




Case Report

Intravascular large B-cell lymphoma in renal cell carcinoma incidentally detected by robot-assisted partial nephrectomy

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Abbreviations & Acronyms

eCT = enhanced computed tomography
 IVLBCL = intravascular large B-cell lymphoma
 RAPN = robot-assisted partial nephrectomy
 RCC = renal cell carcinoma
 R-CHOP = rituximab plus cyclophosphamide, doxorubicin, vincristine, and prednisone

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Introduction: Intravascular large B-cell lymphoma is a rare and aggressive type of extranodal large B-cell lymphoma. Although intravascular large B-cell lymphoma can invade various organs, renal involvement has been rarely reported. Synchronous occurrence of intravascular lymphoma with renal cell carcinoma is extremely rare. We herein report a case of intravascular large B-cell lymphoma in a renal cell carcinoma incidentally detected by robot-assisted partial nephrectomy.

Case presentation: A 69-year-old female with recurrent fever lasting 4 years underwent robot-assisted partial nephrectomy for small renal cell carcinoma. Histological findings led to the diagnosis of intravascular large B-cell lymphoma, which involved the normal tissue of right kidney as well as clear cell renal cell carcinoma. She received six cycles of chemotherapy without major complications and achieved complete remission.

Conclusion: We encountered a rare case of synchronous intravascular lymphoma with renal cell carcinoma.

Key words: intravascular large B-cell lymphoma, renal cell carcinoma, robot-assisted partial nephrectomy.

Keynote message

We report a case of IVLBCL in a renal cell carcinoma. It was incidentally detected by robot-assisted partial nephrectomy. This is the first case of synchronous IVLBCL with renal cell carcinoma detected by robot-assisted partial nephrectomy.

Introduction

IVLBCL is a rare and aggressive type of extranodal large B-cell lymphoma, with an estimated annual incidence of 0.5–1 per 100 thousand population. Most cases occur in adults, with a median age of 67 years. Men and women are equally affected. It often involves the brain and skin, however it can invade almost any organ.¹ Renal involvement of IVLBCL is rarely reported, with <50 previous cases.² Furthermore, the synchronous occurrence of intravascular lymphoma with RCC is extremely rare. There have been only four cases of IVLBCL involving the microvasculature of RCC.^{3–6}

We present the case of a 69-year-old female who underwent RAPN for a clear cell RCC sized 1.7 cm, in which IVLBCL was incidentally detected. To our knowledge, this is the first case of synchronous IVLBCL with RCC detected by RAPN.

Case presentation

A 69-year-old female with a surgical history of maxillary sinus cancer and uterine fibroids visited the Department of Infectious Diseases of a nearby general hospital for examination of severe pitting edema and recurrent fever of 39°C. Although the recurrent fever lasted 4 years, she had not previously undergone detailed examinations.

Infectious diseases or autoimmune diseases were not detected by close examinations. However, eCT revealed an enhanced mass measuring 17 × 17 × 11 mm in her right kidney,

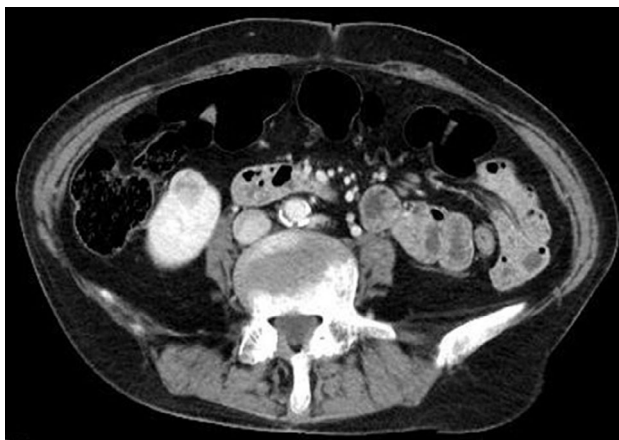


Fig. 1 Computed tomography scan findings of an enhanced mass at the lower pole of her right kidney.

which was suspected to be an RCC (Fig. 1). She also had a right adrenal adenoma (28 × 20 × 21 mm), which was revealed to be nonfunctioning. No metastatic mass was observed, then she was referred to the Urology Department of our hospital for surgical resection of the renal mass. At the time of admission, the physical findings were as follows: height, 148.5 cm; weight, 43.6 kg; body temperature, 37.3°C; blood pressure, 98/73 mmHg; heart rate, 110/min; and oxygen saturation at rest on room air, 98%. Laboratory findings at admission are presented in Table 1.

She underwent RAPN along with right adrenalectomy at the same time. After surgery, she experienced a continuous fever of over 38°C. Therefore, eCT and echocardiography were performed, however there were no abnormal findings.

Two weeks after surgery, histological examination revealed clear cell carcinoma and adrenal adenoma. The RCC was pT1a, G1, INFa, lymphatic invasion (–), vascular invasion (–), resected margin (–). Vessels of clear RCC are packed with atypical lymphoid (Fig. 2a). Atypical cells were positive for CD20 expression. (Fig. 2b) The Ki-67 index increased to

80%. Atypical cells also infiltrated the vessels of the normal tissue of the kidney, however they did not infiltrate the adrenal sinusoid. Histological findings led to the diagnosis of IVLBCL, which involved the right kidney and clear cell RCC.

She underwent bone marrow and skin biopsies to examine the clinical stage of IVLBCL to determine whether IVLBCL invades other sites in her body. Although the bone biopsy did not show evidence of lymphoma invasion or any other hematological abnormalities, including hemophagocytic syndrome, the skin biopsy showed atypical lymphoid cells involving the vessels in subcutaneous tissue. Therefore, she was diagnosed as stage IVB IVLBCL with an International Prognostic Index of 4 (high risk) due to advanced age, clinical stage IV, two extranodal sites, and elevated serum lactate dehydrogenase above the upper limit of normal. Treatment with oral steroids (prednisone, 50 mg/day) was initiated to mitigate the symptoms associated with IVLBCL. Her body temperature turned to be normal, and she was tentatively discharged 1 month postoperatively.

Two months after the surgery, chemotherapy with R-CHOP was initiated. She had received six cycles of R-CHOP without any major complications and achieved complete remission. She has no recurrence of RCC or IVLBCL 1.5-year after RAPN.

Discussion

IVLBCL can invade various organs and manifest diverse symptoms according to the types of organ involved. A definitive diagnosis of IVLBCL requires histopathological examination. Random skin biopsies, including the abundant vascular structures of adipose tissue, could be helpful for diagnosis.

Chemotherapy is the first option of treatment for patients with IVLBCL. As an aggressive form of lymphoma, it has been classically treated with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP regimen). The introduction of the R-CHOP regimen has significantly improved the therapeutic effects.⁷ It has recently been reported that autologous stem cell transplantation following high-dose chemotherapy can improve the outcome of patients with IVLBCL. However, this aggressive treatment approach appears to be practical only in a limited proportion of patients.⁸

The recently updated World Health Organization classification of tumors of hematopoietic and lymphoid tissues has suggested considering variants of IVLBCL according to their clinical characteristics: classical variant, cutaneous variant, and hemophagocytic syndrome-associated variant.¹ The classical variant IVLBCL often presents with various nonspecific symptoms, such as fever, fatigue, and altered consciousness. In cutaneous variant, involved lesions are limited to the skin and disease progressions are less aggressive. The hemophagocytic syndrome-associated variant showed the most rapidly aggressive onset and progression, with a median survival time of 2–8 months. We diagnosed this case as a classical variant due to IVLBCL in her kidney and skin with recurrent fever for a long time, and complete remission was achieved with R-CHOP chemotherapy.

The occurrence of synchronous IVLBCL with RCC is very rare, with only four cases reported to date (Table 2). IVLBCL

Table 1 Laboratory data upon admissions

WBC	8600	/μL	AST	101	IU/L
Neutrocyte	79	%	ALT	52	IU/L
Lymphocyte	5.5	%	ALP	844	IU/L
Monocyte	13.5	%	γGTP	57	IU/L
Eosinophils	0	%	LD	333	IU/L
Basophils	0.5	%	Total bilirubin	0.6	mg/dL
RBC	384	× 10 ⁴ /μL	Total protein	6	g/dL
Hb	10	g/dL	Albumin	2.2	g/dL
Platelets	14.9	× 10 ⁴ /μL	CRP	21.9	mg/dL
Sodium	133	mEq/L	Ferritin	584	ng/dL
Potassium	4.7	mEq/L	PT-INR	1.17	
Chloride	98	mEq/L	APTT	35.6	sec
BUN	15	mg/dL	D-dimer	3.4	ng/dL
Creatinine	0.65	mg/dL	NT-pro BNP	318	pg/mL
			sIL-2R	1810	U/mL

NT-pro BNP, N-terminal pro-brain natriuretic peptide; sIL-2R, soluble interleukin-2 receptor.

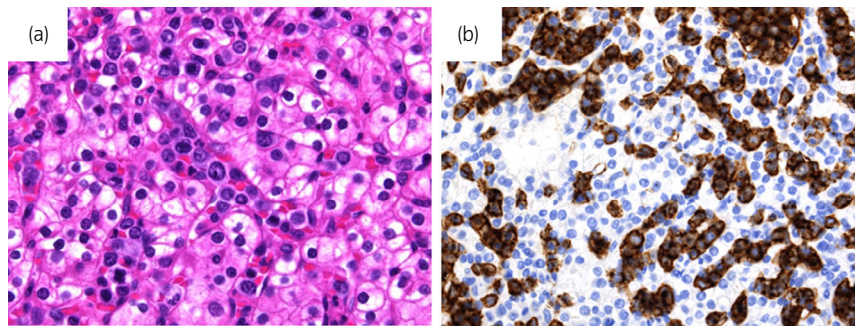


Fig. 2 Histology of the tumors. (a) Hematoxylin and eosin findings: Tumor vessels are packed with atypical lymphoid cells in clear RCC. (b) Immunohistochemistry of CD20: Higher magnification showing lymphoma cells in the vasculature positive for the B-cell marker.

Table 2 Cases of synchronous IVLBCL with RCC

Author	Age (sex)	Symptoms	Stage (RCC)	Invasion of IVLBCL	Treatment	Outcome
Wang <i>et al.</i> ³	77 (F)	Dizziness	pT2a	Skin	Chemotherapy	Died 6 months later
Zhang <i>et al.</i> ⁴	72 (M)	–	Not available	–	Not available	Not available
Kurek <i>et al.</i> ⁵	82 (F)	–	pT3a	–	–	Died 18 months later
Serrano <i>et al.</i> ⁶	72 (M)	Back pain	pT1b	Brain	–	Died 1 month later
Current case	69 (F)	Fever, edema	pT1a	Skin	R-CHOP	No recurrence for 18 months

was incidentally detected by radical nephrectomy in all the four previous cases. The reported prognoses for these cases are poor; they died at 1, 6, and 18 months, respectively.

Our case is unique in two aspects. Firstly, the progression of disease was less aggressive. She had recurrent fever for 4 years before the diagnosis, the clinical response to R-CHOP treatment was excellent without recurrence for 18 months. Secondly, this is the first case of synchronous IVLBCL with RCC detected by RAPN. Compared with the four reported cases, in this case, the renal mass was small and resected with RAPN.

To diagnose IVLBCL at an early stage, it is essential that we suspect lymphoma when facing with constitutional symptoms. It is also important that if we find a patient with a renal mass and recurrent fever or fatigue for a long time, we suspect the possibility of IVLBCL.

Conclusion

We herein report a case of IVLBCL in an incidentally detected RCC by RAPN. To our knowledge, this is the first case of synchronous IVLBCL with RCC detected by RAPN.

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Author Contributions

Michio Noda: Conceptualization; Data curation; Formal analysis; Investigation; Methodology; Writing – original draft. Yutaka Enomoto: Conceptualization; Formal analysis;

Funding acquisition; Investigation; Methodology; Supervision; Writing – review & editing. Yukari Shirasugi: Investigation; Methodology; Writing – review & editing. Sumiyo Ando: Investigation; Methodology; Writing – review & editing. Yukimasa Matsuzawa: Supervision; Writing – review & editing. Haruki Kume: Writing – review & editing.

Conflict of interest

The authors declare no conflict of interest.

Approval of the research protocol by an Institutional Reviewer Board

Not applicable.

Informed consent

Informed consent was obtained from the patient for publishing this case report.

Registry and the Registration No. of the study/trial

Not applicable.

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