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Effective management of a rare case of primary renal intravascular large B-cell lymphoma with modified R-CHOP regimen: A case report

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Abstract

Intravascular large B-cell lymphoma, known for its diverse organ involvement, presents significant diagnostic challenges, particularly when it affects the kidneys. This report highlights a rare case of primary renal intravascular large B-cell lymphoma in a 60-year-old male patient, who presented with persistent fever and renal dysfunction. The case underscores the intricacy of diagnosis and the efficacy of personalized treatment. Following the identification of primary renal intravascular large B-cell lymphoma, a modified R-CHOP regimen was administered, resulting in notable amelioration of symptoms and renal function following the initial treatment cycle. The patient achieved sustained complete remission without any complications after completing five subsequent R-CHOP cycles and two additional cycles of rituximab monotherapy, as confirmed by recent assessments. He is currently under regular follow-up for ongoing monitoring and improvement. This case adds to the limited yet expanding pool of knowledge concerning intravascular large B-cell lymphoma, emphasizing the necessity for personalized therapeutic strategies in atypical presentations. It also highlights the importance of early detection and customized intervention in managing rare lymphoma subtypes with unique organ involvement.

Keywords

Primary renal lymphoma, intravascular large B-cell lymphoma, modified R-CHOP regimen

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Introduction

Intravascular large B-cell lymphoma (IVLBCL) is a rare subtype of lymphoma. Its capacity to affect diverse organs has earned it the moniker "great imitator."[1] Although IVLBCL can present in various organs, renal involvement remains notably rare and poses diagnostic challenges. The incidence of renal IVLBCL is highest among Asians, and its clinical presentation often includes fever, anemia, mild acute kidney injury, and proteinuria.[2,3] A median survival of 21 months further highlights the disease's aggressive course.[4] Herein, we present a case of primary renal IVLBCL, initially manifested as fever and renal dysfunction, and elucidate its diagnostic and therapeutic journey.

Case report

A 60-year-old male with a history of pulmonary tuberculosis, emphysema, and a post-trauma splenectomy, presented with persistent afternoon fever for 40 days, peaking at 38.5°C, and accompanied by fatigue, night sweats, and

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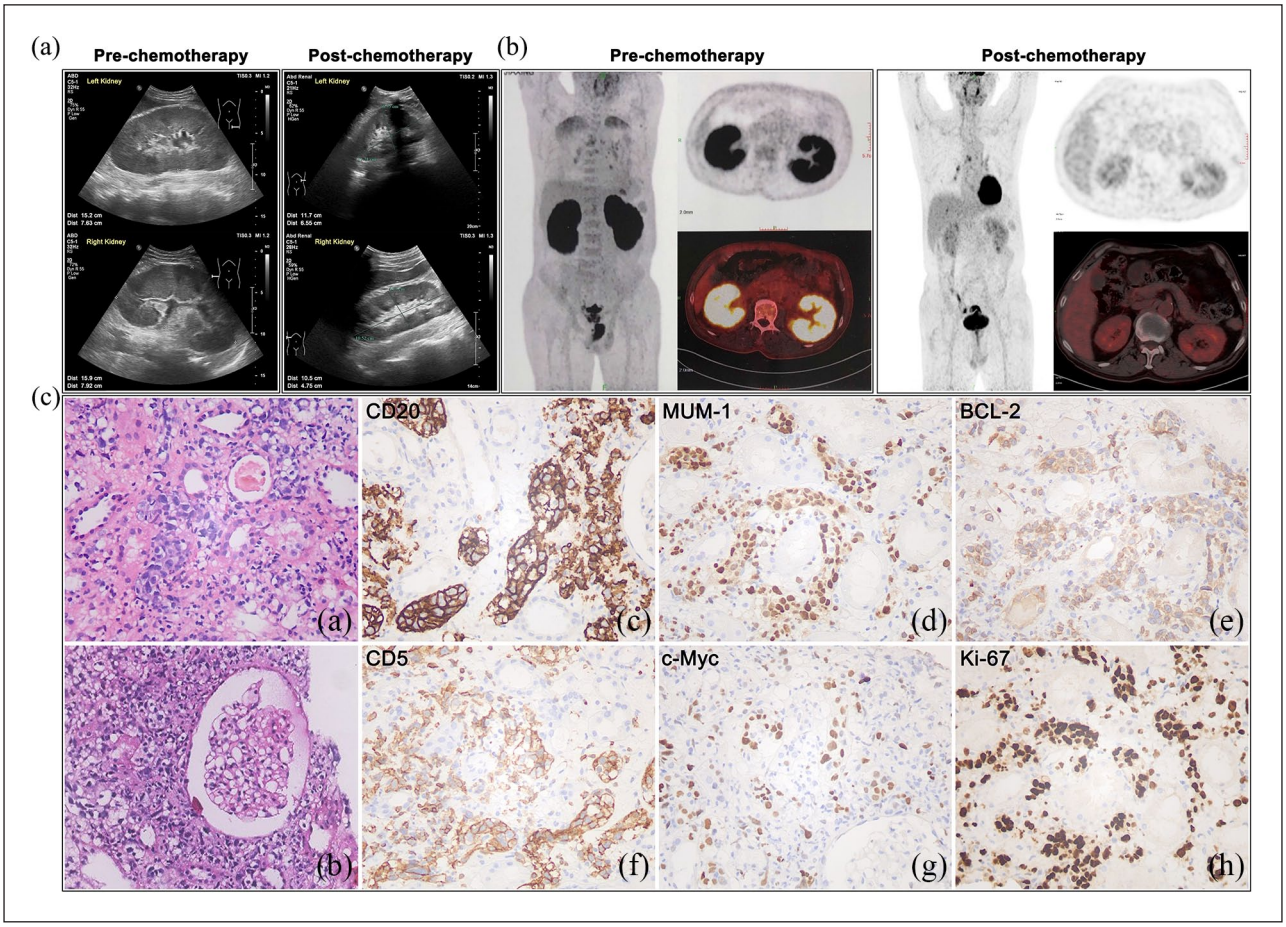

Figure 1. (a) Ultrasonography findings both pre- and post-chemotherapy are notable. Initially, ultrasonography revealed bilateral renal enlargement, with measurements approximately 15.9 × 7.9 cm for the right kidney and 15.2 × 7.6 cm for the left kidney. After 4 cycles of chemotherapy, there were reductions in renal sizes, measuring 11.7 × 6.6 cm for the right kidney and 10.5 × 4.8 cm for the left kidney, respectively. (b) PET/CT scan images. Prior to chemotherapy, the PET/CT scan displayed an increase in bilateral renal volume and diffuse, high FDG uptake, with a maximum standardized uptake value (SUVmax) of 11.4. Additionally, a mildly elevated FDG metabolism was noted in the systemic bone marrow cavity, exhibiting an SUVmax of 1.9; following 4 cycles of chemotherapy, the scan results indicated no observed increase in abnormal FDG metabolism in either the bilateral renal areas or the skeletal structures within the scanning area. (c) Histological images from an ultrasound-guided renal biopsy specimen revealed large atypical lymphomatous cells populating the blood vessels and glomerular capillary lumina, as shown in hematoxylin and eosin stains (a–b). Immunostaining highlighted these cells as CD5, CD20, c-Myc, Bcl-2, and MUM1-positive, predominantly within the vessels (c–g), and they exhibited a high Ki67 proliferation index (h).
FDG: fluorodeoxyglucose; PET: positron emission tomography.

post-activity chest tightness. He had lost 5 kg over the past month and initially attributed his symptoms to a common cold, receiving ineffective anti-infective therapy at a local clinic before seeking help at our hospital. Physical examination revealed moderate bilateral lower limb edema but no superficial lymphadenopathy, hepatomegaly, or other abnormalities. Blood tests identified elevated creatinine (356 μmol/L) and lactate dehydrogenase (532 U/L). Chest computed tomography (CT) scan disclosed minimal bilateral pleural effusion and scattered pulmonary inflammation. Abdominal CT and renal ultrasonography unveiled bilateral renal enlargement, with measurements approximately 15.9 × 7.9 cm and 15.2 × 7.6 cm for the right and left kidneys, respectively, along with heterogeneous parenchymal density (Figure 1(a)). 18F-fluorodeoxyglucose (FDG) positron emission tomography (PET) scanning demonstrated increased bilateral renal volume and diffuse high FDG uptake, with a maximum standardized uptake value (SUVmax) of 11.4. Mildly elevated FDG metabolism was observed in the systemic bone marrow cavity, with an SUVmax of 1.9 (Figure 1(b)). Ultrasound-guided renal biopsy revealed abnormal lymphoid proliferation, aligning with primary IVLBCL (non-germinal center B-cell-like subtype), corroborated by immunohistochemical findings (Figure 1(c)). The tumor cells exhibited positivity for CD5, CD20, c-Myc, Bcl-2, MUM1, and negativity for CD3,

CD10, and CD30, with a Ki-67 proliferation index surpassing 90% (Figure 1(c)). A subsequent bone marrow biopsy excluded lymphoma infiltration.

A diagnosis of primary renal IVLBCL was made, and modified R-CHOP was initiated, considering the risk of acute renal failure due to rapid tumor regression. This regimen consisted of cyclophosphamide (0.45 g on days 1–3), liposomal doxorubicin (20 mg on day 1–3), vindesine (2 mg on days 1–2), rituximab (600 mg on day 4), and dexamethasone (5 mg on days 1–5). In this modified R-CHOP regimen, we distributed the day 1 doses of cyclophosphamide, liposomal doxorubicin, and vindesine over 2–3 days and delayed rituximab to day 4, to mitigate the risk of rapid tumor lysis syndrome and ensure the patient's safety and facilitate a smooth transition to the standard R-CHOP regimen in subsequent cycles. Following treatment, the patient exhibited marked improvements, including the normalization of body temperature, alleviation of fatigue, and resolution of bilateral lower limb edema. Creatinine levels decreased to 120 μmol/L after the initial modified R-CHOP cycle, indicating substantial improvements in renal function. The patient attained complete remission (CR) with normalized renal size and function after three additional standard R-CHOP cycles (Figure 1(a) and (b)), improving quality of life and gaining 5 kg. The patient remains in a state of CR, as confirmed by the latest assessment, following the successful completion of all 6 cycles of the R-CHOP regimen and two additional cycles of rituximab monotherapy. Presently, the patient is undergoing regular follow-up examinations to ensure the maintenance of improvement and to monitor for any potential developments or changes in his condition.

Discussion

IVLBCL, a rare and enigmatic subtype of non-Hodgkin lymphoma, is characterized by its low incidence of approximately 0.5–1 cases per million individuals annually and a generally poor prognosis.[1] The rarity of this disease, coupled with its typically aggressive course, underscores the urgency for accurate diagnosis and effective treatment. Renal involvement in IVLBCL, found in about 2%–13% of cases, presents unique clinical challenges due to its atypical presentation and the critical function of the kidneys.[5,6]

The management of IVLBCL, particularly with renal compromise, necessitates a careful balancing act between efficacy and toxicity. The R-CHOP regimen,[7] a mainstay in diffuse large B-cell lymphoma (DLBCL) treatment, has shown notable effectiveness in IVLBCL, achieving an 88% CR rate, 91% overall response, and 81% 3-year overall survival rate.[8] Despite limited data, chemotherapy also shows promise in renal IVLBCL.[9] However, the standard R-CHOP protocol may require modifications when renal involvement is significant, as seen in our case. This is particularly crucial to mitigate the risk of acute renal failure, a potential complication due to rapid tumor lysis.[10] The modified R-CHOP regimen, as employed in our case, demonstrates the feasibility and success of adapting established protocols to meet the specific needs of individual patients, especially in the context of renal function preservation.

The lack of large-scale, prospective clinical trials in IVLBCL, especially concerning its renal manifestation, leads to a reliance on case reports and retrospective studies for guiding treatment strategies. This gap in literature emphasizes the need for more focused research on this lymphoma subtype. Our case contributes to the growing body of evidence suggesting that early diagnosis, followed by tailored chemotherapy, can lead to substantial improvement in renal function and overall patient outcomes. It also highlights the potential for organ-specific treatment responses, an area that warrants further exploration.

Conclusions

In conclusion, our report of a primary renal IVLBCL case underscores the critical importance of early detection and individualized treatment approaches in managing this rare and complex lymphoma subtype. The significant renal improvement observed following the initial modified R-CHOP regimen in our case reinforces the need for personalized therapeutic strategies. It also calls attention to the broader implications of understanding IVLBCL's diverse presentations and tailoring interventions accordingly. Future research should aim to develop more nuanced treatment protocols and establish clearer guidelines for managing IVLBCL, particularly in cases with rare organ involvement.

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Ethics approval

This case was reviewed and approved by the First Affiliated Hospital, College of Medicine, Zhejiang University (IIT20230869A).

Informed consent

Written informed consent was obtained from the patient for the publication of potentially identifiable images or data in this article.

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