Non-Wilms' Renal Tumors In Children:A 139 Cases Series

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Abstract

Background: Pediatric non-Wilms renal tumors (NWRs) which comprise a small proportion of renal tumors, are a heterogeneous group of neoplasms with variable malignant potential, mortality, and response to treatment. This study aimed to determine the clinical characteristics, management and prognosis of children with non-Wilms' renal tumors (NWRs).

Methods: Medical records of all patients (n = 139) treated for NWRs over a 12-year period (2008.01 – 2019.10) at a single center were reviewed retrospectively.

Results: The histopathological groups of NWRs included malignant rhabdoid tumor of the kidney (MRTK) (n: 30, 21.6%), renal cell cancer (RCC) (n: 26, 18.7%), clear cell sarcoma of the kidney (CCSK) (n: 24, 17.3%), congenital mesoblastic nephroma (CMN) (n: 21, 15.1%), cystic nephroma (CN) (n: 16, 11.5%), metanephric tumours (n: 12, 8.6%), renal angiomyolipoma (RAML) (n: 3, 2.2%), renal primitive neuroectodermal tumor (rPNET) (n: 2, 1.4%), renal hemangioma (n: 2, 1.4%), inflammatory myofibroblastic tumor (IMT) (n: 2, 1.4%), ossifying renal tumor of infancy (ORTI) (n: 1, 0.7%). 123 children were followed up with an average of 42 months. 16 children were lost to follow-up. Tumor-free survival was observed in 94 children. 28 children (22.8%) were died.

Conclusions: Pediatric NWRs comprises 19.1% of all renal tumors in our single center. Accurate diagnoses along with appropriate management are important factors in improving patients outcome. The mainstay treatment of malignant NWRs including MRTK, CCSK, RCC and PNET is comprehensive treatment. The mainstay treatment of benign NWRs including RAML, CN, ORTI, CMN, metanephric tumours, and renal hemangioma is surgical resection alone.

Background

Wilms tumor is the most common renal tumor in children and represents approximately 80%-90% of all pediatric renal tumors. Pediatric non-Wilms renal tumors (NWRs) which comprise a small proportion of renal tumors, are a heterogeneous group of neoplasms with variable malignant potential, mortality, and response to treatment [1]. There is few reports of NWRs in the literature due to their rarity. We retrospectively analyzed the medical records of NWRs in our single center to assess their clinical characteristics, management and prognosis.

Methods

This study was carried out in the department of urology at a comprehensive 3A pediatric hospital in China. Following institutional review board approval, the medical records of children with renal tumors in our hospital between January 2008 and October 2019 were retrospectively analyzed. The patients inclusion criteria required NWRs that were first diagnosed and treated in our hospital and the patients' medical records were complete. Patients that were first treated in other hospitals or had no pathological diagnosis in our hospital were excluded in our study.

Results

Over a 12-year period, a total of 729 cases of pediatric renal tumors were reviewed, of which 590 (81%) were Wilms tumor and 139 (19%) were NWRs. The histopathological groups were malignant rhabdoid tumor of the kidney (MRTK) (n: 30, 21.6%), renal cell cancer (RCC) (n: 26, 18.7%), clear cell sarcoma of the kidney (CCSK) (n: 24, 17.3%), congenital mesoblastic nephroma (CMN) (n: 21, 15.1%), cystic nephroma (CN) (n: 16, 11.5%), metanephric tumours (n: 12, 8.6%), renal angiomyolipoma (RAML) (n: 3, 2.2%), renal primitive neuroectodermal tumor (rPNET) (n: 2, 1.4%), renal hemangioma (n: 2, 1.4%), inflammatory myofibroblastic tumor (IMT) (n: 2, 1.4%), ossifying renal tumor of infancy (ORTI) (n: 1, 0.7%).

The mean age of the 139 children with NWRs was 3.5 years (range: 7 days–15 years and 2 months). Age distribution according to tumor histology are different. 90.5% of those with CMN presented at an age <1 year, all of those MRTK presented at an age <3 years, most of the patients with CCSK presented at 0–3 years of age, 62% of those with RCC presented at an age >7 years and all of those RAML, rPNET presented at an age >7 years. 83 were male and 56 were female. The left kidney was involved in 63 cases and the right kidney in 76 cases.

Clinical presentations included 50 cases with hematuria, 37 cases were detected incidentally by ultrasound examination, 34 cases with abdominal mass, 10 cases with abdominal pain, 4 cases with hematuria and abdominal pain and 4 cases with vomiting and abdominal distension. The most commonly seen presenting symptoms in malignant NWRs such as MRTK, CCSK, RCC and PNET were hematuria, which in 60% of MRTK, 53.8% of RCC and 33.3% of CCSK. The most commonly seen presenting symptoms in benign NWRs were abdominal mass and detected incidentally by ultrasound examination.

Preoperative imaging including ultrasound (US) and computed tomographic (CT) were underwent in all patients. The tumors size were range from 2.0cm × 1.5cm × 1.5cm to 17.7cm × 12.8cm × 19.1cm, with an average of 7.5cm × 6.1cm × 6.9cm. There were 100 cases (71.9%) of solid tumors and 28 cases (20.1%) of cystic tumors. There were 10 cases of tumor rupture in this study, including 5 patients with MRTK, 4 patients with RCC and 1 patient with CMN. There were total 9 cases of tumor thrombus, including 6 cases of renal venous tumor thrombus, 1 case of renal vein and inferior vena cava (IVC) tumor thrombus, and 2 cases of tumor thrombus extension through the renal vein to the right atrium. Image specific signs: 16 patients (53.3%) with typical subcapsular fluid in MRTK.

In most of these cases, NWRs can readily be distinguished using a range of immunohistochemical markers. Molecular genetic profiling has allowed much progress in the understanding of this group of tumours. Among the 26 patients with RCC, 20 (76.9%) were associated with Xp11.2 translocations /TFE3 gene fusion, 3 with papillary carcinoma, 2 with clear cell carcinoma, and 1 with chromophobe cell carcinoma. All the 26 RCC patients underwent TFE3 immunohistochemical assay, and 20 were positive for TFE3. The immunohistochemistry and Fluorescence in situ hybridization (FISH) results of the 30 MRTK cases were that 28 cases were INI-1 negative. All the 24 CCSK cases were positive for vimentin expression and negative for neuron specific enolase (NSE), CD34, CD99 etc. Among 21 cases of CMN,

classical, cellular and mixed subtypes were 7 cases respectively. There were 5 cases of metanephric stromal tumor (MST) and 7 cases of metanephric adenoma (MA) in metanephric tumours.

Distribution of all malignant NWRTs including MRTK, CCSK, RCC and PNET according to stages were as follows: Stages I (n=26), II (n=16), III (n=29), and IV (n=11).

Neoadjuvant chemotherapy was offered to 9 patients, including CCSK (n:4), MRTK (n:2), RCC (n:1), renal hemangioma (n:1) and rPNET (n:1) which were considered for preoperative diagnosis of Wilms' tumor. Nephron-sparing surgery (NSS) was performed in 28 patients. In patients who underwent NSS, the malignant tumor diameter was less than 6 cm in all 8 cases, and the benign tumor diameter was less than 6 cm in 14 of 20 cases. 107 patients were offered radical nephrectomy including 9 patients' tumor thrombus were removed en bloc with the kidney (Figure 1).

123 children (88.5%) were followed up with an average of 42 months (range 13 months to 72 months). 16 children were lost to follow-up. Tumor-free survival was observed in 94 children. 28 children (22.8%) were died. The follow-up of malignant NWRTs including ultrasound and chest X-ray examination every 3 months to postoperative 5 years. Patients with MRTK had the highest mortality rate which only 5 (20.8%) patients survived with an average follow-up of 24 months. The overall tumor-free survival was 73.9% (17/23) for CCSK with an average follow-up of 49 months. 21 of 26 patients with RCC were followed up with an average of 37 months. 19 patients (19/26, 73.9%) survived including 1 patient underwent targeted cancer therapy and 7 patients underwent NSS. For 2 cases of rPNET, one presented with tumor extension through the renal vein to the right atrium died 2.5 years after surgery due to lung metastasis, the other case survived for 4 years till now. So the mortality rate of malignant NWRTs was 40% (28/70).

The follow-up of benign NWRTs including ultrasound examination every six months after surgery. 53 of 57 patients were followed up and no tumor recurrence was found including 20 patients underwent NSS.

Discussion

Non-Wilms' renal tumors (NWRTs) that occur in children include CMN, CCSK, RCC, MRTK, lymphoma, angiomyolipoma, teratoma, hemangioma and other more rare entities [2]. Different literatures reported that the proportion of NWRTs in renal tumors were different ranged from 13.6–18.7% [3–4].

The patient's age at presentation, clinical features, and imaging characteristics are often sufficient to presume a diagnosis [5]. NWRTs can occur in children of all ages, but the peak age of different tumor types is different. MRTK predominantly affects young children in general, International Society for Pediatric Oncology (SIOP) reported a median age of 13 months and the National Wilms Tumor Study Group (NWTSG) reported median ages of 10.6 months [6–7]. The median age at presentation in children with RCC is 9 years [8]. The CCSK occurs with a peak incidence at 1–4 years of age [9]. CMN is the most frequent renal neoplasm of newborns and young infants, which the median age at diagnosis is 2 months [10]. CN is an uncommon benign renal lesion that occurs most commonly in children younger than

24 months of age. RAML and rPNET predominantly affects young adults in general. ORTI occurs most commonly in infancy.

Different NWRs have distinct presentations. Most NWRs present with non-specific features of an abdominal mass, abdominal pain, haematuria and detecting incidentally by ultrasound[11]. In our group, hematuria was found in 50 cases (36.0%), asymptomatic ultrasound in 37 cases (26.6%), abdominal mass in 34 cases (24.5%) and abdominal pain in 10 cases (7.2%).

Preoperative imaging is often sufficient to presume a diagnosis which can be performed to determine the anatomic location and extent of the mass[12]. CCSK is generally unilateral and unicentric with solid and occasionally cystic areas. Many CMN cases are diagnosed on prenatal US and can give rise to polyhydramnios, hydrops, and premature delivery. CN is well-encapsulated multilocular tumors composed of various cysts with thin septation that compress the normal kidney. RAML can be detected fat content within the mass. ORTI is a well-defined, often calcified mass located in the renal pelvis and calyces. A prominent and eccentric crescent with attenuation of fluid-representing subcapsular renal haemorrhage or fluid can be identified in MRTK. Chest X-ray can help determine whether the tumor has lung metastasis. In our group, subcapsular fluid was seen in 16 cases (53.3%) of MRTK. Tumor thrombus were seen in 9 cases including 3 of CCSK, 3 of MRTK, 1 of RCC and 1 of rPNET.

NWRs can be diagnosed by histologic examination and be distinguished using a range of immunohistochemical markers. MRTK accounts for 2% of pediatric renal tumours and they consist of sheets of cells showing nuclear pleomorphism and characteristic morphologic features of open vesicular nuclei, prominent nucleoli, and scattered hyaline eosinophilic cytoplasmic inclusions. The presence of mutations in the hSNF5/INI1 gene on chromosome 22 is the hallmark of MRTK. It results in a marked reduction in nuclear expression of the gene product which is detectable immunohistochemically[13]. In our group, 28 cases (93.3%) of MRTK were INI-1 negative. RCC accounts for 2–5% of all pediatric renal tumors. RCC associated with Xp11.2 translocations/TFE3 gene fusions is the main pathological type which are characterized by the chromosomal translocations involving the TFE3 gene on Xp11.2217-219 or the TFEB gene on 6p21. The most characteristic pathological manifestation is a papillary structure composed of clear cells, which is rarely seen in adult patients and is often associated with nest-shaped structures composed of tumor cells containing eosinophilic granules. Immunohistochemistry can detect aberrant expression for TFE3 or TFEB and can thus be useful in establishing the diagnosis. Other RCC cell types include papillary renal cell carcinoma, clear cell carcinoma and chromophobe cell carcinoma[14]. In our group of RCC, 20 cases (76.9%) were RCC associated with Xp11.2 translocations/TFE3 gene fusions. CCSK accounts for 3% of renal tumors. CCSK has been described as soft and tan-grey in colour, and is well-delineated macroscopically, being composed of small round and oval cells and stellate and spindle cells that had bland nuclei. It is characteristically composed of a mixture of cord cells and septal cells with an extensive capillary network. But other patterns including myxoid, sclerosing, cellular, epithelioid, palisading, spindle-cell, storiform, and anaplastic patterns are also noted. CCSK is characterized by bone and brain metastases. In our group, all the 24 CCSK cases were positive for vimentin expression. Three patients with CCSK presented with metastases to the brain (n = 1), the lung (n

= 1), and bone (n = 1). CMN is a low malignant potential that may exhibit several subtypes including classical, cellular and mixed. Classical variant is composed of fibroblastic spindle cells arranged in bundles and fascicles that infiltrate into the normal renal parenchyma [15]. CN is well-encapsulated multilocular tumors composed of various sized cysts with thin septations that compress the normal kidney. The identifying feature of CN is that of mature well-differentiated cell types within the septa of the cyst wall. There are no blastemal or embryonal elements [16]. CN has been reported associating with pleuropulmonary blastoma and the DICER1 mutation. This is in contrast to adult CN which lack DICER1 mutations [17]. PNET is a high-grade malignant neoplasm which has a characteristic translocation t(11;22)(q24;q12) results in EWS-FLI fusion gene [18]. PNET is composed of primitive-appearing undifferentiated round cells in diffuse dense cellular sheets or vaguely lobulated pattern. The cytoplasm is indistinct except in those areas where the cells are more mature and the elongated hairlike extensions coalesce to form Homer-wright rosettes. Histologically, metanephric stromal tumour is unencapsulated and composed predominantly of spindle or stellate cells, with hypo- and hypercellular areas giving the tumour a characteristic nodular appearance. Metanephric adenoma is a purely epithelial lesion composed of regular, closely packed tubular structures formed of small uniform ovoid cells with no nuclear atypia or mitoses. RAML is the most common benign solid renal tumor and is almost always associated with the tuberous sclerosis complex [19]. Histologically, RAML is composed of different proportions of mature fat, smooth muscle and thick-walled malformed blood vessels. ORIT has a benign clinical behavior [20]. Grossly, the tumor had a nodular or irregular appearance, often partially calcified and located in the renal pelvis and calyces. Histologically, ORIT is composed of osteoblast-like cells, spindle cells and an osteoid core. IMT is a rare entity that tends to aggressive behavior and local recurrence [21]. It is characterized by proliferation of typical spindle-shaped cells accompanied by inflammatory infiltration of plasma cells, eosinophils, and lymphocytes. Immunohistochemistry is positive for ALK (50%-60%), vimentin (95%-100%), desmin (5%-80%).

For the management of NWRTs, the treatment principle of malignant NWRTs is the need for comprehensive treatment including surgery, chemotherapy, and if necessary, radiotherapy [22]. Preoperative treatment which for these MRTK patients consisted of vincristine and actinomycin D for a period of 4 weeks for stages I-III tumours and vincristine, actinomycin D and doxorubicin (VAD) for stage IV tumours for a period of 6 weeks. However, this did not seem to improve outcome [6]. The extent of radical surgical excision followed by chemotherapy and radiotherapy are important determinants for long-term survival. Recent reports have shown encouraging results with a combination of ifosfamide/carboplatin/etoposide (ICE) alternating with vincristine/doxorubicin/cyclophosphamide (VDC) [23]. Since identifying the MRTK patients with hSNF5/SMARCB1/INI1 gene mutations, urgently exploring targets for development of novel treatment strategies is warranted in future. As RCC is often resistant to chemotherapy and radiotherapy, no studies exist that support the use of adjuvant or neoadjuvant chemotherapy or radiotherapy. So surgical resection is the mainstay of therapy. Cook reported NSS was suitable for children with RCC [24]. Current therapy for CCSK includes a combination of nephrectomy, chemotherapy and radiotherapy [25]. It would possibly enhance cure by enabling early inclusion of doxorubicin in the chemotherapy regimen. Renal PNET is a

rare but highly aggressive neoplasm with poor prognosis. Definite metastases occurring at the time of diagnosis in approximately 25% of patients. Effective treatment methods include a combination of surgery, chemotherapy and radiotherapy. VDC (vincristine, doxorubicin, cyclophosphamide) alternating with using of IE (ifosfamide, VP-16) in combination is currently recommended treatment[26].

The mainstay of treatment in most benign NWRTs is surgery and when the conditions permit NSS can be undertaken. CMN generally follows a benign course even with local spillage. Radical nephrectomy alone is usually sufficient. 95% of patients do not relapse and most of the 5% who do have the cellular variant of this disease[10]. CN is adequately treated by radical nephrectomy alone, with an excellent prognosis and no chemotherapy required. Except when spillage or rupture occur, and local recurrence has been reported. With its benign nature some have advocated nephron sparing surgery in polar lesions. Surgery with radical excision or NSS is considered to be the treatment of metanephric tumours. For RAML, if possible partial nephrectomy rather than total nephrectomy is the preferred surgical management. Angioinfarction of the tumors is also an option. ORIT have a benign clinical behavior. NSS or partial resections of the kidney may be considered. IMT is a very rare benign reactive proliferative lesion. Surgery with radical excision is still considered to be the best treatment, although steroid therapy has been reported to regress IMT[27].

The prognosis of malignant NWRTs is poor. For MRTK, age is an important prognostic indicator as younger patients have worse prognosis compared with older patients[28]. The MRTK in our group was followed up for 24 months on average, and the survival rate was 20.8%. Survival for children with RCC is largely affected by stage of disease at presentation and completeness of resection at radical nephrectomy, with overall survival at around 64-87%. In our group of RCC the survival rate was 73.9%. The NWTS reported that the 5-year relapse-free and overall survival rates for CCSK were 79% and 89%, respectively[29]. With an average follow-up of 49 months, the survival rate of CCSK in our group was 73.9%. The prognosis of benign NWRTs is good. In our group of benign NWRTs, a total of 53 cases including 20 cases underwent NSS were followed up, and no tumor recurrence was found.

Conclusion

Pediatric NWRTs comprises 19.1% of all renal tumors in our single center. The histopathological groups including MRTK, RCC, CCSK, CMN, CN, metanephric tumours, RAML, rPNET, renal hemangioma, IMT and ORTI. NWRTs can occur in children of all ages, but the peak age of different tumor types is different. The treatment principle of malignant NWRTs is the need for comprehensive treatment including surgery, chemotherapy, and if necessary, radiotherapy. The mainstay of treatment in most benign NWRTs is surgery and when the conditions permit NSS can be undertaken which can achieve good results, a good prognosis, long-term survival without tumor.

Abbreviations

non-Wilms' renal tumors=NWRTs;malignant rhabdoid tumor of the kidney=MRTK;renal cell cancer=RCC;clear cell sarcoma of the kidney=CCSK;congenital mesoblastic nephroma=CMN; cystic nephroma=CN;renal angiomyolipoma=RAML;renal primitive neuroectodermal tumor=rPNET; inflammatory myofibroblastic tumor=IMT;ossifying renal tumor of infancy=ORTI; ultrasound=US;International Society for Pediatric Oncology=SIOP; National Wilms Tumor Study Group=NWTSG.

Declarations

Acknowledgements

Not applicable.

Ethics approval and consent to participate

This study was approved by the Institutional Review Board of Beijing Children's Hospital,Capital Medical University(IRB No. 2020–Z-070) with waiver of informed consent.

Consent for publication

Not applicable.

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Competing interests

The authors declare that they have no competing interests.

Author's Contribution

Yi Wei Fang☒Data Collection, Manuscript writing

Hong Cheng Song☒Project development, Data analysis

Ning Sun☒Manuscript editing

Wei Ping Zhang☒Data management

Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

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Figures

Figure 1

<p>rPNET patient presented with tumor extension through the renal vein to the right atrium which removed en bloc with the kidney.</p>