



## Oncology

## A rare case of retroperitoneal primitive neuroectodermal tumor (PNET)

P. Genov<sup>\*,1</sup>, I. Serbezova, D. Georgieva, G. Koleva, I. Hristova

University of Ruse "Angel Kanchev" Ruse, 8 "Studentska" str.7000, Bulgaria

## ARTICLE INFO

## Keywords:

Primitive neuroectodermal tumor (PNET)  
Kidney  
Radical nephrectomy

## ABSTRACT

Retroperitoneal primitive neuroectodermal tumors (PNET) is a rare neoplastic disease of high malignancy with a tendency towards early metastasis, affect young adults irrespective of the gender. We present 81 year old woman, who was admitted in the Urology Department with symptoms of right flank pain and hematuria. Contrast-enhanced computerized tomography scan (CT-scan) showed a large heterogeneous right kidney mass around 12 cm in diameter. The final diagnosis of primitive neuroectodermal tumor (PNET) was established based on clinical, pathological, and molecular results.

## Introduction

Retroperitoneal primitive neuroectodermal tumor (PNET) is a rare disease with high malignancy and with tendency to give early metastasis. It affect young people irrespective of the gender. The clinical symptoms are nonspecific and patients most frequently are presenting with abdominal pain or hematuria. Also radiology imaging is nonspecific and generally shows a heterogeneous renal mass. Less than 100 cases of PNET have been published in the medical literature, since its first description in 1975.

## Case presentation

We present a 81 year old woman, who was admitted in the Urology Department with symptoms of recurrent right flank pain and hematuria. The blood chemo indicators and biochemical investigations were in normal ranges, except the hemoglobin-108 G/L. Contrast-enhanced computer tomography scan (CT-scan) showed a large heterogeneous right kidney mass around 12 cm in diameter, engaging the lower pole and enhanced its density after the contrast. The mass was containing large central cystic component and homogeneous peripheral hypodense zones, suggesting necrosis. (Fig. 1).

The patient underwent laparoscopic radical right nephrectomy with preliminary diagnosis of renal cell carcinoma. The whole tumor was removed using sharp and blunt dissection. Macroscopically the tumor replaced almost completely the kidney parenchyma and its weight was around 800 g. Histopathology results showed a malignant tumor

composed of monomorphic cells forming structures like rosettes, with irregular nuclei and dense chromatin. From immunochemistry, the cells were positive for CD99, for Vimentin anticorps and protein S-100, but negative for Epithelial membrane antigen (EMA), Leucocyte common antigen (LCA) and Desmine (Fig. 2). Based on all clinical and histopathological results the final diagnosis was primitive neuroectodermal tumor (PNET).

## Discussion

Primary renal primitive neuroectodermal tumor (PNET) is a very rare condition. The first case was reported by Seemayer et al., in 1975 and since then, there have been less than 100 cases reported in the literature. The origin of this tumor is unknown, but it seems that it derives from cells that migrated from the neural tube with variable capability of ectodermal or neuronal differentiation.<sup>1</sup>

The PNET is effecting mostly young people around their thirties, with a slight predominance in male. There are no specific clinical signs and symptoms, and the diagnosis of PNET is based on histopathology. Histologically, PNET is composed by monomorphic small cell rounds which form Homer right rosettes, also the immunohistochemistry examination is always required and it shows immature primitive malignant tumour, strongly positive for CD99.<sup>2</sup>

Due to the small count of cases published in the literature, there is no standard guideline for diagnosis and treatment of PNET. Therefore a multimodal treatment is required and the tumor has a trend to relapse locally, and to develop metastasis in the regional lymph nodes, lung,

\* Corresponding author. UMHAT "Kanev" Ruse, 2 "Nezavisimost" str.7000, Bulgaria.

E-mail addresses: [genov\\_p@abv.bg](mailto:genov_p@abv.bg) (P. Genov), [vania\\_serbezova@abv.bg](mailto:vania_serbezova@abv.bg) (I. Serbezova), [dpgeorgieva@uni-ruse.bg](mailto:dpgeorgieva@uni-ruse.bg) (D. Georgieva), [gkoleva@uni-ruse.bg](mailto:gkoleva@uni-ruse.bg) (G. Koleva), [ihristova@uni-ruse.bg](mailto:ihristova@uni-ruse.bg) (I. Hristova).

<sup>1</sup> Present/permanent address: 97 "Aleksandrovska" str. Ruse Bulgaria 7000.

<https://doi.org/10.1016/j.eucr.2020.101554>

Received 6 December 2020; Received in revised form 19 December 2020; Accepted 24 December 2020

Available online 30 December 2020

2214-4420/© 2020 The Authors.

Published by Elsevier Inc.

This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

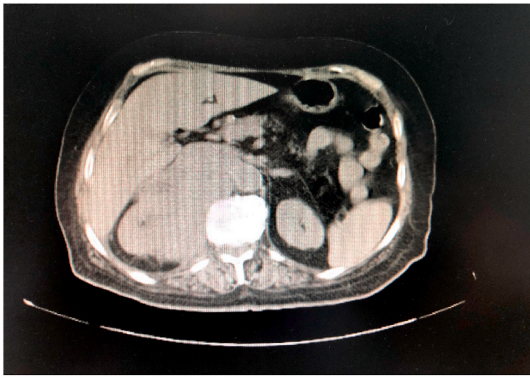


Fig. 1. CT scan of tumor mass at the right kidney.

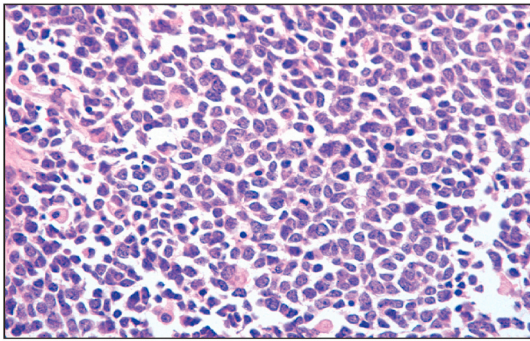


Fig. 2. Histopathology image of PNET in kidney.

liver and bones. The prognostic factors are based on the tumor stage,

grade, patient's age, extent of the surgery, surgical margin status, treatment beginning time and etc. There are some studies showed that the tumor is radio and chemotherapy sensitive and they can be used as a standard treatment if necessary.<sup>3,4</sup>

### Conclusions

Because of its rarity, the management of retroperitoneal PNET remains difficult, and the results are uncertain. The prognosis is poor, despite aggressive treatment. Also there is a high chance for late local relapses and due to this it requires multimodality treatment with surgery, radiotherapy and chemotherapy.

### Section headings

Oncology.

### Declaration of competing interest

The authors declare that they have no competing interests.

### References

1. Nicolaus Friedrichs, Vorreuther Roland, Poremba Christopher, et al. Primitive neuroectodermal tumor (PNET) in the differential diagnosis of malignant kidney tumors. *Pathol Res Pract.* 2002;198(8):563–569.
2. Kairouani Mouna, Mokrim Maha, Mellas Nawfel, et al. Metastatic Ewing's sarcoma/ PNET of kidney in 40 year old patient. *Int J Surg Case Rep.* 2012;3(6):215–217.
3. AlAhmadi Hassan H, Al Essa Ahmed, Ahmed Ayesha, et al. Primary ewing sarcoma/ primitive neuroectodermal tumor of the kidney. *J Pediatr Surg Case Rep.* October 2020;61:101608.
4. Marinova Lena. Retroperitoneal primitive neuroectodermal tumour (PNET). A case report and review of the literature. *Rep Practical Oncol Radiother.* November–December 2009;14(6):221–224.