Case Report

Primary renal peripheral T-cell lymphoma, not otherwise specified, treated with partial nephrectomy

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Abbreviations & Acronyms
CHOP = cyclophosphamide, doxorubicin, vincristine, and prednisone
COVID-19 = coronavirus infectious disease, emerged in 2019
CT = computed tomography
EBER = Epstein–Barr virus-encoded RNA
FDG-PET = [18F]-2-fluoro-2-deoxy-D-glucose (FDG) – positron emission tomography
HE = hematoxylin and eosin
HTLV-1 = human T-cell leukemia virus type 1
IPI = International Prognostic Index score
LDH = lactate dehydrogenase
NCCN = National Comprehensive Cancer Network
NHL = non-Hodgkin's lymphoma
PTCL-NOS = peripheral T-cell lymphoma, not otherwise specified
RAPN = robotic-assisted partial nephrectomy
TIA = T-cell intracytoplasmic antigen
WHO = World Health Organization

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How to cite this article: Miki A, Tani M, Tsutsui K *et al*. Primary renal peripheral T-cell lymphoma, not otherwise specified, treated with partial nephrectomy. *IJU Case Rep*. 2024; 7: 11–13.

Received 25 April 2023; accepted 29 September 2023.
Online publication 18 October 2023

Introduction: Renal involvement by non-Hodgkin's lymphoma is very rare, and the kidney as the primary site of this lymphoma is much more uncommon. We report a case of primary renal peripheral T-cell lymphoma, not otherwise specified, treated with partial nephrectomy.

Case presentation: A 63-year-old man was hospitalized with coronavirus infectious disease, emerged in 2019 in the emergency department. Computed tomography examination showed a 2-cm renal mass in the right kidney. Abdominal enhanced computed tomography examination revealed that the noted mass showed good enhancement in the corticomedullary phase and washout in the nephrogenic phase. No metastatic lesions were found. He was diagnosed as having cT1aN0M0 renal cell carcinoma, and robotic-assisted partial nephrectomy was carried out. The pathological diagnosis was peripheral T-cell lymphoma, not otherwise specified. He has been followed for 20 months after robotic-assisted partial nephrectomy without additional treatment and recurrence.

Conclusion: We experienced a primary renal peripheral T-cell lymphoma, not otherwise specified that was followed up without treatment after surgery.

Key words: RAPN, T-cell lymphoma.

Keynote message

Primary renal peripheral T-cell lymphoma is rare. While it is generally considered to have a poor prognosis, among cases of extranodal lesions in some organs, there are cases with a good prognosis.

Introduction

Renal involvement caused by NHL is very rare, and the kidney as the primary site of NHL is even rarer. Also, patients with PTCL-NOS, generally have a poor prognosis. Here, we report a case of PTCL-NOS in which the patient has survived for 20 months after surgery without recurrence.

Case presentation

A 63-year-old man was hospitalized with COVID-19 in the emergency department. CT examination showed a 2-cm renal mass in the right kidney. He had no palpable lymphadenopathy, and blood tests showed low lymphocytes and hemoglobin and a normal LDH (white blood cell count 6.8 × 10^3/μL, 70.2% neutrophils, 11.5% lymphocytes, hemoglobin 12.0 g/dL, LDH 219 U/L). Anti-HTLV-1 antibodies in the serum were negative. Abdominal enhanced CT examination was performed that showed good enhancement of the noted mass in the corticomedullary phase and washout in the nephrographic phase (Fig. 1). He was diagnosed as having cT1aN0M0 renal cell carcinoma, and RAPN using a retroperitoneal approach was

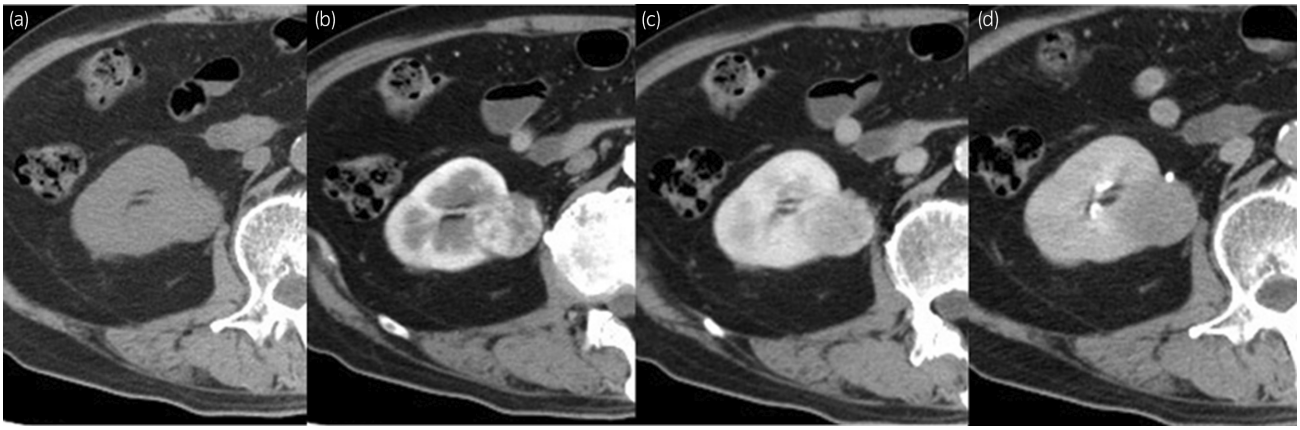

Fig. 1 Abdominal enhanced CT examination. (a) Non-enhanced CT, (b) corticomedullary phase, (c) nephrographic phase, and (d) excretory phase.

carried out. The resected specimen was a tumor with a dark red cross-section and indistinct borders. HE staining of the tumor showed diffuse infiltration of intermediate-sized atypical lymphocytes. With further immunohistochemical staining, it was found that the lymphocytes were CD3(+) and CD20(−) (Fig. 2), indicating that the neoplastic lymphoid cells were considered to be of T-cell origin. Immunostained lymphocytes were CD4(−), CD8(+), TIA-1(+), and EBER(−) (Fig. 3). We diagnosed the patient as having PTCL-NOS. Postoperative FDG-PET did not show metastasis. From the above, the disease was considered to be in the IE stage of the Lugano classification. The patient has been followed for 20 months after RAPN without additional treatment and recurrence.

Discussion

PTCL is a group of mature T-cell malignancies recognized by several entities in the current WHO classification[1] and accounts for 10–15% of all NHL. PTCL is a difficult disease to study due to its heterogeneity and rarity. According to the WHO classification, PTCL is classified as nodal, extranodal, and leukemic types, each of which includes several different pathologies. Those not fulfilling these criteria are called PTCL-NOS.[2] PTCL-NOS accounts for about 25% of all PTCL.

PTCL-NOS can occur in a variety of sites, but there are only a few reports describing its occurrence in the kidney. Two cases of renal PTCL-NOS have been previously reported. One case was diagnosed as tubulointerstitial nephritis due to tubulointerstitial infiltration of PTCL-NOS performed renal biopsy to investigate acute kidney injury of unknown cause with systemic symptoms.[3] The abdominal CT showed both kidneys slightly enlarged, but no masses. Other extranodal lesions of the skin, liver, and eye were observed. In the second case, a patient with generalized multiple abscesses was treated by antibiotics and developed acute kidney injury suspected to be drug-induced acute interstitial nephritis; a renal biopsy was performed and revealed a tubulointerstitial infiltration of PTCL-NOS.[4] The abdominal CT showed bilateral renal enlargement and no other extranodal lesions or lymphadenopathy. The current case was definitely different from the above two cases in that the renal function was normal and a solitary mass of the kidney could be

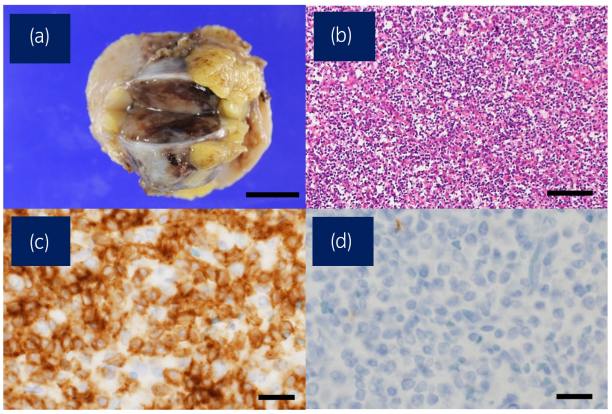

Fig. 2 (a) Cross-section of the tumor revealed a dark red interior, and the borders were indistinct. Scale bar: 1 cm (b) HE staining of the tumor showed a diffuse infiltrate of intermediately sized atypical lymphocytes. Scale bar: 100 μm Immunohistochemical staining showed that the lymphocytes were (c) CD3(+) and (d) CD20(−). Scale bar: 20 μm.

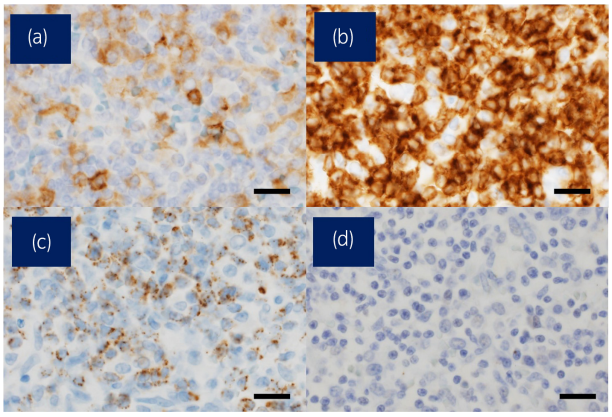

Fig. 3 Immunohistochemical staining: (a) CD4(−), (b) CD8(+), (c) TIA-1(+), (d) EBER(−). Scale bar: 20 μm.

identified. In the abdominal CT of this case, the tumor was contrasted in the arterial phase and washed out in the late phase, making it difficult to differentiate it from renal cancer as a preoperative diagnosis.

PTCL-NOS generally exhibits aggressive behavior and is often resistant to standard therapies.[5] The 5-year overall survival rate for PTCL-NOS is 50%, even at low risk in the IPI, a poor prognosis.[6,7] However, Hayashi *et al*.[8] suggested that there was a group of PTCL-NOS with slow progression and good prognosis, which should have been classified as indolent PTCL and differentiated from the other groups. The pathological findings of indolent PTCL feature small to medium-sized lymphocytes and low Ki-67 positivity (<10%).[9] Indolent PTCL sometimes presents with extranodal lesions in the thyroid, spleen, liver, and elsewhere. Lee *et al*.[9] summarized 22 reported cases of indolent PTCL in the literature. Among them, 3 cases underwent only surgical resection, and 5 cases received conservative treatment including steroid therapy. Most cases had an indolent clinical course at follow-up. In our patient, medium-sized lymphoid cells were seen, and his Ki-67 was low (6%). These suggest the possibility that ours is a case of low-grade PTCL.

PTCL-NOS remains asymptomatic until the disease progresses to advanced stages. At the time of diagnosis, systemic involvement is often already present. Complete resection of involvement is difficult, and after definitive diagnosis by biopsy, chemotherapy is usually instituted. In this case, if residual disease had been identified on postoperative FDG-PET, CHOP therapy would have been introduced. However, no residual disease was found on postoperative FDG-PET. According to NCCN guidelines,[10] low-risk patients who achieve complete remission after primary therapy can be followed up. Low-risk PTCL-NOS with complete resection of the involved lesion, as in this case, may be eligible for postoperative follow-up.

Conclusion

PTCL-NOS is generally considered to have a poor prognosis, but it is now known that among extranodal lesions in some organs, there are cases with a good prognosis. There have been no reports of cases with resectable extranodal lesions in the kidney with a good prognosis. The molecular classification of PTCL-NOS remains controversial because the genetic background of the tumor is not well understood.

Author contributions

Airi Miki: Conceptualization; writing – original draft. Masaru Tani: Writing – review and editing. Kenji Tsutsui: Writing – review and editing. Teppei Wakita: Writing – review and editing. Yuki Horibe: Writing – review and editing. Yoichi Kakuta: Writing – review and editing. Koichi Tsutahara: Writing – review and editing. Jun Ishiko: Supervision. Kyohei Yamada: Supervision. Hiroaki Miyoshi: Supervision. Koichi Ohshima: Supervision. Tetsuya Takao: Writing – original draft.

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.

Registry and the Registration No. of the study/trial

Not applicable.

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