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Renal cell carcinoma in children, report of a new case

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A B S T R A C T

Renal cell carcinoma (RCC) is rare in children and is usually found in late children. We present a case of a 14 year-old boy who presented with right lumbar pain. CT-scan showed a tumor in the upper pole of the right kidney measuring 15 cm. He underwent radical nephrectomy and histopathologic examination revealed RCC. No adjuvant therapy was given. After three years and half, there is no evidence of recurrence.

1. Introduction

Renal cell carcinoma (RCC) is a rare tumor in children. Only 0.3%–1.3% of RCC develop in children. It accounts for 2–6% of kidney tumors in children compared to 85–87% for nephroblastoma.[1] However, the distinction between these two types of tumor before treatment is difficult to make in practice, while it remains insensitive to radio-chemotherapy. We report a new case of RCC in a child treated with radical nephrectomy, after ineffective reduction chemotherapy prescribed for suspected nephroblastoma.

2. Observation

This is a 14 and a half year old boy with no notable pathological history who consulted for right back pain that had progressed for 2 months, associated with transit disorders consisting on alternation between constipation and diarrhea. The physical examination found an arching of the right hypochondrium, with a right lumbar mass, painless and mobile on breathing. Abdominopelvic ultrasound showed an upper right polar kidney mass 15 cm long. Computed tomography confirmed the existence of a tumor process developing at the expense of the upper pole of the right kidney with a long axis of 15 cm, with hypodense areas of tumor necrosis, hyperdense areas of bleeding and calcifications. This tumor was intensely enhanced after injection of contrast product, with doubt about invasion of segments IV and V of the liver as well as the gall bladder (Fig. 1).

Given the patient's age and the appearance of the tumor, the diagnosis of nephroblastoma was made and reduction chemotherapy was indicated. After 4 cycles of chemotherapy, no reduction in the size of the tumor was noted and the child was then operated on by the right subcostal approach.

The exploration did not find any involvement of the liver or the gallbladder (Fig. 2) and an radical nephrectomy was performed associated with lymph node dissection (Fig. 3). The postoperative follow-up was uncomplicated. Pathological examination of the surgical specimen revealed renal cell carcinoma with intact renal capsule and a Fuhrman grade of 2. There was no lymph node metastasis and the renal vein was permeable.

No adjuvant treatment was prescribed and the outcome was favorable after a follow-up of 3.5 years, without any tumor recurrence.

3. Discussion

RCC is very rare in children, in whom the majority of kidney tumors are nephroblastomas.[2] The relative frequency with respect to nephroblastoma (Wilms tumor) is 2–6.6%. The incidence of RCC increases with age. While the peak incidence of nephroblastoma is around 3 years, that of RCC is between 9 and 15 years, as is the case in our observation. That is why in most reports, the incidence of RCC vs Wilms in teens is actually higher. As in the case of adults, a male predominance has been reported by some pediatric series.

The clinical picture is dominated by lower back pain (57%), followed by massive hematuria (45%) and lumbar mass (38%).[3] This classic triad is only found in 6% of cases, especially in advanced stages of the disease.[3] In fact, medical imaging has limited value in distinguishing RCC from other childhood kidney tumors.[2] The case we have reported already brought together most of these characteristics. However, to date, the diagnostic aspects of RCC lack specificity and the identification of this type of cancer without resorting to histology remains difficult. As a result, the use of pre-adjuvant chemotherapy is controversial. The

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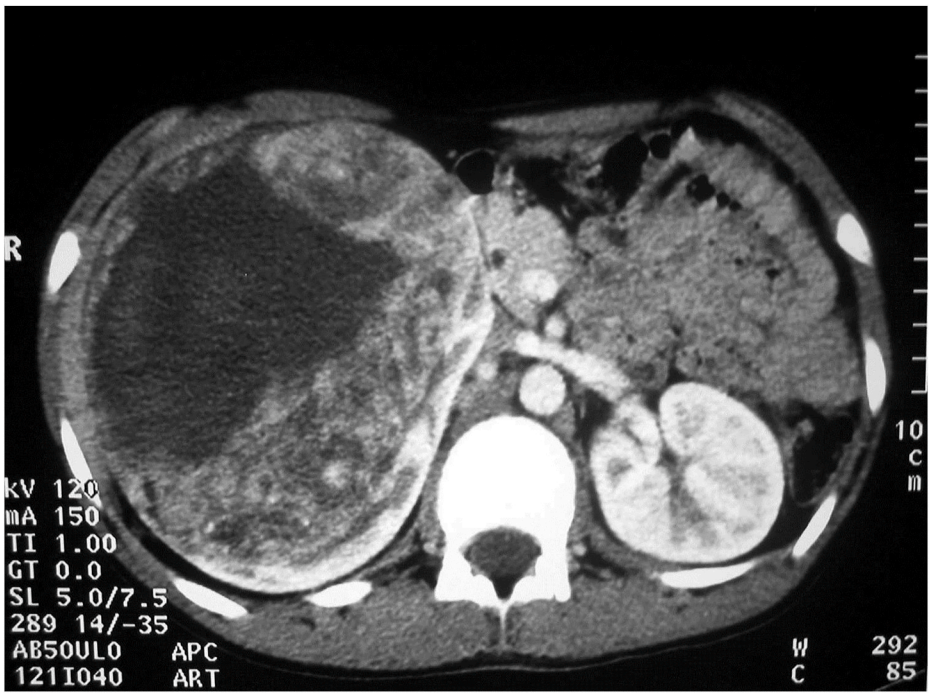

Fig. 1. Tumor of the superior pole of the right kidney with significant uptake of contrast by the tumor which delimits areas of necrosis. Permeable IVC and left renal vein.

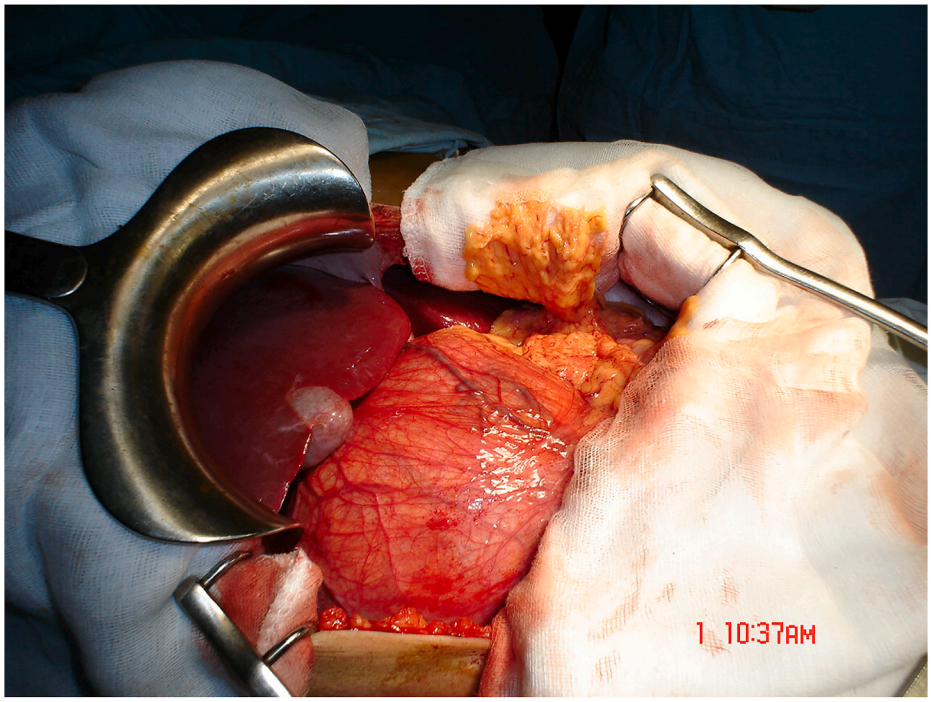

Fig. 2. Intraoperative view, healthy gallbladder liver.

treatment of choice for childhood RCCs is surgical excision, involving an radical nephrectomy combined with lymph node dissection. This lymph node dissection is recommended by certain authors such as Indolfi et al. who find lymph node invasion a factor of poor prognosis and recommend the use of dissection, associated with adjuvant treatment (radiochemotherapy, immunotherapy) in case of positivity.[4] However, these treatments have so far shown questionable effectiveness in addition to their side effects. A lymph node dissection was performed in our patient. If it has not been done during this one, it is not necessary to do it systematically at a second stage (after histological confirmation of the RCC).

The prognosis for this type of tumor in children is similar to that in adults. According to some authors, the factors influencing the patient's prognosis are the size of the tumor, the age of the patient, complete surgical excision, vascular invasion, and clinical and pathological staging.

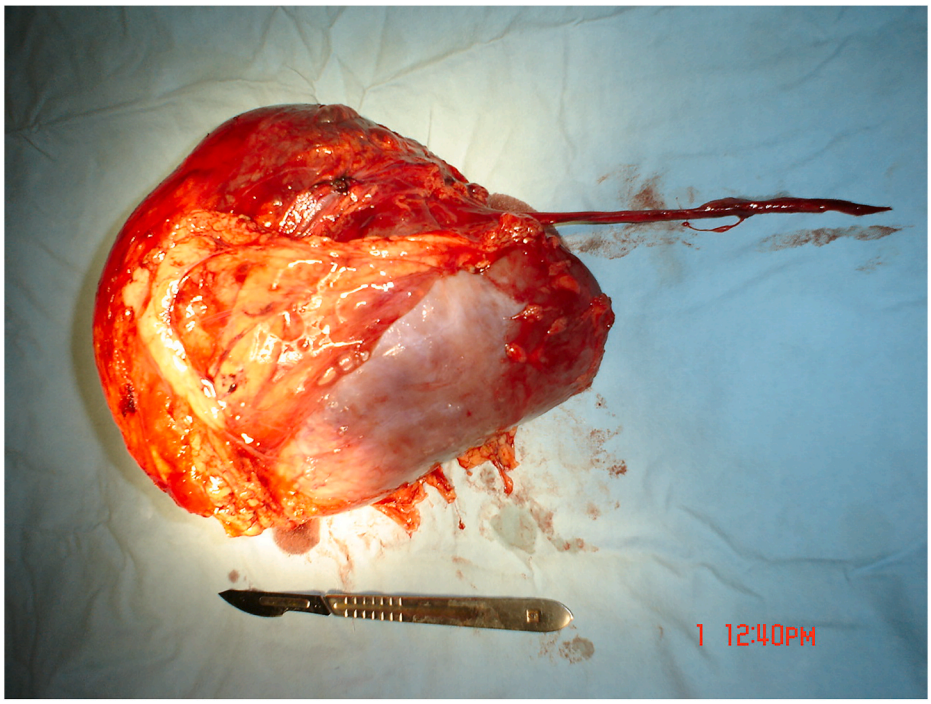

Fig. 3. Enlarged nephrectomy piece after opening the perirenal fat. Ureter folded to the right.

In our patient, after a follow-up of 3 and half years, no suspicious image was found on the control CT scan. This follow-up interval may seem short in the literature. However, most recurrences or deaths occurred within two years of diagnosis.

4. Conclusion

Although very rare in children, RCC should be part of the diagnostic spectrum for kidney tumors occurring in older children. The type of this tumor still poses problems of differential diagnosis with Wilm's tumors preoperatively. Treatment usually consists of an radical nephrectomy, with a prognosis that depends on several factors.

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