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Case Report Open Access

Renal Cell Carcinoma in Children: About Two Case Report

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Abstract

Introduction

Renal cell carcinoma (RCC) is extremely rare in children. Several factors could influence prognosis, including stage, grade, histology, symptomatic presentation, and performance status. Among these, tumor stage is the most important predictor of disease prognosis for RCC. We report our experience with two cases of children with renal cell carcinoma in children.

Cases

Case 1: A 13-year-old girl presented macroscopic hematuria after a right lumbar trauma. The renal ultrasonography and CT urography showed a very limited rounding medio renal mass. Fine needle renal aspiration objectified RCC. Patient underwent transabdominal radical nephrectomy. Histopathology revealed RCC grade 2 of Furhrman T1b N0 M0. The postoperative course was uneventful.

Case 2: A 9-year-old girl presented macroscopic hematuria. The renal ultrasonography showed a horseshoe left kidney with limited rounding mass. CT urography showed limited left renal mass of 31 × 25 mm. Fine needle renal aspiration objectified RCC. Patient underwent transabdominal radical nephrectomy with regional lymphadenectomy. Histopathology revealed RCC associated with translocation Xp11.2 grade 2 of Furhrman T1a N1M0. The postoperative course was uneventful.

Conclusion

RCC is rare in children but in children older than 5 years with renal masses it is very important to suspect diagnosis. Surgery is the best treatment and prognosis is favorable when the tumor is localized and completely eradicated.

Keywords: Renal cell carcinoma; Children; Treatment

Introduction

Renal cell carcinoma (RCC) is extremely rare in children [1]. Incidence of this tumor in childhood is estimated to be 0.1-0.3% of all the neoplasm and 1.8-6.3% of all the malignant renal tumors [2,3]. Several factors could influence prognosis, including stage, grade, histology, symptomatic presentation, and performance status. Among these, tumors stage is the most important predictor of disease prognosis for RCC [4]. No treatment protocols have been defined for children with RCC. Surgery is the mainstay of treatment and results in cure when the tumor is localized and completely resected [1]. We report two cases of children with renal cell carcinoma.

Case Presentation

Case 1

A 13-year-old girl presented macroscopic hematuria after a right lumbar trauma. The renal ultrasonography and CT urography showed a very limited rounding medio renal mass (Figure 1). Fine needle renal aspiration objectified RCC. There were no metastases in the staging. Patient underwent transabdominal radical nephrectomy (Figure 2). The tumor has 5 cm in diameter, the renal capsule was intact, and there was no tumor thrombus in the renal vein or inferior vena cava. Histopathology revealed RCC grade 2 of Furhrman T1b N0 M0. Immunohistochemical explorations have reported Xp11.2 translocations. The postoperative course was uneventful. There was no recurrence after one year control.

Case 2

A 9-year-old girl presented macroscopic hematuria. The renal ultrasonography showed a horseshoe left kidney with limited rounding mass. CT urography showed limited left renal mass of 31 × 25 mm. Fine needle renal aspiration objectified RCC. There were no metastases in the staging. Patient underwent transabdominal a partial nephrectomy of the horseshoe kidney with regional lymphadenectomy. The tumor has 3 cm in diameter, the renal capsule was intact, and there was no tumor thrombus in the renal vein or inferior vena cava. Histopathology revealed RCC associated with translocation Xp11.2

grade 2 of Furhrman T1a N1M0. The postoperative course was uneventful. There was no recurrence after five years control.

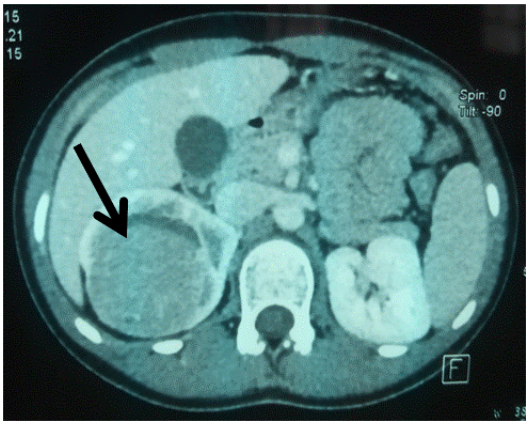

Figure 1: CT urography showed a very limited rounding medio renal mass in the patient number 1.

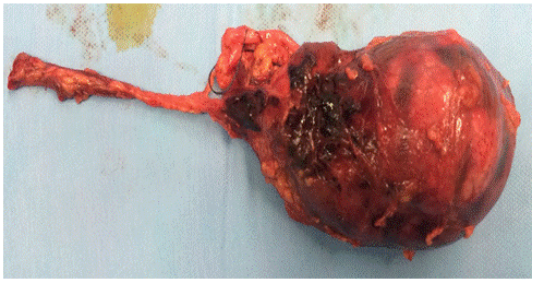

Figure 2: Image objectifying the nephrectomy piece of patient number 1.

Discussion

Literature revealed that renal cell carcinoma corresponds to 1.4% of renal tumors in patients under 4 years old, 15.2% between 5 and 9 years and 52.6% between 10 and 15 years old [2]. Currently, about 50% to 68% of RCC patients are diagnosed incidentally [5]. The most common form of presentation of RCC in children is macroscopic hematuria and abdominal or flank pain. Other less frequent symptoms are palpable abdominal mass, anemia, and fever [6]. Palpable mass occurs in 38%, hematuria in 38% and abdominal pain in 50%, with the classic triad being found in only 6% of cases [7]. Stachowicz-Stencel et al. reported that 52.4% of pediatric RCC cases were asymptomatic and diagnosed during routine examinations [8]. In our two cases diagnosis were done in the exploration of hematuria.

Incidence of metastatic disease, commonly to lung and bones, in children with RCC, similar to adults with half of patients presenting with metastatic disease at the time of diagnosis has been reported. Metastases occur in lungs (40-65%), liver (35-57%), bones (10-42%) or bladder, brain or pleura (7-15%) [9]. Using such immunohistochemical staining, recent studies have reported Xp11.2 translocations in 20% to 70% of pediatric RCC [7]. For that immunohistochemical exploration was made for our two patients and it had founded the Xp11.2 translocations.

Because of the resistance of RCC to systemic therapies and radiotherapy, surgical excision is the main stay of treatment. The completeness of the surgical resection has been shown to be a significant prognostic factor, with survival rates as low as 10% after incomplete resection [10]. The effects of chemotherapy, including immunotherapy, are unclear. Postoperatively, adjuvant radiotherapy and chemotherapy have been used in patients with higher-grade tumors. Although immunotherapies with interferon or interleukin for the treatment of advanced cases have been reported, the beneficial effects of these treatments are uncertain in children because of the lack of any prospective randomized studies [11]. For our two patients it was the renal biopsy which guided us to the best treatment choice and this biopsy was made because of the age of our patients.

Five years survival is more than 90% for patients with stage 1, 50-80% for patients with stages 2 and 3 and less than 10% for stage 4 [7]. Patient age, tumor size, histological pattern, and vascular invasion have all been reported to be the predictors of outcome. Most recurrences and deaths usually occur within the first 2 years after diagnosis, although late recurrences are frequent [1]. Geller et al. suggested that children and adults with RCC have similar overall survival rates [12]. Furthermore, they reported that children with lymph node-positive RCC in the absence of distant metastatic disease had relatively favorable long-term prognoses compared with adults [12]. Geller and Dome conclude that lymphadenectomy in the absence of clinical or radiographic suspicion for nodal involvement confers no benefit [13]. Tumor stage appears to be the most important factor for the survival. The patients with a localized stage (stages 1 and 2) have the best prognosis [10]. Whatever the stage, patients need long-term follow-up after being treated for pediatric RCC.

Conclusion

RCC is rare in children, it is very important to suspect diagnosis in children older than 5 years with renal masses. Surgery is the best treatment and prognosis is better when the tumor is limited and completely removed.

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