CASE REPORT

Baby No More: A Rare Case of Papillary Renal Cell Carcinoma in a One Year Old Female

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Renal cell carcinoma is the most common renal malignancy in adults and extremely rare in children. It may present with hematuria, flank pain and palpable mass. Treatment protocols for renal cell carcinoma in children have not yet been well-defined due to the rarity of the desease, however surgery remains the mainstay treatment for tumors that are resectable. Presented here is a case of a 1 year old female presenting with left hemiabdominal mass, who underwent transabdominal left radical nephrectomy. Histopathology showed a papillary renal cell carcinoma type 1, with positive immunohistochemical stains for Vimentin, CK7 and AMACR.

Keywords: Renal cell carcinoma (RCC), transabdominal radical nephrectomy, papillary renal cell carcinoma type 1, Vimentin

Introduction

Renal cell carcinoma is the most common malignancy of the kidneys in adults however it is extremely rare in children with an estimated incidence of 0.1 to 0.3% of all tumors and 1.8 to 6.3% of all malignant renal tumors in childhood.[1] The biologic behavior and the prognostic factors of RCC are not well-known but may resemble that in adults, so far no treatment protocols have been defined for children with renal cell carcinoma but surgery is the mainstay of treatment when the tumor is resectable.[2] Presented is a 1 year old female with renal cell carcinoma presenting with left hemiabdominal mass.

The Case

This is a case of S.A. a 1 year old female born to a then 29 year old mother via a cesarean section with no feto-maternal complications. Patient has unremarkable birth and maternal history. At one year of age, her mother noticed a palpable soft mass on patient's left hemiabdomen, which was not accompanied by any other symptoms. They consulted a pediatrician wherein a whole abdominal ultrasound was done which showed a 7cm x 6cm mass on the inferior pole of the left kidney. A whole abdominal CT scan with contrast was done and showed an enhancing mass mid to inferior pole of the left kidney measuring 7cm x 6cm with areas of necrosis (Figure 2). Patient was then referred to NKTI for further evaluation and management. Patient underwent a transabdominal left radical nephrectomy. Intra-operatively the authors noted a 7cm x 7cm solid mass occupying mid to inferior pole of the left kidney (Figure 2). Neither thrombus nor palpable lymph nodes were noted. Histopatholigical report of the mass showed a papillary renal cell carcinoma Type 1 (Figure 3)

with positive immunohistochemical stains for CK7, Vimentin, AMACR and CK (Figure 4). All eight nodes were negative for tumor and the margins were negative. Patient was then discharged stable on the sixth post operative day.

Discussion

Renal cell carcinoma represents 2% of malignant tumors in adults and is the third most frequent tumor of the urinary tract after prostate and bladder tumors. On the other hand, in pediatric ages, only 2% to 3% of malignant renal tumors have been proven to be renal cell carcinoma.[3] The incidence of RCC increases with age, according to the survey of the Japanese Society of Paediatric Surgeons, RCC accounted for 1.4% of all renal tumors in patients younger than 4 years, 15.2% in patients aged 5 to 9 years, and 52.6% in patients aged 10 to 15 years.[4]

Generally, there is no sex predominance for this renal tumor type in children. In the literature unlike in adults, the tumor predominates in males. The most common form of presentation of RCC in children is macroscopic hematuria and

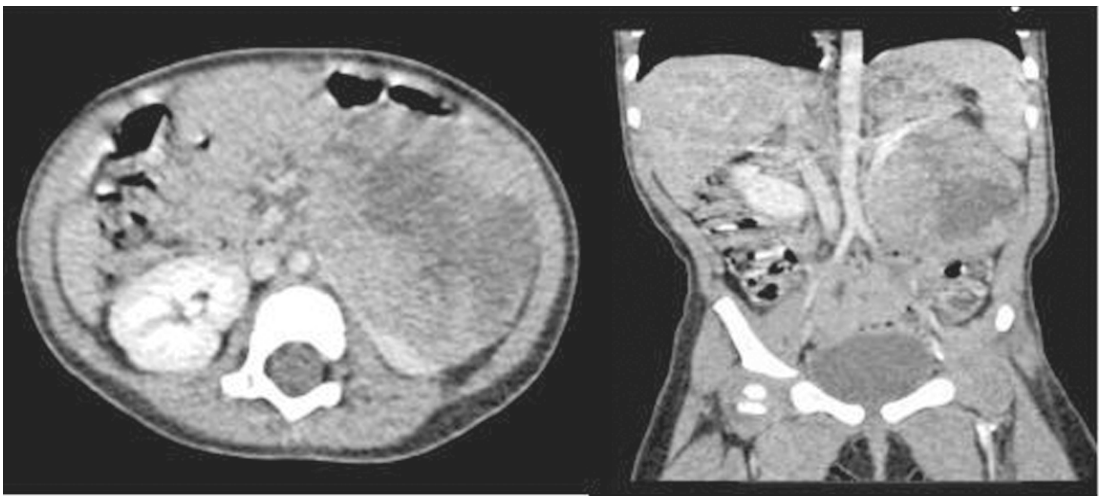

Figure 1. Whole abdominal CT scan showing a 7cm x 6cm mass occupying mid to inferior pole of left kidney, (Right) axial cut, (Left) coronal cut.

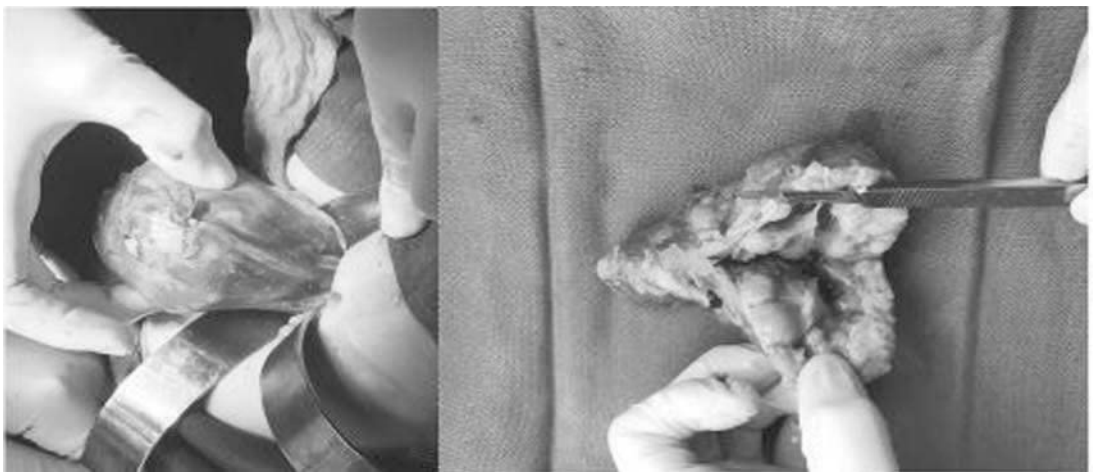

Figure 2.Right: Intraoperative picture of the 7cm x 7cm left renal mass occupying the mid to inferioe pole. Left: Cut section showing the mid to inferior left renal mass

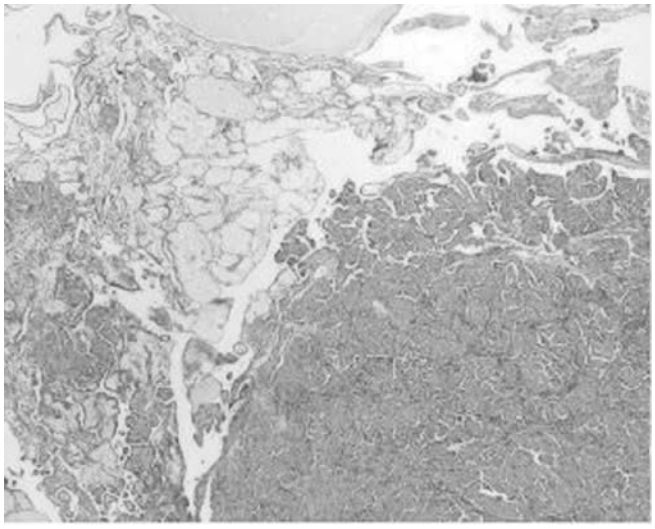

Figure 3. Histopath of the tumor showing a complex papillary formation with sheets of macrophages.

abdominal or flank pain. Other less frequent symptoms are palpable abdominal mass, anemia, and fever.[5] Palpable mass occurs in 38%, hematuria in 38% and abdominal pain in 50%, with the classic triad being found in only 6% of cases.[6]

Two subtypes of papillary RCC are recognized based on their histologic features. Type 1 papillary RCC is the most frequent, accounting for approximately two-thirds of all PRCCs and is composed of papillae covered with a single layer of small cells and scant clear or pale cytoplasm and uniform nuclei with inconspicuous nucleoli.[7] Type 2 PRCC is composed of tumor cells with voluminous cytoplasm and pseudostratified high-grade nuclei with prominent nucleoli. These subtypes also differ in their immunohistochemical phenotypes. CK7 is positive in 87% of type 1 and 20% of type 2 lesions[8] ; EMA, Vimentin, and AMACR are typically positive in both types.[9] In the present case, the specimen was positive for CK7, Vimentin and AMACR. So far, no treatment protocols have been defined for children with renal cell carcinoma but surgery is the mainstay treatment when the tumor is localized.[2]

Overall survival rate of paediatric RCC is around 63%, with survival rates for stages I to IV at 92.4%, 84.6%, 72.7%, and 13.9%, respectively. Patient age, tumor size, histological pattern, and vascular invasion have all been reported to be predictors of outcome.[10]

Conclusion

Renal cell carcinoma in children is rare, and there has not been an established standard treatment protocol. However, immunostains to diagnose such rare entity are available in the Philippines. Treatment for renal cell carcinoma for children remains to be surgery once resectable.

References

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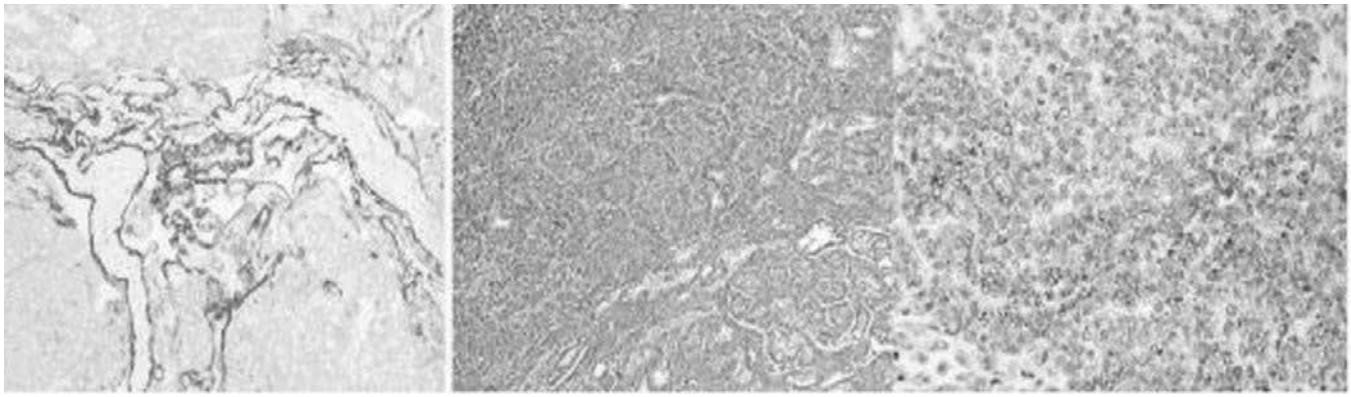

A B C

Figure 4. Immunohistochemical stain of the specimen positive for(A) CK 7, Vimentin (B), AMACR (C)

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