly of the urogenital system; a duplication of the upper urinary tract occurs in approximately 1 in 125 individuals. A duplex system describes a renal unit where the kidney is composed of two pelvicalyceal systems that are associated with the incomplete, partial, or complete duplication of the ureters.^[4] There is an increase in ureterorenal pathologies including ectopic ureter, ureterocele, VUR, and UPJO; howbeit, the association with renal tumors is unknown.

The development of a tumor within a duplicated collecting system is infrequent. The presence of RCC in a duplex system has been reported in adult.^[5] However, the presence of RCC in such an anomalous kidney was not reported in children.

CONCLUSION

Surgery of RCC in duplex kidney dose not differ from those arising in single system. The underlying pathogenesis of this



Figure 1: Exophytic mass in the right kidney



Figure 2: Well-defined solid heterogeneously enhancing right renal mass measured 3 cm × 2.8 cm × 2.8 cm arised from the lower moiety of a duplex collecting system



Figure 3: Specimen showed the tumor arising from the lower moiety of the right kidney

association is unknown. However, the limitation number of those cases and publications, making the screening for

malignancy in children with duplex system challengeable, therefore, multicenters' collaboration study is required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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