



## Primitive Neuroectodermal Tumor (PNET) of Kidney: A Rare Tumor in Differential Diagnosis of Renal Mass in Adolescents

Farzad Allameh<sup>1</sup>, Hamidreza Qashqai<sup>1</sup>, Farideh Mousavi<sup>2</sup>, Ali Asghar Halimi-Asl<sup>2</sup>, Amirhossein Rahavian<sup>1</sup>

1- Department of Urology, Shohada-e-Tajrish Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

2- Department of Pediatric Oncology, Shohada-e-Tajrish Hospital, Shahid Beheshti University of Medical Sciences, Tehran, Iran

Received: 6 October 2016

Revised: 16 October 2016

Accepted: 13 November 2016

### ARTICLE INFO

Corresponding author:  
Hamidreza Qashqai

Email:  
qashqae@gmail.com

Keywords:  
Primitive Neuroectodermal  
Tumor (PNET);  
Renal tumor;  
Biopsy;  
Metastasis

### ABSTRACT

A thirteen-year-old boy with a right flank mass and painless gross hematuria is presented. The patient underwent open biopsy and the result was a huge primitive neuroectodermal tumor (PNET) of right kidney. Radical nephrectomy was performed for him and the treatment process continued with systemic chemotherapy.

**Citation:** Allameh F, Qashqai H, Mousavi F, Halimi-Asl AA, Rahavian A. **Primitive Neuroectodermal Tumor (PNET) of Kidney: A Rare Tumor in Differential Diagnosis of Renal Mass in Adolescents.** Case Rep Clin Pract 2016; 1(4): 102-5.

### Introduction

Paediatric renal masses are some of the most common kinds of childhood solid neoplasms, where both diagnosis and treatment are highly dependent on the histopathological findings.

Primitive neuroectodermal tumor (PNET) is a rare malignant tumor that originates from neural crest and it may have distant

metastasis. This usually occurs in young people and has a poor survival rate (1). PNETs of the kidney have been sporadically reported before (2). These tumors display characteristic translocations (the t(11;22)(q24;q12)) with the fusion of EWS1 to FLI1 genes (3, 4). The PNET histopathologic features have similarities to other small round cell tumors, including

desmoplastic small-round-cell tumors (DSRCTs) and Wilms' tumor (WT) of pure blastemal type (5).

DSRCTs are rare malignant tumors that tend to affect the abdominal serosa and are usually found in adolescent males that has recently been described in the kidney in pediatric patients (aged 6–8 years) (6, 7). The diagnosis should only be made if the EWS-WT1 t(11;22)(p13;q12), a unique reciprocal translocation is demonstrated, which fuses the Ewing sarcoma gene EWS1 to the Wilms' tumor suppressor gene WT1 (4, 8).

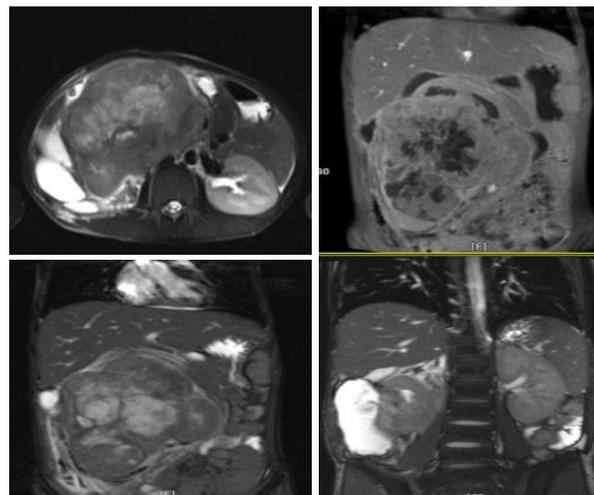
Wilms' tumor is a malignant neoplasm derived from nephrogenic blastema. It represents approximately 85% of renal tumors and about 7% of all pediatric cancers (9). Most of the cases occur in young children, while rare congenital and adult cases have been sporadically reported (4). Wilms' tumor usually shows a distinctive triphasic appearance with epithelial, stromal, and blastemal elements. The blastemal component exhibits the features of a classical embryonal small round blue cell tumor including round to ovoid cells, a high nucleo-cytoplasmic ratio, closely packed nuclei, nuclear overlapping, and mitoses (4).

### Case Report

A thirteen-year-old boy was referred to our medical center with an abdominal mass. The patient had a history of painless gross hematuria from 5 months before the entrance that was diagnosed as a right flank mass after 2 months. The patient was suffering from flank pain, on and off fevers, constipation, and weight loss of 4 kg during one month.

His medical history was otherwise uneventful. His family history was negative for any kind of cancer. In physical examination, he was febrile and had a firm, tender abdominal mass. Urinalysis revealed gross hematuria and renal function tests were in normal range. Abdominal and pelvic computed tomography (CT) scan revealed a huge right kidney mass with pressure effect

on liver and a mild right hydronephrosis was detected. Lung CT scan showed at least two metastatic nodules in both lungs. Abdominal and pelvic magnetic resonance imaging (MRI) confirmed a huge right kidney mass measuring 205 x 180 mm with extension to left para-aortic area and displacing bowel loops. Inferior vena cava (IVC) and renal vein were also engaged by tumor (Figure 1).



**Figure 1.** Magnetic resonance imaging (MRI) shows a huge mass in right kidney (a, b, c, and d)

A needle biopsy was done to start chemotherapy, but it was not conclusive. The Wilms' tumor was suspected but was not proved certainly. Therefore, a laparotomy was planned to do radical nephrectomy. At the time of operation, we found a huge non-resectable mass with extreme extension to adjacent organs and IVC encasement. Therefore, the patient underwent an open biopsy of the mass, only.

Pathologist's report was very helpful and changed the path of treatment. Sections showed parts of renal cortex infiltrated by malignant, cancerous growing cells, composed of sheets, and clusters of small round uniform cellular material intervening renal tubular tissue without epithelial and stromal elements. PNET was diagnosed which was verified by immunohistochemistry (IHC) staining. WT1, CD45, and Synaptophysin were negative and CD99 was positive.

After the establishment of diagnosis, chemotherapy was started with vincristine, doxorubicin, cyclophosphamide, etoposide, and ifosfamide. After five courses of chemotherapy, lung metastases were disappeared and tumor was decreased in dimensions significantly (Figure 2).



Figure 2. Magnetic resonance imaging (MRI) after chemotherapy shows tumor regression (a, b, and c)

Then, we performed right radical nephrectomy for him. Our pathologist evaluated the final specimen and reported that macroscopic tumor dimensions were  $9.5 \times 6.5 \times 4.5$  cm and Gerota's fascia was intact. Tumor was unifocal, including renal tissue in inferior pole and renal sinus soft tissue. The almost total necrotic neoplasm was present with only a few numbers of viable cancerous growing cells (malignant small-round-cell neoplasm). Recognition of renal vein intrusion and also tumor expansion to adjacent organs was negative and adrenal tissue was intact. Identification of nephrogenic rest was negative. Surgery margins including Gerota's fascia, renal, and ureter were free of tumor. Lymph node was not determined.

Chemotherapy was continued after second operation and follow-up visits continued 18 months after operation and showed neither local recurrence nor metastasis.

## Discussion

Because the accurate diagnosis of renal tumors may be difficult by using the needle biopsy, this may present a set of diagnostic challenges to pathologists. In a study, over 241 cases of renal masses, despite epidemiologic, clinic, and radiologic findings pointed to Wilms' tumor; in 12% of cases, needle biopsy revealed different histologies

other than Wilms' tumor and about 18% of cases were of pure blastemal Wilms' tumor; finally, biopsy material was inadequate for diagnosis in 4% (10).

As PNET of kidney is uncommon, we could about to point out a handful of cases below:

Pomara et al. presented a woman, twenty seven years of age, who had developed mild flank pain and hematuria. The person was diagnosed with PNET of kidney, so she went through radical nephrectomy, lymphadenectomy, and chemotherapy, and after twenty four months, no recurrence was detected (11).

Mandal et al. presented four cases of PNET in the age group of 30-50 years of age who complained about flank pain, mass, chronic fever, and hematuria. Radical nephrectomy and lymphadenectomy were performed on all of them and following that, the patients received chemotherapy and radiotherapy. All the patients ended up in good condition (12).

Kairouani et al. presented a 40-year-old woman with metastatic PNET suffering from severe back pain and abdominal mass. After radical nephrectomy and pathologic analysis, she was diagnosed with PNET. Chemotherapy was done and the patient got it tolerated. 4 months later, she was in an effective condition (13).

As conclusion, in adolescents, almost all renal masses are likely to be Wilms' tumor or renal cell carcinoma; but the presence of an uncommon tumor like PNET is probable. So, this point emphasizes the importance of a conclusive biopsy that has a very important role in correct diagnosis.

## Conflict of Interests

Authors have no conflict of interests.

## Acknowledgments

We should appreciate Dr. Mohammadreza Razzaghi, as the head of our department for his close observation and guidance, and Dr. Saman Najafi for his efforts and helps in preparation of this paper.

## References

1. Hamidi N, Esen B, Kivrak H, Sertcelik A, Gulpinar O. A large and metastatic primitive neuroectodermal tumor of the kidney. *Turk J Urol* 2015; 41(3): 152-4.
2. Chu WC, Reznikov B, Lee EY, Grant RM, Cheng FW, Babyn P. Primitive neuroectodermal tumour (PNET) of the kidney: a rare renal tumour in adolescents with seemingly characteristic radiological features. *Pediatr Radiol* 2008; 38(10): 1089-94.
3. Saxena R, Sait S, Mhawech-Fauceglia P. Ewing sarcoma/primitive neuroectodermal tumor of the kidney: a case report. Diagnosed by immunohistochemistry and molecular analysis. *Ann Diagn Pathol* 2006; 10(6): 363-6.
4. Sebire NJ, Vujanic GM. Paediatric renal tumours: recent developments, new entities and pathological features. *Histopathology* 2009; 54(5): 516-28.
5. Chang F. Desmoplastic small round cell tumors: cytologic, histologic, and immunohistochemical features. *Arch Pathol Lab Med* 2006; 130(5): 728-32.
6. Gerald WL, Rosai J. Case 2 Desmoplastic Small Cell Tumor with Divergent Differentiation. *Pediatr Pathol* 1989; 9(2): 177-83.
7. Lae ME, Roche PC, Jin L, Lloyd RV, Nascimento AG. Desmoplastic small round cell tumor: a clinicopathologic, immunohistochemical, and molecular study of 32 tumors. *Am J Surg Pathol* 2002; 26(7): 823-35.
8. Sawyer JR, Tryka AF, Lewis JM. A novel reciprocal chromosome translocation t(11;22)(p13;q12) in an intraabdominal desmoplastic small round-cell tumor. *Am J Surg Pathol* 1992; 16(4): 411-6.
9. Birch JM, Breslow N. Epidemiologic features of Wilms tumor. *Hematol Oncol Clin North Am* 1995; 9(6): 1157-78.
10. Vujanic GM, Kelsey A, Mitchell C, Shannon RS, Gornall P. The role of biopsy in the diagnosis of renal tumors of childhood: Results of the UKCCSG Wilms tumor study 3. *Med Pediatr Oncol* 2003; 40(1): 18-22.
11. Pomara G, Cappello F, Cuttano MG, Rappa F, Morelli G, Mancini P, et al. Primitive Neuroectodermal Tumor (PNET) of the kidney: a case report. *BMC Cancer* 2004; 4: 3.
12. Mandal PK, Mukherjee S, Roy S, Bhattacharyya NK. PNET of kidney: Report of four cases. *Indian J Med Paediatr Oncol* 2012; 33(2): 130-3.
13. Kairouani M, Mokrim M, Mellas N, Khennoussi B, M'rabti HE, Boutayeb S, et al. Metastatic Ewing's sarcoma/PNET of kidney in 40 year old patient. *Int J Surg Case Rep* 2012; 3(6): 215-7.