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Case Series

Renal primitive neuroectodermal tumor. The first case series from Syria

Ahmad Al Mousa [a], Mohammad Nour Kitaz [b], Muhamad Zakaria Brimo Alsaman [c,*], Vairy Rezkallah [d], Lina Ghabreau [d], Ibrahim Al-Hadid [a]

[a] Department of Urology, Aleppo University Hospital, Aleppo, Syria
[b] Neurosurgery Department, Aleppo University Hospital, Aleppo, Syria
[c] Faculty of Medicine, University of Aleppo, Aleppo, Syria
[d] Department of Pathology, Aleppo University Hospital, Aleppo, Syria

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ABSTRACT

Primitive neuroectodermal tumor (PNET) mainly arises from soft tissues of the extremities such as humerus, femur, C tibia. It rarely arises from kidney; less than 200 cases have been reported in the literature. The clinical presentation and radiography findings are not specific. Here we first report two cases of renal primitive neuroectodermal tumor in Syria.

the first patient was 26-year-old- female that presented to urology clinic complaining of right flank pain. Ultrasonography of the abdomen showed a large mixed heterogeneous mass in the right kidney with no hemorrhage or calcification and MSCT of abdomen and pelvis demonstrate a mixed well-demarcated heterogeneous mass measuring (74*117) mm in the right kidney right radical nephrectomy was performed.

The second patient 19-year-old-male presented with left flank pain. Ultrasonography of the abdomen showed mixed large mass involving the left kidney, with unmarked border. The CT of the abdomen and pelvis demonstrating a (30*110*90) mm left renal mass and periaortic lymphadenopathy measuring (45*28) mm. The patient underwent Left radical nephrectomy with periaortic lymphadenectomy dissection. The final diagnosis for both cases was Renal PNET based on microscopic and immunohistochemistry examination.

In patient with suspected renal mass in the radiographic images, the diagnosis of renal primitive neuroectodermal tumor should be kept in the mind despite its rarity. The final diagnosis is done by histopathological study in association with immunohistochemical examination.

1. Background

Primitive neuroectodermal tumors mainly occur in the soft tissues of the extremities such as humerus, femur, C tibia [1]. However, it can be found in other sites such as bladder, prostate, testis, ovary, uterus and less common in the kidney [2].

Renal PNET accounting only for 1% of sarcoma and there are less than 200 cases have been reported in the literature. They are aggressive neoplasias with poor prognosis and their metastases emerge early to the lung, bone and lymph node [3]. The clinical presentations of renal PNET is similar to other renal tumors, so the diagnosis is made by histopathological study in association with immunohistochemical (IHC) examination [4]. Here we will first report two cases of renal PNET in Syria using PROCESS Guideline 2020 [5].

2. Case presentation

2.1. Case 1

A 26-year-old-rural female, presented to urology clinic complaining of right flank pain, anorexia and fever from one week ago. The patient was non-smoker and non-alcoholic with body mass index (BMI) 20.2Kg/m2. Tonsillectomy in her past surgical history. Physical examination revealed soft abdomen, no flank or abdominal mass palpable. Ultrasonography of the abdomen showed a large mixed heterogeneous mass in the right kidney with no hemorrhage or calcification. The patient underwent MSCT of abdomen and pelvis with and without intravenous contrast to study the renal tumor and to investigate presence of regional lymph node hypertrophy, MSCT demonstrate a mixed well-demarcated heterogeneous mass measuring (74*117) mm in the right kidney that

* Corresponding author.
E-mail addresses: ahmad.almousa@gmail.com (A. Al Mousa), mohnour97ki@gmail.com (M.N. Kitaz), dr.zakariaBrimo@gmail.com (M.Z. Brimo Alsaman), vairygr1993@gmail.com (V. Rezkallah), Ibrahimalhadid@alepuniv.edu.sy (I. Al-Hadid).

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has poor measurable contrast enhancement with no regional lymphadenopathy. In addition, there were no evidence of distant metastases on chest CT scan. Urine analysis showed microscopic hematuria (120_150) RBC in HPF. Right radical nephrectomy was performed.

The diagnosis was primitive neuroectodermal tumor based on microscopic (Fig. 1, A) and immunohistochemistry examination (Fig. 1, B).

Tumor stage was (T3a, N0, M0) and group-staging 3 according to TNM 2017. The patient deteriorated after surgery and she died.

2.2. Case 2

A 19-year-old-male presented to urology department with left flank pain, fever and weight loss from 10 day ago. The patient was a farmer that lived in a rural area, with body mass index (BMI) 23.4 kg/m2. Ultrasonography of the abdomen showed mixed large mass involving the left kidney, with unmarked border. The patient underwent multiphasic renal mass protocol CT of the abdomen and pelvis demonstrating a (30*110*90) mm left renal mass and periaortic lymphadenopathy measuring (45*28) mm. In addition, there were metastases to both navel lung on chest CT scan. He was smoker; packet 10 year, with negative familial history for renal malignancies and no past medical or surgical history. Laboratory values were as follows: Hb: 12.2 g/dl– WBC: 9.1 (10^3/mm^3) _ Palettes: 90 (10^3/mm^3) _ K: 3.6 (mEq/l) _ Na: 132 (mEq/l) _ Cr: 0.1 (mg-100 m) _ Urea: 40 (mg-100 m) _ Ca: 8.8 (mEq/l). Urine analysis: RBC: 10 _ WBC: 3.

Left radical nephrectomy with periaortic lymphadenectomy dissection has been done.

The diagnosis was primitive neuroectodermal tumor based on microscopic (Fig. 2, A)and immunohistochemistry examination.

Immunohistochemical stains: CD99: positive, CK, LCA: negative (Fig. 2, B).

Tumor stage was (T3a,N1,M1) and group staging 3 according to TNM 2017.He died two weeks following surgery.

3. Discussion

Seemayer et al. first described the Primitive neuroectodermal tumor (PENT)/Ewing sarcoma (ES) of kidney in 1975 [6]. Because PENT tumor is difficult to differentiate from an extra skeletal ES, the actual number of reported cases is still unknown [7]. Here we will first discuss two rare cases of renal PNET in Syria.

The clinical signs and symptoms are not specific, but the most common symptom is acute flank pain (73%), that mimics renal stone colic [6]. Fever, weight loss, bone pain, palpable abdominal or flank mass may be presented [8]. In our cases the main symptoms were acute flank pain and fever.

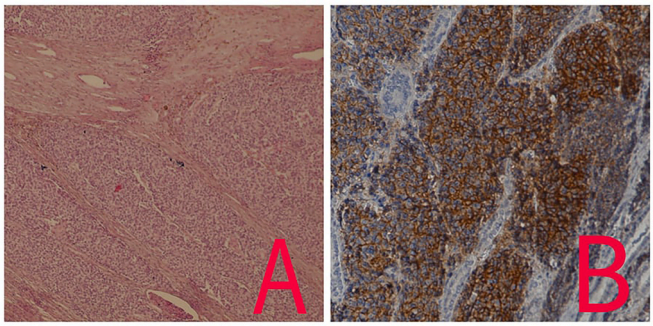

Fig. 1. A: Histological examination revealed solid sheets, cords and trabeculae infiltrating the renal parenchyma. The tumor was composed of small round blue cells with scant cytoplasm, uniform nuclei and stippled chromatin. Mitosis was numerous.
B: Immunohistochemical stains: CD99: positive/CK, LCA: negative. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

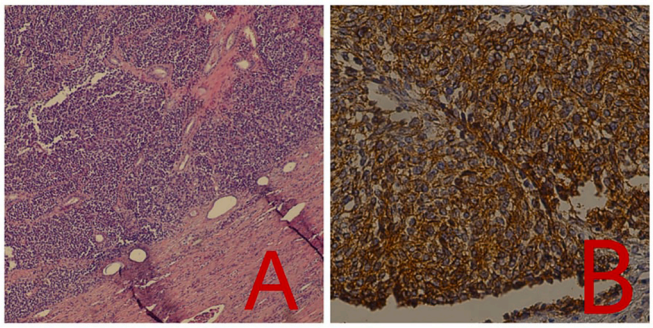

Fig. 2. A: Histological examination revealed solid sheets, cords and trabeculae infiltrating the renal parenchyma. The tumor was composed of small round blue cells with scant cytoplasm, uniform nuclei and stippled chromatin. Mitosis was numerous.
B: Immunohistochemical stains: CD99: positive/ CK, LCA: negative. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

EWS/FLI-1 gene fusions in renal ES/PNET is the gold standard of the diagnosis, which is the same gene in the extrarenal tumors [4], the cytogenetic studies showed that the translocation of long arms of chromosome 11 and 22, t (11; 22) (q22; q12) in 90% of PNET/ES cases [9].

Pathologically, we can see small round blue cells that involve necrotic areas, also we may detect diffuse uniform membranous staining with CD99, vimentin, and CD117 [9].

In general, radiologic findings are not specific, so histologic and immunohistochemistry studies still consider for diagnosis [10].

PENT/ES should be considered as differential diagnosis when advance renal tumor presents especially in young patients [11].

The combination of surgery, chemotherapy, and radiotherapy is considered for renal PENT. The neoadjuvant chemotherapy (IVAD regimen: Adriamycin, vincristine, ifosfamide with mesna; four cycle) is recommended in locally advanced, inoperable renal PNET [12].

Others suggested that the best choice for long-term survival is the chemotherapy after the surgery [13].

4. Conclusion

Primitive neuroectodermal tumor is a rare localized to the kidney. It is aggressive neoplasia with poor prognosis and their metastases emerge early. In patient with suspected renal mass in the radiographic images, the diagnosis of renal primitive neuroectodermal tumor should be kept in the mind despite its rarity. The final diagnosis is done by histopathological study in association with immunohistochemical examination.

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Author contributions

AM & IA: managed the patients and did the surgery.

MZBA & MNK: design of the study, revising critically, wrote the manuscript, and analysis.

VR & LG: Interpretation and providing pathological data.

All authors read and approved the final version of manuscript.

Registration of research studies

1. Name of the registry: not applicable
2. Unique Identifying number or registration ID: not applicable
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Guarantor

Ahmad Al Mousa.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2022.104740.

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