

Oncology

Primary renal lymphoma: An incidental finding in an elderly male

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A B S T R A C T

Primary renal lymphoma (PRL) is a rare disease process which represents less than 1% of all renal masses and is an uncommon type of Non-Hodgkin's Lymphoma. PRL is of clinical significance due to the fact that unlike the more commonly seen renal cell carcinoma, PRL is treated with neoadjuvant chemotherapy followed by nephrectomy. This challenges the long held notion that preoperative biopsies are not necessary prior to nephrectomy for renal masses. In this paper, we present a case of a primary renal lymphoma and discuss its clinical significance.

Introduction

Non-Hodgkin's Lymphoma (NHL), a cancer that originates in the lymphatic tissue, accounts for 4.3% of all cancers in the United States.¹ Primary renal lymphoma (PRL) is a rare type of NHL, with B cell lymphoma being the most common subtype; to date, no more than 100 cases of PRL have been described.² PRL accounts for 0.68% of all extranodal NHL masses in North America and makes up less than 1% of all renal masses identified.³

The exact pathophysiology of PRL is not fully understood as the kidney lacks lymphoid channels. Theories propose an origin in subcapsular lymphatic tissue, dissemination via a hematogenous route, an extension of inflammatory disease, or secondary to mucosa-associated lymphoid tissue (MALT) renal lymphoma tissue.² PRL can have many growth patterns within the kidney with the most common being multiple renal masses (60%), followed by diffuse infiltration (25%), and less commonly as a solitary mass (15%).⁴ PRL is reported to occur more commonly in men with a median age of 45–65. Patients may present with flank pain as well as the B symptoms (fevers, night sweats, and unintentional weight loss) typically seen in lymphomas. Clinical findings often include hematuria, proteinuria, and renal failure. There are several imaging studies that can be utilized to workup renal masses and detect PRL, such as ultrasound, intravenous urography, nuclear medicine, magnetic resonance imaging (MRI), and computed tomography (CT). Although all of these techniques can provide valuable information in the diagnosis, CT scan remains the most sensitive and comprehensive modality.⁴ However, if renal failure or allergy to contrast is present, MRI becomes the diagnostic modality of choice.

Case presentation

A 79-year-old male was brought in by ambulance to the emergency department after sustaining a fall at home. The patient was found by his family several hours after the fall lying on the floor and unable to ambulate. He had a past medical history of coronary artery disease, obesity (body mass index of 46.35 kg/m²), hypertension, and atrial fibrillation as well as a 32-pack-year history of smoking. The patient's only complaint upon arrival was diffuse myalgias and physical examination was unremarkable. On review of systems, the patient denied fevers, chills, night sweats, or weight loss. Initial workup showed a creatinine of 2.8 mg/dL, blood urea nitrogen of 55 mg/dL, potassium of 6.2 mEq/L, white blood cell count of 16.91 × 10³ cells, and microhematuria. Labs from several months earlier showed normal renal function with a creatinine of 1.08 mg/dL and a potassium of 4.9 mEq/L.

Upon further workup for acute kidney injury, a renal ultrasound was performed which revealed a 4.7 × 4.3 × 3.9 cm hypoechoic region within the midpole of the left kidney. This warranted further investigation with a CT abdomen and pelvis study which demonstrated an enhancing left perinephric soft tissue density with similar appearing soft tissue encasing the left renal artery [Fig. 1]. Following the suspicion of a renal mass, lactate dehydrogenase levels were ordered which were shown to be elevated at 278 units/L.

The patient underwent CT guided biopsy of the mass, which revealed B cell lymphoma of follicular center cell origin [Fig. 2]. The histopathological examination showed lymphoma cells that were small and in a diffuse pattern. Immunohistochemistry analysis showed that the tumor cells were positive for CD20, CD10, Bcl-2, Bcl-6, and negative for Cyclin D1 [Fig. 3]. The diagnosis of primary renal lymphoma was

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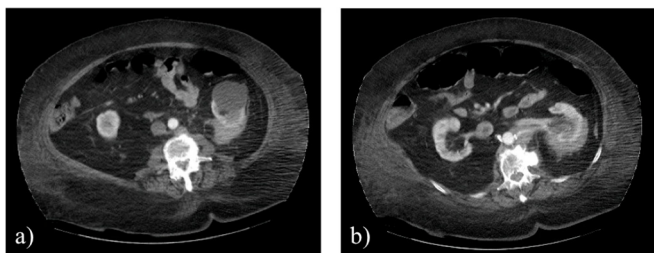


Fig. 1. Contrast enhanced CT scan of the abdomen showing a) enhancing left perinephric soft tissue density b) soft tissue density encasing the left renal artery.

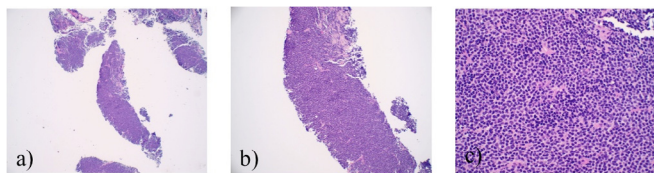


Fig. 2. Primary renal lymphoma pathology slide at a) 4X magnification b) 10 × magnification c) 20 × magnification.

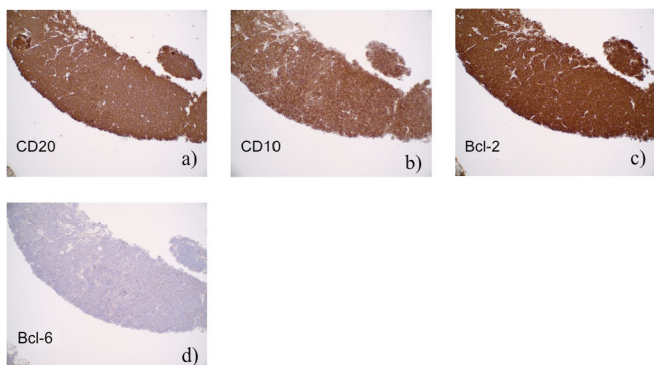


Fig. 3. Primary renal lymphoma pathology slide with staining positive for a) CD20 b) CD10 c) Bcl-2 d) Bcl-6.

subsequently made. Upon discharge, the patient was scheduled for follow-up with oncology for treatment and further evaluation.

Discussion

PRL is a rare disease, constituting only a small percentage of all types of renal masses and all forms of NHL. The etiology of PRL is a debated topic as the kidney is an extranodal organ that lacks lymphatic channels. It is unknown how lymphoma originates in the kidneys but there are several theories that try to explain this phenomenon. Some

believe that PRL originates in the lymphatics of the renal capsule with invasion into the renal parenchyma. Others believe it is spread through hematogenous dissemination, which is a possible explanation of why many of the cases of PRL described in the literature occur bilaterally, or occurs from extension of inflammatory tissue. MALT tissue found in the kidney is also a potential origin site of PRL. Due to the rarity and unclear etiology of PRL, it is often overlooked in the differential for renal masses in favor of more common pathologies such as renal cell carcinoma (RCC), renal cysts, and nephroblastoma.

PRL is most commonly misidentified as RCC, which can present with similar symptoms, including the classic triad of hematuria, flank pain, and a palpable mass in the flank or abdomen. However, on diagnostic imaging RCC tends to have a more heterogeneous appearance than PRL. Standard management of renal masses typically includes nephrectomy. PRL differs in that it is first treated with neoadjuvant chemotherapy followed by nephrectomy. Therefore, diagnostic and confirmatory studies are key to differentiating between RCC and PRL, which may present similarly. Early diagnosis and treatment with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone (R-CHOP) may improve renal function within 2–4 weeks of initiating therapy.² However, despite oncologic treatment, the 1-year mortality rates of PRL can be as high as 75% with a 5-year survival rate of only 40–50%.³

Of the reported cases of PRL, a majority of the patients presented with symptoms such as fever, night sweats, weight loss, anorexia, nausea, vomiting, flank pain, hematuria, proteinuria, and hydronephrosis. According to a literature review of 49 cases of PRL, only 4 cases were noted to be older than the age of 75.⁵ This patient differs from other cases in that he was asymptomatic at presentation, did not exhibit preceding B symptoms, is older than the median age of 45–65, and the mass was unilateral.

Conclusion

The patient in this study was diagnosed incidentally with PRL during a routine workup for acute kidney injury. This case report attempts to highlight the rarity of PRL while demonstrating the significance of early diagnosis and treatment.

References

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