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Central nervous system involvement of primary renal lymphoma with diffuse large B-cell type lymphoma

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Manuscript Preparation E
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Patient: Female, 27
Final Diagnosis: Central nervous system leukemia
Symptoms: Abdominal mass • fever • nausea • vomiting
Medication: —
Clinical Procedure: —
Specialty: Hematology

Objective: Unusual clinical course

Background: Primary lymphoma of urinary system is an uncommon entity including primary renal lymphoma, primary ureter lymphoma, bladder lymphoma, and urethra (including prostate) lymphoma. The pathology of PRL is usually diffuse large B-cell lymphoma.

Case Report: In this study we describe a 27-year-old Chinese woman with central nervous system leukemia due to PRL. It was classified as diffuse large B-cell lymphoma non-GCB type according to kidney biopsy. We selected R-CHOP as a regimen and it showed good curative effect. However, the patient suffered recurrence involving the central nervous system with complaint of intolerable iliac-sacral pain at 26 days after 6 courses of R-CHOP.

Conclusions: This case shows that prognosis of PRL is poorer than nodal lymphoma. There is an urgent need for a more effective strategy with higher blood-brain barrier permeability.

Key words: lymphoma • diffuse large B-cell type • primary renal lymphoma • central nervous system involvement

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Background

Primary lymphoma of urinary system is an uncommon entity caused by absence or inadequate amounts of lymphatic tissues in the urinary tract [1]. It may misdiagnosed as kidney cancer and include primary renal lymphoma (PRL), primary ureter lymphoma, bladder lymphoma, and urethra (including prostate) lymphoma. Both male and female patients can suffer from it. The accurate mechanism is unclear. PRL does not show specific manifestations. The common symptoms include uncertain abdominal pain, fever, night sweats, weight loss, anemia, hematuria, pyuria, and azotemia. The signs include hypertension and abdominal mass. The pathology of PRL is usually diffuse large B-cell lymphoma (DLBCL) [2].

Case Report

Here we describe a case of central nervous system (CNS) leukemia due to PRL. To our knowledge, this is a rare case.

A 27-year-old Chinese woman was admitted with nausea, vomiting, and fever of 2 week duration.

She denied night sweats and weight loss. Two weeks before the present admission, she had nausea, vomiting, and fever. The highest temperature was 38.4°C. At the same time, whole-body edema and reduced urine volume with 600 ml per day occurred. On examination, she had a 5-cm tough mass in the lower left abdomen, with tenderness and good movement and no lymphadenopathy.

Serum lactate dehydrogenase was 644 U/L (125–243 U/L). Serum β 2 micro-globulin was 17.9/L (0.7–1.8 mg/L). CA125 was 35.42 U/mL (0–35 U/mL). Complete blood count was normal. Urea was 14.66 mmol/L (2.85–7.14 mmol/L), creatinine was 458 μ mol/L (44.2–88.4 μ mol/L), and uric acid was 549 μ mol/L (142–420 μ mol/L). Bone marrow cell analysis was normal. The PET-CT showed diffuse bilateral kidney enlargement and elevated 18-FDG activity (Figure 1). Kidney biopsy showed DLBCL with CK(-), CD3 scatter(+), CD20 diffuse(+), MPO(-), CD34(-), Vimentin(-), HMB45(-), CD79a(+), MUM1(+), Bcl-6(-), and CD10(-). It was classified as diffuse large B-cell lymphoma non-GCB type. The proliferation fraction as detected by Ki67 was 60%+.

This profile assessed the diagnosis of stage III according to the Ann Arbor system. The international prognosis index (IPI) was 3 scores. Chemotherapy was administrated, consisting of 6 courses R-CHOP regimen (Rituximab 375 mg/m², cyclophosphamide 750 mg/m², doxorubicin 50 mg/m², vincristine 1.4 mg/m², and dexamethasone 10 mg every 21 days) [3]. A CT after 6 courses of treatment showed a complete response and the patient showed no discomfort.

Discussion

Stallone et al developed 3 criteria to differentiate primary renal lymphoma from renal involvement of systemic NHL [4]: (1) lymphomatous renal infiltration, (2) non-obstructive uni- or bilateral kidney enlargement, and (3) no extra-nodal localization at the time of diagnosis.

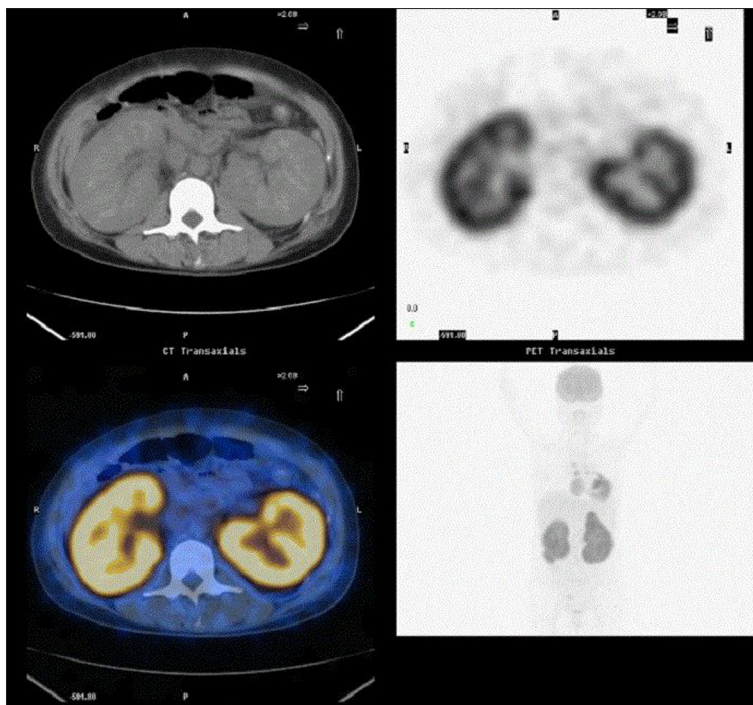


Figure 1. The PET-CT showed diffuse largeness of bilateral kidney and elevated 18-FDG activity.

Prognosis of PRL is usually poorer than nodal lymphoma, not only due to inaccurate or delayed diagnosis, but also due to its high invasiveness. There is no standard treatment protocol for PRL due to its rarity. According to its histology showing CD20(+), we selected R-CHOP as a regimen and it showed good curative effect. This case demonstrates that Rituximab possesses good permeability in renal tissues and may be a better choice than traditional chemotherapy regimen.

However, the patient had a recurrence involving the CNS with complaint of severe iliac-sacral pain at 26 days after 6-course administration of R-CHOP. Lumbar vertebra showed lumbar iliac herniation from L4 to S1 and enlargement of the cauda equine. The patient received lumbar puncture and intrathecal chemotherapy (Cytosar 50 mg, dexamethasone 5 mg, methotrexate 10 mg) of 4 courses. Leukemia cells disappeared after 2 courses and the Pandy test was negative after 4 courses. Six more courses of intrathecal chemotherapy (2 courses/week) were implemented for further remission. The patient was undergoing regular follow-up. One year after the onset, the patient progressed to lymphoma leukemia and has achieved remission after VDLP (vincristine 2 mg at day 1, 8, 15, and 21, doxorubicin 40 mg/m² at day 1–3 and 16–18, L-asparagine

1000 U/d at day 19–28, dexamethasone 10 mg at day 1–14, and 5 mg at 15–28) regimen.

Conclusions

This case shows that prognosis of PRL is poorer than nodal lymphoma. Although Rituximab can salvage the patient from lymphoma, she had CNS leukemia and lymphoma. NNCN guidelines recommend regular intrathecal chemotherapy for lymphoma patients with external node involvement. However, new clinical research showed both Rituximab and intrathecal chemotherapy cannot prevent CNS involvement in a lymphoma patient [5]. A more effective strategy with higher blood-brain barrier permeability is urgently needed.

Conflict of interest

All the authors declare no conflict of interest.

Acknowledgments

None.

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