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Case Report

A large and metastatic primitive neuroectodermal tumor of the kidney

Böbreğin büyük ve metastatik primitif nöroektodermal tümörü

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ABSTRACT

Primitive neuroectodermal tumor (PNET) of the kidney is rare. Mostly patients with renal PNET are young adults and the survival rates are poor. Although radiological and pathological investigations, differential diagnosis from other kidney tumours is very difficult. The treatment is often delayed because of difficulties with diagnosis. In most cases of renal PNET, as in this case, prognosis is poor. Particularly, in young adults with large renal masses, it must be diagnosed and treatment should be started immediately.

Keywords: Kidney; primitive neuroectodermal tumor; survival rate.

ÖZ

Böbreğin primitif nöroektodermal tümörü (PNET) nadirdir. Renal PNET hastalarının çoğu genç erişkindir ve sağkalım oranları kötüdür. Radyolojik ve patolojik incelemelere rağmen, diğer böbrek tümörlerinden ayırıcı tanısı çok zordur. Tanı esnasındaki zorluklar nedeniyle tedavi çoğu zaman gecikir. Bu vakada da olduğu gibi renal PNET olgularının çoğunda prognoz kötüdür. Özellikle büyük renal kitle saptanan genç erişkinlerde tanı hemen koyulmalı, tedaviye mümkün olduğunca erken başlanmalıdır.

Anahtar kelimeler: Böbrek; primitif nöroektodermal tümör; sağkalım oranı.

Introduction

Primitive neuroectodermal tumor (PNET) is thought to arise from primitive cells of neural crest and commonly involves the bone and soft tissue of adolescents.[1] PNET of the kidney is rare, and the differential diagnosis from other malignancies of the kidneys is crucial for management. The patients are mostly young adults, and the clinical course is very aggresive.[2] We aimed to present a case of renal PNET admitted to our clinic with metastatic disease.

Case presentation

A 35-year-old male was admitted with complaints of back pain for 1 month. On clinical examination, a large fixed abdominal mass was palpable. Ultrasound showed a 15 × 13 cm sized lesion on the right kidney. Computed tomography (CT) scan showed a 150 × 145 × 115 mm sized right renal mass, invasive to Gerota's fascia and perirenal fat tissue. Multiple metastatic lymph nodes in the para-aortic, interaortocaval, and paracaval area (largest, 30 × 21 mm in size) was noted (Figure 1). MRI was performed for a better assessment of tumor thrombus, which showed no tumor trombus. Laboratory examinations indicated an increase in the liver function tests. The preoperative diagnosis of the patient was renal cell carcinoma (RCC). Radical nephrectomy and retroperitoneal lymph node excision was performed.

Macroscopically, right radical nephrectomy material of the size of 22 × 18 × 11 cm and weighing 1.77 kg was excised. The tumor was invasive to Gerota's fascia, renal pelvis, perirenal fat tissue, and surrenal gland. Histologically, the tumor cells are uniform, small round blue cells with scant cytoplasm, uniform nuclei, fine chromatin, indistinct cytoplasmic membranes, and were arranged in solid sheets. Mitotic activity (20 mitos/10 HF) and an extensive zone of necrosis were observed. A panel of immunohistochemical markers was used, which included neuron-specific enolase, synaptophysin, chromogranin-A, CD56, CD45, CD3, CD20, Bcl-2, CDX2, TTF-1, CD99, Fli-1, vimentin, and PANCK.

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The tumour cells were positive for synaptophysin, CD99, and Fli-1, whereas they were negative for the other markers. Tumors have high (approximately 85%) Ki-67 indices (Figure 2).

Bone scintigraphy was performed, and multiple bone metastasis that includes cranium, bilateral femur, and pelvic bones were detected. Medical oncology and radiation oncology consultations for final treatment suggested radiotherapy to cranium and pelvis at 300 cGy/day for palliative purposes. The patient died on the fourth day of radiotherapy.

The required consent for publishing this case was obtained from the patient's relatives because patient died.

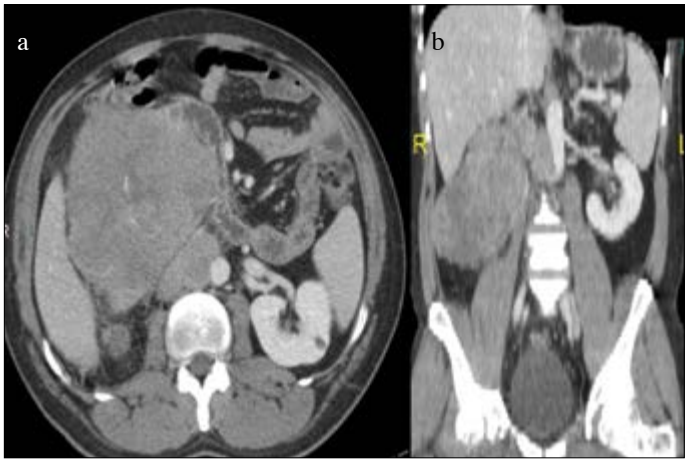

Figure 1. a, b. Right renal mass and metastatic lymph nodes in the interaortocaval and paracaval area on computed tomography [transverse plane (a) and frontal plane (b)]

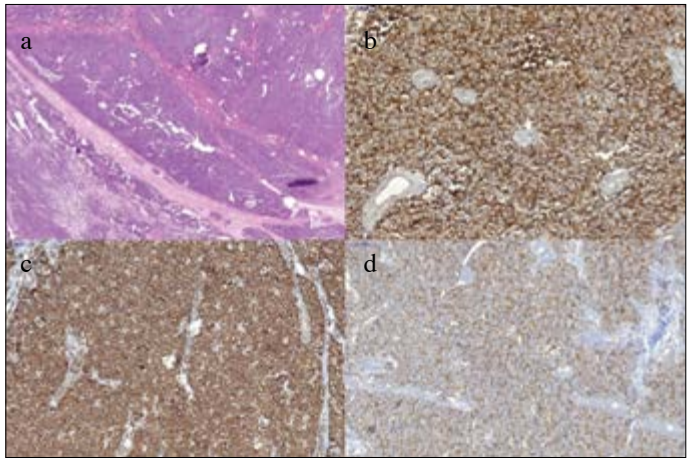

Figure 2. a-d. The tumor cells were arranged in solid sheets (a). The tumor cells showed strong positive expression of CD99 (b). The tumor cells showed strong positive expression of Fli-1 (c). The tumor cells showed strong positive expression of Synaptophysin (d)

Discussion

PNET is related to the Ewing sarcoma family of tumors typically manifesting in the bone or soft tissues of extremities, trunk, head, and neck, but it can also rarely manifest in the viscera or kidneys.[3] Primary renal PNET is a rare condition. Renal PNET can manifest in a wide age range; however, most of the patients are observed in a range of 10–39 years.[4]

Ellinger et al.[4] reported that patients with renal PNET complained about pain (85%), palpable masses (60%), and hematuria (37%), similar to a classic triad of RCC. The advanced disease rate was 57.6% and the most frequent metastases were lymph node (25%), lung (20%), and liver (14%). In another study, Yuvaraja et al.[2] reported that the most common symptoms are abdominal pain (68%), abdominal mass (37%), and hematuria (31%) in their case series of 16 patients. In this case series, 10 patients (63%) had localized disease, 5 (31%) had metastasis (2 lungs, 1 lung and lymph nodes, 1 lymph nodes, and 1 liver), and 1 (6%) had locally advanced disease. Three- and 5-year overall survival rates were 60% and 42%, respectively.

Despite recent improvements in imaging modalities, there are no characteristic radiologic properties detectable for renal PNET. Imaging of renal PNET is generally non-specific and typically reveals a large mass with necrotic and hemorrhagic areas partially or completely replacing the kidney. It typically shows a weak, heterogeneous enhancement on CT and MRI with intravenous contrast.

Differential diagnosis from other renal masses is challenging. Because renal PNET includes Blastemal Wilm's tumor and other small blue round cell tumors such as neuroblastoma, rhabdomyosarcoma, small cell carcinoma, poorly differentiated synovial sarcoma, desmoplastic small round cell tumor, nephroblastoma, and lymphoma, diagnosis of renal PNET is therefore based on histopathology, immunohistochemistry, and cytogenetic studies.[2] On microscopic examination, renal PNET typically shows small round cells that may form characteristic Homer–Wright rosettes. CD99 expression is not pathognomonic for PNET, although overexpression of the surface membrane protein CD99 is almost always observed in these tumors. Because it is also found in synovial sarcomas and gastrointestinal stroma tumors,[5] we therefore need other markers such as synaptophysin and Fli-1 for differential diagnosis.

The tumour in the kidney is no different than the more common counterpart in soft tissues. The cells are relatively monotonous polygonal cells and have a hyperchromatic rounded nucleus. A finely dispersed chromatin and a micronucleolus in some cases are the nuclear characteristics. Mitotic figures may be numerous. Although the nuclear to cytoplasmic ratio is high, a rim of clear cytoplasma and discrete cell membranes are often apparent in well-fixed tumours without extensive degenerative

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changes. The presence of clear cytoplasm is often associated with abundant glycogen as demonstrated by diastase sensitive PAS-positivity.

Renal PNET is a rare entity presented with an agressive clinical behavior. Despite multimodal treatment protocols combining surgery, chemotherapy, and radiotherapy to the renal bed, prognosis is poor with a 5-year disease-free survival rate of 45%–55%.[2,4] Before the routine use of adjuvant chemotherapy, the 5-year survival rate in patients with Ewing family tumors were <10%.[2] Unfortunately, our patient died in the early postoperative period because of an advanced disease. Standard protocol includes vincristine, doxorubicin, and cyclophosphamide alternating with ifosfamide and etoposide. Postoperative radiotherapy must be added in the case of inadequate surgical margins.

In conclusion, the prognosis and survival rate are poor, despite multimodal treatment protocols combining surgery, chemotherapy, and radiotherapy to the renal bed. Immunohistopathological and cytogenetic studies are very important in the diagnosis of renal PNET because of the similarity of radiological finding on RCC. Particularly, in young patients with large renal masses, this rare entity should be taken in to consideration in the differential diagnosis and treatment should be started immediately.

Informed Consent: The required consent for publishing this case was obtained from the patient's relatives due to patient had died.

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References

1. Rodriguez-Galindo C, Marina NM, Fletcher BD, Parham DM, Bodner SM, Meyer WH. Is primitive neuroectodermal tumor of the kidney a distinct entity? Cancer 1997;79:2243-50. [CrossRef]
2. Thyavihally YB, Tongaonkar HB, Gupta S, Kurkure PA, Amare P, Muckaden MA, et al. Primitive neuroectodermal tumor of the kidney: a single institute series of 16 patients. Urology 2008;71:292-6. [CrossRef]
3. Maly B, Maly A, Reinhartz T, Sherman Y. Primitive neuroectodermal tumor of the kidney. Report of a case initially diagnosed by fine needle aspiration cytology. Acta Cytol 2004;48:264-8. [CrossRef]
4. Ellinger J, Bastian PJ, Hauser S, Biermann K, Müller SC. Primitive neuroectodermal tumor: rare, highly aggressive differential diagnosis in urologic malignancies. Urology 2006;68:257-62. [CrossRef]
5. Perlman EJ, Dickman PS, Askin FB, Grier HE, Miser JS, Link MP. Ewing's sarcoma-routine diagnostic utilization of MIC2 analysis: a Pediatric Oncology Group/Children's Cancer Group Intergroup Study. Hum Pathol 1994;25:304-7. [CrossRef]