Case Report

Primary renal lymphoma: a case report and review of the literature

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Abstract: Although renal involvement is common in non-Hodgkin's lymphoma (NHL), primary renal NHL is a rare disease. Diffuse large B-cell lymphoma (DLBCL) is the most common subtype of NHL and is responsible for 30–40% of adult NHL cases globally. Here, we present a case of a 68-year-old man with right flank pain who underwent retroperitoneal laparoscopic radical nephrectomy for an enlarged right kidney, with evidence of an infiltrating mass. Postoperative histopathological examination revealed primary renal DLBCL. Primary renal lymphoma (PRL) is quite rare, and clinicians should try to ensure early diagnosis and treatment with standardized and systematic chemotherapy.

Keywords: Primary renal lymphoma (PRL); non-Hodgkin's lymphoma (NHL); diffuse large B-cell lymphoma (DLBCL); case report

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Introduction

Diffuse large B-cell lymphoma (DLBCL) is the most common subtype of non-Hodgkin's lymphoma (NHL) and is responsible for 30–40% of adult NHL cases globally, according to the World Health Organization's (WHO) classification of neoplastic diseases of the hematological and lymphatic systems (1). This tumor is clinically, morphologically, and genetically heterogeneous. At present, the advent of genomic technologies has allowed molecular subtyping of this heterogeneous disease process in both human and dogs (2-4). Although renal involvement has been reported in 30–60% of all patients diagnosed with NHL (5), primary renal lymphoma (PRL) is a very rare disease and is often mistaken for renal cell carcinoma (RCC). Occasionally, patients with PRL present with nonspecific symptoms and signs, as well as flank pain, weight loss, fever, hematuria, and a palpable mass (6). In addition, the B-cell immunophenotype of PRL is a common pathological type (7), and so far, more than 50 cases have been reported in the literature.

Here, we report a 68-year-old man who presented with right flank pain and an approximately 9 cm solid mass in the right kidney on computed tomography (CT). After retroperitoneal laparoscopic radical nephrectomy, postoperative histopathological examination revealed a primary renal DLBCL.

Case presentation

A 68-year-old Chinese man visited the local hospital because of persistent pain on the right side of his waist. The pain lasted for half a year and recently worsened. He underwent an enhanced CT examination (*Figure 1*) in the local hospital, and a mass (8.5×6.8 cm in diameter) was noted in the right kidney. He was then transferred to our hospital for surgical treatment. During the course of the disease, he did not lose weight. He had a history of hypertension, and he had undergone cholecystectomy in 2008. He denied any history of familial infective or genetic disease.

His laboratory results were as follows: blood glucose level, 6.2 mmol/L (reference range, 3.9–6.10 mmol/L); lactate dehydrogenase level, 305 U/L (reference range, 140–271 U/L); serum creatinine level, 72.0 μmol/L (reference range, 44.0–133.0 μmol/L); albumin level, 37.7 g/L (reference range, 40.0–55.0 g/L); erythrocyte

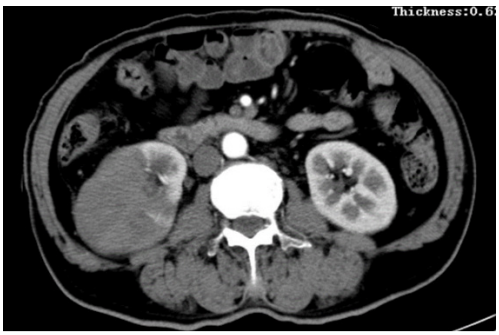

Figure 1 A contrast-enhanced CT finding at the disease onset shows a huge mass (8.5×6.8 cm in diameter) of the right kidney.

sedimentation rate, 32.00 mm/h (reference range, 0–15 mm/h); leukocyte count, 5.07×10^9/L (reference range, 3.50×10^9–9.50×10^9/L); erythrocyte count, 3.85×10^{12}/L (reference range, 4.30×10^{12}–5.80×10^{12}/L); hemoglobin level, 112 g/L (reference range, 130–175 g/L). Routine urine and stool tests were normal.

He underwent radical nephrectomy and lymph node dissection. Postoperative immunohistochemical staining (*Figure 2*) showed DLBCL with the following results: CD3(-), CD20(+++), Pax-5(+++), Ki-67(>80%,+), CD10(+), Bcl6(+), MUM1(+), Bcl2(+), CMyc(+), CD30(-), CK(-), CD21(-), CD5(-), EMA(-). He was considered to

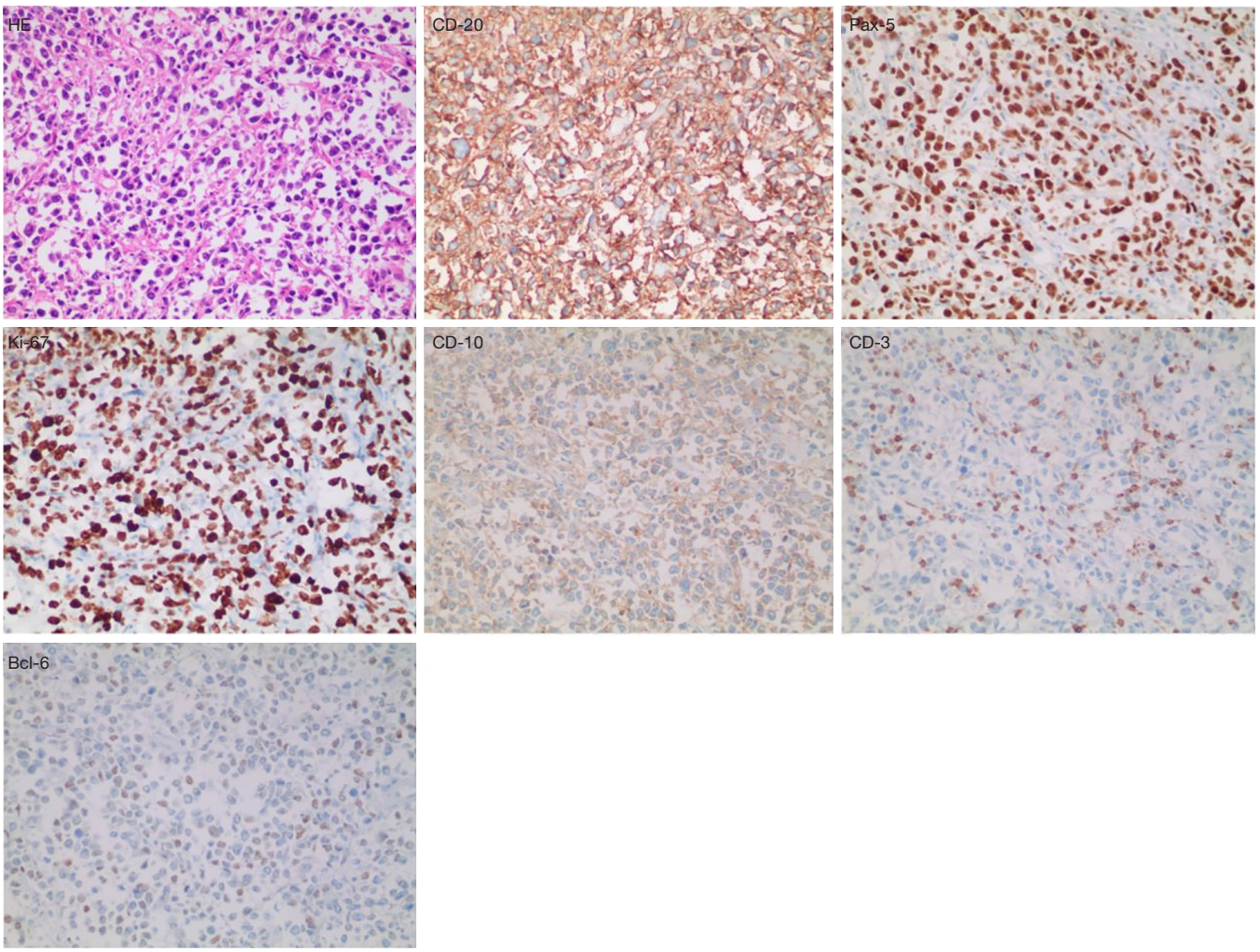

Figure 2 Immunohistochemistry staining of the resected right renal kidney. The antibodies used were as listed in the right below boxes of each representative graph (×200).

have DLBCL (origin at the germinal center). Postoperative pathology and immunohistochemistry indicated DLBCL, which only localized in the kidney. Bone marrow biopsy and CT of the chest region did not show any evidence of lymphoma invasion. According to the findings, he was diagnosed with PRL. After operation the patient received the R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone) regimen for eight standard courses, and achieved remission to a large extent. Follow-up results showed no recurrence and metastasis. Usually, PRL patients show improved outcome after the addition of rituximab to the CHOP regimen (8).

Discussion

It is common for NHL to occur in the kidney. However, PRL is an argumentative and rare disease because the renal parenchyma lacks lymphatic vessels (7). Several cases have been reported in the literature, but consistent diagnostic criteria of PRL have not been established. Stallone *et al.* assessed 29 cases of PRL that satisfied the following three diagnostic criteria: (I) lymphoma renal infiltration; (II) non-obstructive single or bilateral kidney enlargement; (III) no extra renal localization at the time of diagnosis (9).

As shown in *Table 1*, a total of 46 cases of PRL (10-12) have been reported from 1990 until now. Among these cases, nearly 90% involved patients who were aged more than 50 years. The male-to-female ratio was about 2:1, indicating that this disease is more common in men. Additionally, the proportion of patients with unilateral renal involvement was higher than the proportion of patients with bilateral renal involvement. The most common histology type of PRL was DLBCL and the second most common was marginal zone lymphoma (MZL). The most

Table 1 Case reports of primary renal lymphoma since 1990

Gender	Age	Symptoms	Surgical treatment	Pathology	Chemotherapy	Prognosis
M	75	High fever	Nephrectomy	DLBCL	Chemo	Dead
M	22	Edema	No	DLBCL	EPOCH	Remission
M	84	Undefined	No	DLBCL	R-CHOP	Remission-recurrence
M	56	Pain, fever	Nephrectomy	DLBCL	R-chemo	Remission
F	82	Undefined	Nephrectomy	MZL	R-chemo	Remission
M	42	Pain	No	DLBCL	R-chemo	Remission
M	39	Pain	Nephrectomy	NHL	Chemo	Remission
M	77	Hematuria	No	MZL	Radiation	Remission
M	78	Pain	Nephrectomy	DLBCL	Chemo	Remission
M	73	Undefined	No	DLBCL	Undefined	Undefined
M	77	Malaise	Nephrectomy	DLBCL	R-chemo	Remission
F	57	Pain, fever	No	B cell	CHOP	Undefined
F	62	Pain	Nephrectomy	DLBCL	R-chemo	Remission-recurrence
M	58	Headache	Nephrectomy	DLBCL	R-chemo	Undefined
M	83	Undefined	No	MZL	Chemo	Remission
M	66	Undefined	Nephrectomy	MZL	No	Remission
F	75	Undefined	Nephrectomy	MZL	Chemo	Remission
M	54	Undefined	No	MZL	Chemo	Remission
M	65	Undefined	No	MZL	No	Remission
F	77	Undefined	No	MZL	Undefined	Remission

Table 1 (*continued*)

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Gender	Age	Symptoms	Surgical treatment	Pathology	Chemotherapy	Prognosis
M	21	Pain, fever	No	DLBCL	Chemo	Remission
Undefined	77	Undefined	No	DLBCL	R-chemo	Remission
F	71	Pain	Nephrectomy	DLBCL	CHOP	PD
M	68	Fever	No	DLBCL	No	Dead
M	53	ARF	Nephrectomy	MZL	No	Remission
F	83	Pain, fever	No	MZL	R-chemo	Remission
M	82	Pain	No	DLBCL	Chemo	Undefined
M	45	Pain, fever	Nephrectomy	MZL	Chemo	Remission-recurrence
M	62	Hematuria	No	FL	Chemo	Dead
M	77	Undefined	Nephrectomy	MZL	No	Remission
M	50	Undefined	Nephrectomy	MZL	No	Remission
F	51	Pain	Nephrectomy	SNC	Chemo	PD
F	70	Fever	Nephrectomy	DLBCL	R-chemo	Remission
F	27	Fever	No	DLBCL	R-chemo	Remission
M	61	Undefined	Nephrectomy	DLBCL	Chemo	Undefined
F	63	Pain	No	DLBCL	Chemo	PD
M	60	Pain	Nephrectomy	DLBCL	Chemo	Remission-recurrence
F	60	Pain	Nephrectomy	DLBCL	Chemo	Remission
M	59	Pain	Nephrectomy	SNC	Chemo	Remission
F	52	Pain	Nephrectomy	DLBCL	Chemo	Remission-recurrence
M*	68	Pain	Nephrectomy	DLBCL	R-CHOP	Remission
M	78	Pain	Nephrectomy	DLBCL	No	Dead
M	4	Fatigue	No	DLBCL	CHOP	Remission
M	37	Pain, distention	No	DLBCL	No	Dead
M	51	Pain	Nephrectomy	NHL	Chemo	Remission
M	13	Fatigue, arthralgia	No	T lymphoblastic lymphoma	Chemo	Dead

*, present case. M, male; F, female; DLBCL, diffuse large B cell lymphoma; MZL, marginal zone lymphoma; SNC, small non-cleaved cell lymphoma; NHL, non-Hodgkin lymphoma; R, rituximab; PD, progress of disease; ARF, acute renal failure.

common symptoms of PRL were pain and fever, and the condition of the majority of patients showed remission to some extent after chemotherapy. Compared with other urological tumors, PRL has some unique imaging features. Usually plain CT scan shows that primary renal DLBCL is homogeneous or slightly low-density lump, with unclear border. While enhanced CT shows mild enhancement, and the degree of enhancement is lower than that of normal renal parenchyma. In addition, PRL often lacks a blood supply and rarely invades the inferior vena cava as shown by CT. Moreover, the center of the PRL tumor is outside the renal collection system. These characteristics can help differentiate from other urologic tumors.

In 2008, Ladha *et al.* reported that a 62-year-old man who presented with right flank pain and a right renal mass on CT left against medical advice and underwent

nephrectomy elsewhere. Histopathology revealed DLBCL. Although it is a rare disorder, overlooking PRL as one of the causes of a renal mass can lead to disseminated disease and unnecessary nephrectomy (5). In 2016, Fujizuka *et al.* reported that a 75-year-old man presenting with high fever and swelling of the left kidney was initially treated for severe pyelonephritis and the pathological diagnosis after left kidney nephrectomy was primary renal DLBCL (12). In view of the aggressive nature and poor prognosis of this condition, early diagnosis and timely treatment is essential. Currently, many examinations, including bone marrow biopsy, renal biopsy, immunohistochemistry, thoracoabdominal CT (9), and PET/CT (13), can help in the diagnosis of PRL. Although patients with PRL have poor prognosis, combined and intensive therapy, such as chemotherapy followed by radiation, may increase survival and improve quality of life (14). Patients who do not receive standard courses of chemotherapy or radiation treatment because of severe complications and other special circumstances may have limited disease free survival. In this case, we present a patient with a unilateral renal mass due to massive lymphomatous infiltration of the right kidney, which was diagnosed as primary renal diffuse large B-cell NHL according to postoperative pathological findings. Clinicians should try to ensure preoperative diagnosis and avoid unnecessary radical diagnostic nephrectomy, as patients with PRL can have a positive outcome after standardized and systematic chemotherapy.

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Footnote

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at http://dx.doi.org/10.21037/acr.2019.12.03). The authors have no conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. The study was approved by the Institutional Review Board of the First Affiliated Hospital of Nanjing Medical University. Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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