A case of chromophobe renal cell carcinoma in a 12-year-old girl

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Abstract Renal cell carcinoma (RCC) accounts for only 2–6 % of pediatric renal tumors. Chromophobe RCC is very rare in pediatric patients, with only a few cases reported. A 12-year-old girl who had abdominal and left flank pain visited the pediatric clinic of a local hospital. A uniformly enhanced tumor measuring 2.5 cm × 2.3 cm was found on computed tomography. On magnetic resonance imaging, T2-weighted images showed a solid tumor with hyperintensity suggesting intratumoral necrosis. We considered RCC and Wilms' tumor as differential diagnoses, but we could not reach a definitive diagnosis from the radiographic findings. We then performed right partial nephrectomy with regional lymph node dissection. The histopathological diagnosis was pT1aN0 chromophobe RCC. After the surgery, no recurrence has been observed for 15 months.

Keywords Renal cell carcinoma · Nephron-sparing surgery · Chromophobe renal cell carcinoma · Pediatric

Introduction

Renal cell carcinoma (RCC) in children is generally thought to be rare, accounting for only 2–6 % of pediatric malignant renal tumors [1]. Previous reports showed that pediatric RCC presents histological features different from adult RCC, although papillary RCC is the most frequent

type [2]. Chromophobe RCC in children is very rare with, to the best of our knowledge, only a few cases reported in the literature.

Case report

A 12-year-old girl presented in a pediatric clinic with abdominal and back pains. She had no personal or family history of malignant disease. Ultrasonography of the abdomen found a 2.5-cm mass with blood flow in the right kidney. Contrast-enhanced computed tomography (CT) showed a uniformly enhanced tumor measuring 2.5 cm × 2.3 cm in the early phase, and attenuation in the delayed phase (Fig. 1). On magnetic resonance imaging (MRI), T1-weighted images showed a well-defined hypointense mass. T2-weighted images displayed a heterogeneously hyperintense tumor with necrosis. She was referred to our department for treatment of the right renal tumor (Fig. 2).

There were no abnormal data, including neuron-specific enolase (NSE) alpha-fetoprotein (AFP), in her initial laboratory test. After discussion with radiologists, we considered RCC and Wilms' tumor as differential diagnoses from the radiological findings. We decided to treat her with nephron-sparing surgery (NSS), since we thought that the tumor was most likely to be RCC judging from her age, the tumor size and radiographic information. Therefore, we performed right partial nephrectomy. The right renal hilar and paracaval nodes were dissected for staging in case of Wilms' tumor or Xp11 translocation RCC. Macroscopically, the specimen was a 2.5 cm × 2.0 cm, uniformly ocher and glossy tumor with a negative surgical margin (Fig. 3). Microscopically, the tumor cells had clear, eosinophilic cytoplasm and oval, enlarged nuclei with

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Fig. 1 Contrast-enhanced abdominal computed tomography. A tumor measuring 2.5 cm × 2.3 cm uniformly enhanced in the early phase (a) and attenuated in the delayed phase (b)

Fig. 2 Magnetic resonance imaging (MRI). **a** T1-weighted image shows a well-defined hypointense mass. **b** T2-weighted image shows hyperintense mass

Fig. 3 Macroscopic finding of the partial nephrectomy specimen: a uniformly ocher and glossy tumor

perinuclear halos on hematoxylin and eosin staining (Fig. 4). The tumor cells were immunohistochemically positive for cytokeratin AE1/3 and cytokeratin 7. Vimentin, transcription factor E3 (TFE3), Wilms' tumor 1 protein (WT-1), and lymphocyte common antigen (LCA) were negative. The pathological diagnosis was chromophobe RCC, grade 2, pT1a, pN0. She was discharged 8 days after the surgery without any postoperative com-

Fig. 4 Microscopic appearance of the right renal tumor (H&E stain, ×400). Clear, eosinophilic cytoplasm and oval, enlarged nuclei with perinuclear halos are shown

plications. After the surgery, no recurrence has been observed for 15 months. Her serum creatinine level remains within the normal range.

Table 1 Reported cases of chromophobe RCC in children and young adults

References	Sex	Age (y.o.)	Grade	Stage	Tumor size (cm)	Treatment	Outcome (months)
Fujii et al. [12]	M	12	2	I	4.5	RN	NED (12)
Indolfi et al. [11]	F	10	N.I.	I	N.I.	RN	NED (49)
Selle et al. [2]	M	4	N.I.	II	>10	RN	NED (7.2)
Campus et al. [13]	M	7	N.I.	I	3.5 × 3.0	RN	NED (36)
Baek et al. [14]	M	16	N.I.	II	9.0 × 8.0	RN	NED (132)
Our case	F	12	2	I	2.5 × 2.0	PN + LND	NED (12)

N.I., not indicated, *RN* radical nephrectomy, *PN* partial nephrectomy, *LND* lymph node dissection, *NED* no evidence of disease

Discussion

Although we considered the renal masses to be RCC rather than Wilms' tumor as mentioned above, biopsy might be indicated before surgery. According to a recent international consensus panel [3], tumor biopsy is recommended in most cases with a small renal mass except in patients with imaging or clinical characteristics indicative of pathology since there is strong evidence that renal mass biopsy can achieve very high accuracy in identifying malignancy [3]. However, the United Kingdom Children's Cancer Study Group [4] reported that of 182 children who had percutaneous cutting needle biopsy, 20 and 19 % of patients had a fall in hemoglobin and local pain, respectively. Furthermore, one child required emergency nephrectomy due to massive bleeding, another had tumor rupture and subsequently died, and a third developed a needle track recurrence 8 months after the biopsy [4]. Thus renal mass biopsy for children should be carefully indicated and performed only when absolutely necessary.

The standard treatment for localized RCC is surgical resection. NSS is thought to be a favorable approach for patients with T1 disease. Cook et al. reported 5 pediatric patients with RCC smaller than 4 cm who were treated with NSS [5]. All patients obtained 100 % recurrence-free survival (RFS) and overall survival (OS) during the follow-up period. This report suggested that NSS was well tolerated and provided favorable oncological outcomes in pediatric patients with small RCC tumors. A recent study [6] with 304 children aged 0–17 years with RCC from a large national cancer database revealed that survival following partial nephrectomy was 100 % at 1 and 5 years.

In the case of a unilateral Wilms' tumor, radical nephrectomy is considered as standard treatment [7]. Recently, several studies showed the efficacy of NSS against unilateral Wilms' tumors [8–10]. Cost et al. compared the oncologic outcomes of partial nephrectomy (PN) and radical nephrectomy (RN) for unilateral WT patients [8]. In stage I cases, there were no significant differences in RFS (89 versus 83.1 %) and OS (95 versus 95 %) between the

two groups. Furthermore, postoperative renal function in the PN group was better than that in the RN group [9]. Thus NSS is recommended for pediatric patients with renal tumors even if the histology is Wilms' tumor.

We reviewed the literature and found only 5 cases of chromophobe RCC in childhood (Table 1) [2, 10–13]. In the current case, the patient age and tumor grade were in agreement with those of the previous reports. There has been no recurrence during follow-up in any case. The present case suggests that NSS provides favorable outcomes for pediatric patients with localized chromophobe RCC as in adults.

Conflict of interest Naoya Masumori received a research grant fund from Astellas and honoraria from Astellas, Asahi Kasei, GSK and Nippon Shinyaku; Ko Okabe, Hiroshi Kitamura and Naotaka Nishiyama have no conflict of interest.

References

- Pastore G, Znao A, Spreafico F et al (2006) Malignant renal tumours incidence and survival in European children (1978–1997): report from the automated childhood cancer information system project. *Eur J Cancer* 42:2103–2114
- Selle B, Furtwängler R, Graf N et al (2006) Population-based study of renal cell carcinoma in children in Germany, 1980–2005. *Cancer* 107:2906–2914
- Tsivian M, Rampersaud EN Jr, del Pilar Laguna Pes M et al (2014) Small renal mass biopsy—how, what and when: report from an international consensus panel. *BJU Int* 113:854–863
- Vujanic GM, Kelsey A, Mitchell C et al (2003) The role of biopsy in the diagnosis of renal tumors of childhood: results of the UKCCSG Wilms tumor study 3. *Med Pediatr Oncol* 40:18–22
- Cook A, Lorenzo AJ, Salle JL et al (2006) Pediatric renal cell carcinoma: single institution 25-year case series and initial experience with partial nephrectomy. *J Urol* 175:1456–1460
- Rialon KL, Gulack BC, Englum BR et al (2015) Factors impacting survival in children with renal cell carcinoma. *J Pediatr Surg* (in press)
- Ritchey ML, Shamberger RC (2011) Pediatric urologic oncology. In: Wein AJ, Kavoussi LR, Novick AC, Partin AW, Peters CA (eds) *Campbell-Walsh urology*, 10th edn. Elsevier Saunders, Philadelphia, pp 3696–3730
- Cost NG, Lubahn JD, Granberg CF et al (2012) Oncologic outcomes of partial versus radical nephrectomy for unilateral Wilms tumor. *Pediatr Blood Cancer* 58:898–904

9. Cost NG, Sawicz-Birkowska K, Kajbafzadeh AM et al (2014) A comparison of renal function outcomes after nephron-sparing surgery and radical nephrectomy for non-syndromic unilateral Wilms tumor. *Urology* 83:1388–1393
10. Haecker FM, von Schweinitz D, Harms D et al (2003) Partial nephrectomy for unilateral Wilms tumor: result of SIOP 93–01/GPOH. *J Urol* 170:939–942
11. Indolfi P, Terenziani M, Casale F et al (2003) Renal cell carcinoma in children: a clinicopathologic study. *J Clin Oncol* 21:530–535
12. Fujii Y, Kihara K, Tsujii T et al (1998) Chromophobe cell renal carcinoma in childhood. *Int J Urol* 5:92–93
13. Campus R, Nozza P, Dell'Acqua A et al (2006) Eosinophilic renal cell carcinoma in a child with hypospadias. *Pediatr Blood Cancer* 50:413–415
14. Baek M, Jung JY, Kim JJ et al (2010) Characteristics and clinical outcomes of renal cell carcinoma in children: a single center experience. *Int J Urol* 17:737–740