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Primary Renal Lymphoplasmacytic Lymphoma Presenting as a Solitary Renal Mass: A Case Report

M A Suboor Shaherose[1], Vikranth Mummaneni[2], Muqarrab Ali[3], Bhavana Sadhvani[4], Vijaya Tourani[5]

[1]Consultant, Department of Medical Oncology, CARE Hospitals, Hyderabad, India
Corresponding Author Email: *suboorshaherose[at]gmail.com*

[2]Consultant, Department of Surgical Oncology, CARE Hospitals, Hyderabad, India

[3]Consultant, Department of Urology, CARE Hospitals, Hyderabad, India

[4]Consultant, Department of Pathology, CARE Hospitals, Hyderabad, India

[5]Consultant, Department of Pathology, CARE Hospitals, Hyderabad, India

Abstract: ***Background**: Primary renal lymphoma (PRL) is an uncommon extranodal manifestation of non-Hodgkin lymphoma because the kidney lacks native lymphoid tissue. Among PRLs, lymphoplasmacytic lymphoma (LPL) involving the kidney as an isolated lesion is exceptionally rare and may mimic renal cell carcinoma both clinically and radiologically. **Case Presentation**: A 57-year-old woman presented with right loin pain of a few days' duration. Contrast-enhanced imaging demonstrated a right renal mass suspicious for renal malignancy. She underwent right radical nephrectomy. Histopathological examination suggested a lymphoproliferative neoplasm, and immunohistochemistry confirmed low-grade B-cell non-Hodgkin lymphoma consistent with lymphoplasmacytic lymphoma. Whole-body ^18F-FDG PET-CT demonstrated no evidence of nodal or extranodal disease elsewhere, supporting the diagnosis of primary renal lymphoplasmacytic lymphoma. Following multidisciplinary discussion, the patient was commenced on chemoimmunotherapy with rituximab and bendamustine. Cycle 1 was administered uneventfully with pegfilgrastim support. **Conclusion**: Primary renal lymphoplasmacytic lymphoma is an exceptionally rare entity that poses a significant diagnostic challenge because it frequently masquerades as renal cell carcinoma. Histopathological examination with immunohistochemistry is essential for establishing the diagnosis. Although standardized treatment guidelines are lacking because of the rarity of the disease, systemic chemoimmunotherapy with rituximab and bendamustine appears to be an appropriate therapeutic strategy.*

Keywords: Primary renal lymphoma, lymphoplasmacytic lymphoma, bendamustine, rituximab, extranodal lymphoma.

1. Introduction

Primary renal lymphoma is a rare extranodal lymphoma accounting for less than 1% of extranodal lymphomas. The kidney is not considered a lymphoid organ, making primary lymphomatous involvement uncommon. Most renal lymphomas are secondary manifestations of disseminated systemic disease, whereas isolated primary renal lymphoma is exceedingly rare.

Lymphoplasmacytic lymphoma is an indolent B-cell lymphoma characterized by infiltration of small lymphocytes, plasmacytoid lymphocytes, and plasma cells. It most commonly involves the bone marrow and is frequently associated with Waldenström macroglobulinemia. Isolated renal involvement without bone marrow or systemic disease is exceptionally uncommon, with only a limited number of cases reported in the literature.

We report a case of primary renal lymphoplasmacytic lymphoma presenting as a solitary renal mass, initially suspected to represent renal cell carcinoma.

2. Case Presentation

A 57-year-old woman presented with complaints of right-sided loin pain of a few days' duration. There was no history of fever, weight loss, night sweats, hematuria, or lower urinary tract symptoms. Physical examination was unremarkable.

Radiological evaluation demonstrated a right renal mass suspicious for renal cell carcinoma. In view of the imaging findings, she underwent right radical nephrectomy.

Gross pathological examination revealed a renal mass. Histopathological evaluation demonstrated diffuse infiltration by atypical small lymphoid cells suggestive of a lymphoproliferative disorder. Immunohistochemistry confirmed a diagnosis of low-grade B-cell non-Hodgkin lymphoma consistent with **lymphoplasmacytic lymphoma**.

Following histological diagnosis, staging investigations were undertaken. Whole-body 18F-FDG PET-CT revealed no metabolically active lymphadenopathy or extranodal disease elsewhere, suggesting isolated renal involvement consistent with primary renal lymphoplasmacytic lymphoma.

The diagnosis, disease biology, prognosis, and available treatment options were discussed extensively with the patient and her family in a multidisciplinary setting. Considering the diagnosis of localized extranodal lymphoplasmacytic lymphoma following nephrectomy, systemic chemoimmunotherapy with rituximab and bendamustine was planned. The patient tolerated chemotherapy well and at the time of submission of this manuscript she had completed 3 cycles of chemotherapy and was in remission.

Images:

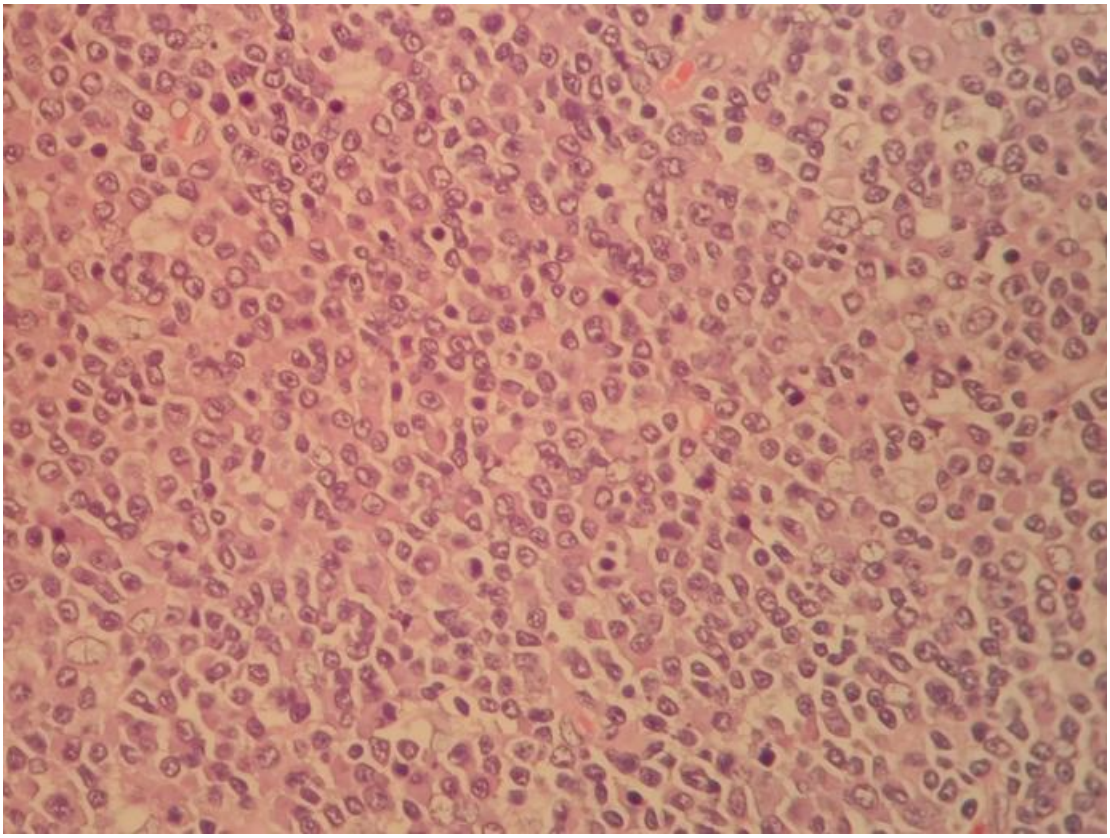

Image 1: Histopathology H&E staining image

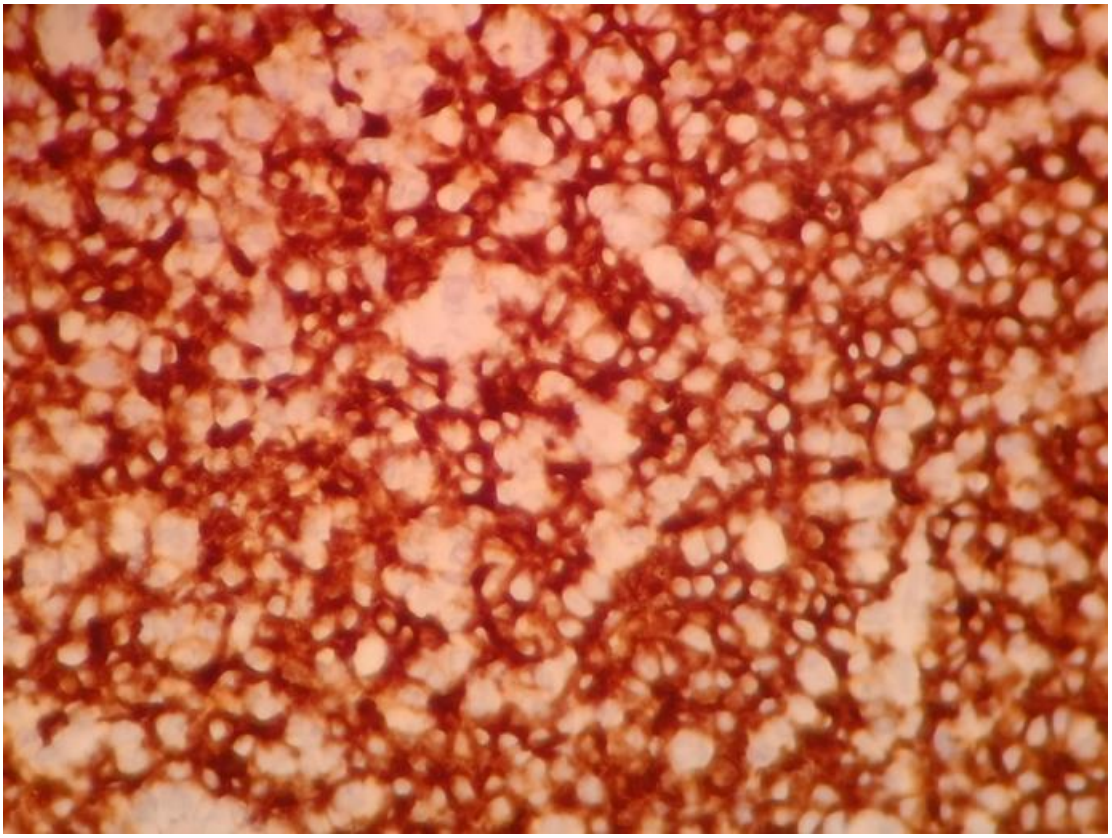

Image 2: IHC CD 20

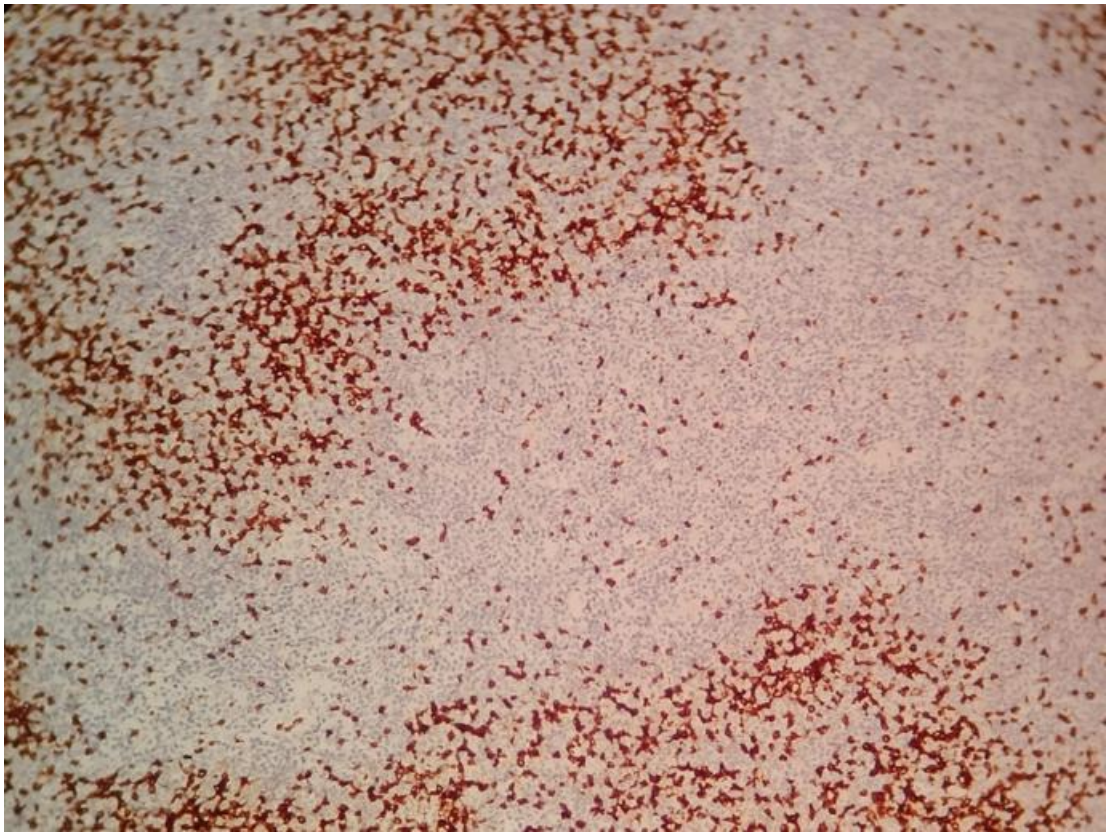

Image 3: IHC CD 5

3. Discussion

Primary renal lymphoma remains a controversial clinical entity because of the absence of native lymphatic tissue within the renal parenchyma. Proposed mechanisms include origin from the renal capsule, lymphatics surrounding the kidney, or chronic inflammatory processes leading to lymphoid proliferation.

Most reported primary renal lymphomas are diffuse large B-cell lymphomas. Low-grade B-cell lymphomas involving the kidney are considerably less common, and lymphoplasmacytic lymphoma presenting exclusively within the kidney is exceptionally rare.

Patients often present with nonspecific symptoms such as flank pain, abdominal discomfort, hematuria, or an incidentally detected renal mass. Radiological imaging cannot reliably distinguish renal lymphoma from renal cell carcinoma, resulting in many patients undergoing nephrectomy before the diagnosis is established.

Histopathological examination supported by immunophenotyping remains the gold standard for diagnosis. Additional investigations, including bone marrow examination, serum protein electrophoresis, immunoglobulin quantification, and molecular testing for *MYD88* L265P mutation (where available), may help confirm the diagnosis and exclude systemic lymphoplasmacytic lymphoma associated with Waldenström macroglobulinemia.

Because of the rarity of this condition, there are no prospective studies defining the optimal treatment approach. Rituximab-based chemoimmunotherapy remains the standard treatment for symptomatic lymphoplasmacytic lymphoma requiring therapy. Bendamustine combined with rituximab has demonstrated high response rates with durable disease control and a favorable toxicity profile in indolent B-cell lymphomas, making it a reasonable choice in this patient.

Long-term surveillance is essential because late relapses may occur even after an excellent initial response.

4. Conclusion

Primary renal lymphoplasmacytic lymphoma is an exceptionally rare cause of a renal mass and represents an important diagnostic consideration in patients undergoing nephrectomy for presumed renal cell carcinoma. Definitive diagnosis depends on histopathology and immunohistochemistry. This case highlights the importance of considering lymphoma in the differential diagnosis of renal masses and demonstrates that rituximab-bendamustine chemoimmunotherapy is a rational treatment option following accurate diagnosis.

Declarations:
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