

Case Report

Case report of a rare presentation of perirenal primitive neuroectodermal tumor

Yi Jiang^{1*}, Gongxian Wang^{1*}, Qiyang Xu^{1*}, Fang Du², Yibing Wang³, Tingxuan Huang⁴, Bin Fu¹

¹Department of Urology, The First Affiliated Hospital of Nanchang University, Nanchang, Jiangxi, P. R. China;

²Department of Hematology and Oncology, The First People's Hospital of Guangzhou Affiliated to Guangzhou Medical University, Guangzhou, P. R. China; ³Department of Emergency, The Second Affiliated Hospital of Nanchang University, Nanchang, Jiangxi, P. R. China; ⁴The Undergraduate of Nanchang University, Nanchang, Jiangxi, P. R. China. *Equal contributors.

Received January 5, 2016; Accepted April 25, 2016; Epub June 1, 2016; Published June 15, 2016

Abstract: Primitive neuroectodermal tumors (PNETs) belong to a group of highly malignant central nervous system tumors that are composed of small, round, undifferentiated neuroectodermal cells. The current study reports a case of perirenal PNET in a 25-year-old female. The tumor presented as an adrenal tumor and caused symptoms of left waist and back pain, which increased over a 2-month period. Open surgery was performed to achieve radical mass resection. The pathological and immunohistochemical diagnosis confirmed that the tumor was a perirenal PNET, which had spread into nervous and adrenal tissue. To our knowledge, the present case of perirenal PNET is the first to be reported in PubMed.

Keywords: CD99, diagnosis, perirenal, primitive neuroectodermal tumors

Introduction

PNET was first proposed and named by Hart and Earle [1] in 1973. The annual PNET incidence rate is low, with approximately 0.2-0.4 cases per 100,000 individuals [2].

Case report

The patient was a 25-year-old female who was admitted to the hospital due to a two-month history of left waist and back pain, which had no obvious cause and intermittently and occasionally radiated toward the lower left abdomen. The patient was healthy previously and did not have any related medical history such as hypertension or hypokalemia. A physical examination indicated that neither kidney exhibited tenderness, percussion pain, or a palpable mass. The computed tomography (CT) plain scan showed a round, lump-shaped shadow adjacent to the anteromedial margin of the median and superior poles of left kidney with a size of approximately 5 × 3 cm. The left renal pelvis was full, and the left perirenal fascia was thickened and blurred. The enhanced CT exam-

ination showed that the tumor was heterogeneously enhanced, and a necrotic area was observed inside the tumor. Computed tomography angiography (CTA) suggested that the local left renal artery and its branches were embedded in the mass, the left renal vein was compressed by the mass and had become flat, and no blood-supplying artery was present inside the mass. The imaging results suggested an adrenal gangliocytoma. A CT examination of other locations and whole body bone imaging did not show signs of tumor metastasis. Laboratory examinations including routine blood tests, routine urine tests, liver and kidney function, and carcinoembryonic antigen (CEA), alpha fetoprotein (AFP), carbohydrate antigen 19-9 (CA19-9), and CA12-5 tests did not show any abnormalities. The preoperative diagnosis was a left adrenal gland tumor. After preoperative preparation of all items, a robot-assisted laparoscopic adrenalectomy was performed in our hospital. During the surgery, a mass was observed in the left renal sinus that wrapped the renal pedicle vessels; in addition, the mass was closely adhered to the pancreatic tail, the left side of the peritoneum, and the splenic vein

Primitive neuroectodermal tumor

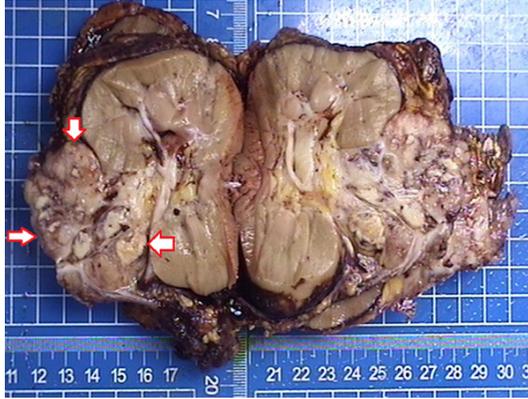


Figure 1. The postoperative specimen showed that the mass was located in the renal hilum and was approximately 6 × 5 × 5 cm in size.

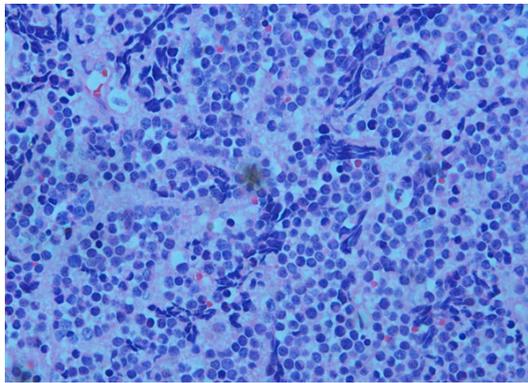


Figure 2. The tumor cells were medium in size, and form a Homer Wright rosette structure (magnification × 400).

and could not be separated. After discussion with general surgery experts, the robot-assisted laparoscopic adrenalectomy was changed to an open surgery. An L-shaped incision was made at the left abdomen, and the mass was observed to have a hard texture and an unclear boundary with surrounding tissues. After the mass was separated from the abdominal aorta, superior mesenteric artery, pancreatic tail, and splenic pedicle vessels using sharp dissection, the mass, left kidney, and adrenal tissues were entirely resected. Intraoperative blood loss was approximately 1500 ml, and the patient was transfused with 2 units of red blood cell suspension and 200 ml of fresh frozen plasma. The patient showed stable vital signs after surgery. Postoperative specimen detection showed that the mass was located at the renal hilum and closely wrapped around the renal pedicle vessels, but it had a clear boundary

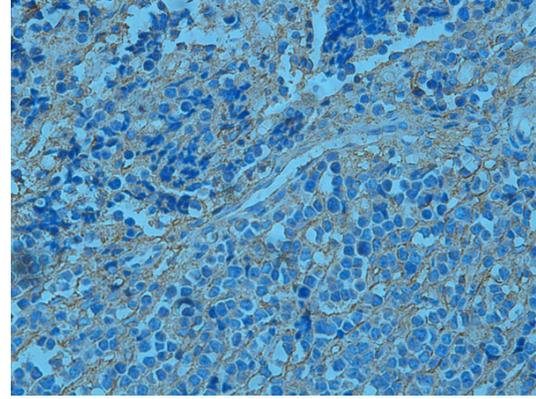


Figure 3. CD99 + (magnification × 400).

with the surface of the renal parenchyma. The size was approximately 6 × 5 × 5 cm (**Figure 1**), the section was gray-red and lobulated, and the focal lesion was gray-white. Observation under a microscope demonstrated that the tumor cells exhibited a nest-like distribution, the cell volume was medium, the cells were round (**Figure 2**), the volume of the cytoplasm was small, some cells seemed to have bare nuclei that were large and darkly stained, and mitotic cells were easily observed using a 4-5/high power field (HPF). The immunohistochemistry results consisted of the following: CD99 (3+) (**Figure 3**), Vim (2+), NSE (+), CD56 (+), Ki-67 (40%+), CK (-), CD10 (-), CgA (-), and EMA (-). The pathological diagnosis indicated left Perirenal primitive neuroectodermal tumors with nerve and adrenal gland tissue involvement; the ureteral stump and renal parenchyma did not exhibit lesion involvement. A systemic CT examination performed one month after surgery did not show metastatic lesions.

Discussion

PNET is a type of highly malignant, undifferentiated tumor composed of small, round cells. It can develop at any age and is mainly observed in children and adolescents. It is classified as a central PNET (cPNETs) or a peripheral PNET (pPNET) in clinical practice. cPNETs originate from the neural tube and mainly occur in the brain and spinal cord. pPNETs originate from the neural crest and mainly occur in the sympathetic nervous system, soft tissues, and bone outside the central nervous system. A search of domestic and foreign literature showed more than 100 cases of pPNETs originating from kidney, while no reports of perirenal pPNETs were found.

Primitive neuroectodermal tumor

In rare cases, preoperative imaging examinations performed for differential diagnosis and determination of the properties and origin of tumors are difficult to conduct and prone to misdiagnosis; therefore, attention should be focused on other features. This case exhibited a unique location at the top inside the kidney. In addition, CT enhanced scanning showed that the boundary of the tumor was located at the surface of the renal parenchyma; therefore, the preoperative imaging diagnosis indicated a left adrenal gland tumor. During surgery, the imaging results were confirmed, which showed that the tumor was closely wrapped in renal pedicle vessels and was adhered to the surrounding tissues including the adrenal gland, kidney, abdominal aorta, and pancreas. A postoperative pathological examination excluded the possibility that the tumor originated from the kidney or adrenal gland.

The final diagnosis for confirmation of a PNET relies on pathological and immunohistochemical examinations. According to a review of the relevant literature and the features of this case, we showed that the perirenal PNET had similar morphological and immunohistochemical features to pPNETs observed at other common locations. A pathological examination showed that tumor cells exhibited a large-patch or nest-shape distribution, the cell volume was small, the cells were round or oval-shaped, the volume of the cytoplasm was small, nuclei were large and darkly stained, heterogeneity was obvious, and many mitotic phases were present. One study [3] reported that PNET tumor cells could form a Homer Wright rosette structure, which is considered the typical histological presentation of a PNET and is conducive to forming a diagnosis. The expression of CD99 is usually positive in all PNETs and shows relative specificity. The positive rate of CD99 is 90%-100%. Vimentin usually exhibits diffuse expression in PNETs. Expression of at least one neural marker, such as NSE and CK, is typically positive [4]. Immunohistochemistry combined with the characteristic presentation of the pathology is sufficient for the diagnosis of a PNET.

Because the number of clinical cases of pPNET is limited, no definitive treatment guidelines have been developed. Most scholars accept the regimen of radical surgical resection combined with chemotherapy as the primary treatment strategy. The main chemotherapy drugs

include doxorubicin, vincristine, cyclophosphamide amides, actinomycin D, ifosfamide and etoposide, while the commonly used combination therapies include CAV, VIDE, VAI, and EI. However, Ishii et al. [5] showed that the 5-year survival rate of patients receiving combined therapy was still in the range of 45% -55%. Due to the lack of reports of perirenal PNETs in the literature, this patient was treated with the regimen of radical tumor resection combined with radiotherapy and chemotherapy.

Consider to this was the first report of a perirenal PNET in pubmed, and its clinical presentations, biochemical indicators, and the imaging examination lacked specificity, these tumors can be easily misdiagnosed as kidney, adrenal gland, or retroperitoneal tumors before surgery. A perirenal PNET is a highly malignant neurological tumor with a short disease course and poor prognosis; therefore, comprehensive treatment regimens mainly based on surgery and assisted with radiotherapy and chemotherapy should be promoted. This regimen highly differs from common gastric cancer treatment regimens. Therefore, it is necessary to summarize the features of the preoperative imaging diagnosis and differential diagnosis and continuously increase the diagnosis level at the early stage to avoid misdiagnosis and mistreatment.

Acknowledgements

This work was supported by the National Natural Science Foundation Program in China (No. 81560419); the Natural Science Foundation of Jiangxi Province (No. 2015BAB205047).

Disclosure of conflict of interest

None.

Address correspondence to: Bin Fu, Department of Urology, The First Affiliated Hospital of Nanchang University, 17 Yongwaizhen Road, Nanchang 330-006, Jiangxi, P. R. China. Tel: +86 791 88698102; Fax: +86 791 88623153; E-mail: fubinncu@163.com

References

- [1] Hart MN and Earle KM. Primitive neuroectodermal tumors of the brain in children. *Cancer* 1973; 32: 890-897.
- [2] Valle JW, Eatock M, Clueit B, Gabriel Z, Ferdinand R and Mitchell S. A systematic re-

Primitive neuroectodermal tumor

- view of non-surgical treatments for pancreatic neuroendocrine tumours. *Cancer Treat Rev* 2014; 40: 376-389.
- [3] Shibuya T, Mori K, Sumino Y, Sato F and Mimata H. Rapidly progressive primitive neuroectodermal tumor of the prostate: A case report and review of the literature. *Oncol Lett* 2015; 9: 634-636.
- [4] Celli R and Cai G. Ewing Sarcoma/Primitive Neuroectodermal Tumor of the Kidney: A Rare and Lethal Entity. *Arch Pathol Lab Med* 2016; 140: 281-285.
- [5] Iwamoto Y. Diagnosis and treatment of Ewing's sarcoma. *Jpn J Clin Oncol* 2007; 37: 79-89.