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Isolated perinephric lymphoma: A distinct presentation of primary lymphoma

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ABSTRACT

Isolated perinephric lymphoma is a rare presentation of primary lymphoma. We report a case of a 77-year-old male who was incidentally found to have isolated left perinephric lymphoma. We outline the diagnostic work up and subsequent diagnosis of lymphoma as well as the considerations that guided management. Ultimately, bone marrow biopsy and PET-CT were used as diagnostic tools to assess for systemic disease and the patient was managed with observation and interval follow-up imaging.

1. Introduction

Non-Hodgkin's Lymphoma (NHL) involving the kidneys is common, however only 0.7% of cases in North America are due to primary renal lymphoma, most commonly diffuse large B cell lymphoma.[1] Even more rare is isolated perinephric lymphoma, wherein lymphoma originates within the renal fascial layers, sparing the renal parenchyma and neighboring lymphatic structures.[2] Renal lymphoma is typically treated with systemic chemotherapy, commonly with cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP).[3] Rituximab, an anti-CD20 monoclonal antibody, can be used in conjunction with other chemotherapies for both indolent and aggressive NHL.[3] Unlike renal lymphoma, primary perinephric lymphoma has no established standard of care and little is known about its prognosis. Herein, we present a patient diagnosed with perinephric lymphoma and the considerations that informed our management.

2. Case presentation

A 77-year-old male with a remote history of prostate cancer managed with external beam radiation therapy presented with acute onset right flank pain. There was no hematuria, flank mass, weight loss, fevers, or adenopathy on further history and examination. Subsequent non-contrast computed tomography (CT) demonstrated a 9 × 5cm posterior subcapsular mass encompassing the left kidney without evidence of urinary tract compression or hydronephrosis (Fig. 1). The differential diagnosis included primary renal malignancy, including renal cell carcinoma, lymphoma, metastasis, perirenal fibroma and retroperitoneal tumors including fibrosarcoma.

Due to the broad differential diagnoses, the patient underwent an uncomplicated percutaneous biopsy of the left perirenal mass which revealed marginal zone lymphoma, a sub-type of low-grade B-cell lymphoma. Pathological analysis demonstrated diffuse proliferation of atypical small lymphoid cells with slightly irregular nuclear contour, moderate amount of cytoplasm and inconspicuous nucleoli (Fig. 2). Immunohistochemistry study demonstrated atypical cells that were positive for CD20, PAX-5, BCL-2 and negative for CD5, CD10, CD23 and Cyclin D1. Concurrent flow cytometry revealed a surface Kappa light chain restricted B-cell population (~65% of total) expressing CD19, CD20, CD22 and CD38 (dim), while negative for CD5, CD10, CD34 and TdT (Fig. 3). The pathologic diagnosis was consistent with marginal zone lymphoma.

The patient was referred to medical oncology, where he underwent a bone marrow biopsy that did not show evidence of lymphoma involvement. ^{18}F-fluorodeoxyglucose (FDG) Positron-Emission Tomography/Computed Tomography (PET-CT) demonstrated increased uptake with a maximum standardized uptake value (SUVmax) of 4.8 by the perirenal mass and no evidence of lymphadenopathy (Fig. 1). At this time, the patient's symptoms had resolved. With a diagnosis of primary perinephric NHL, discussed treatment options included chemotherapy,

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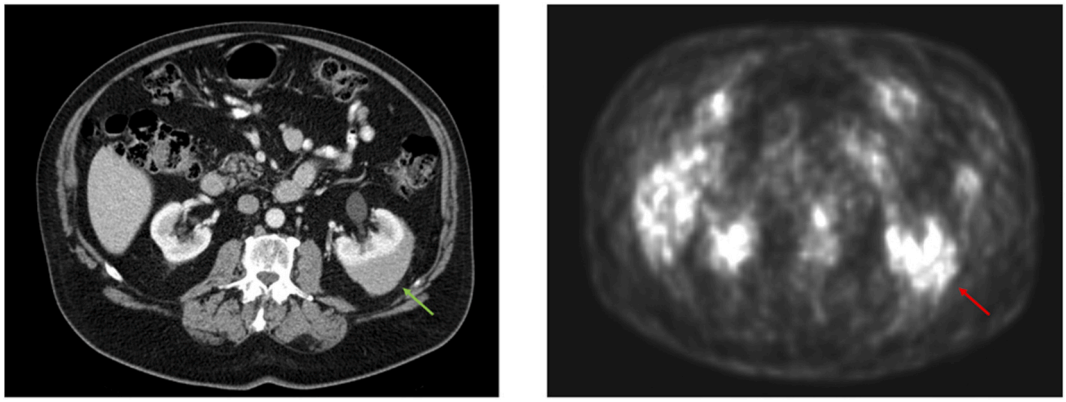

Fig. 1. Computed Tomography (CT) w/contrast showing an enhancing 9 × 5cm left posterior mass (green arrow). Positron-Emission Tomography/Computed Tomography (PET/CT) showing a 3.5 × 6.2cm curvilinear density inseparable from the renal capsule (red arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

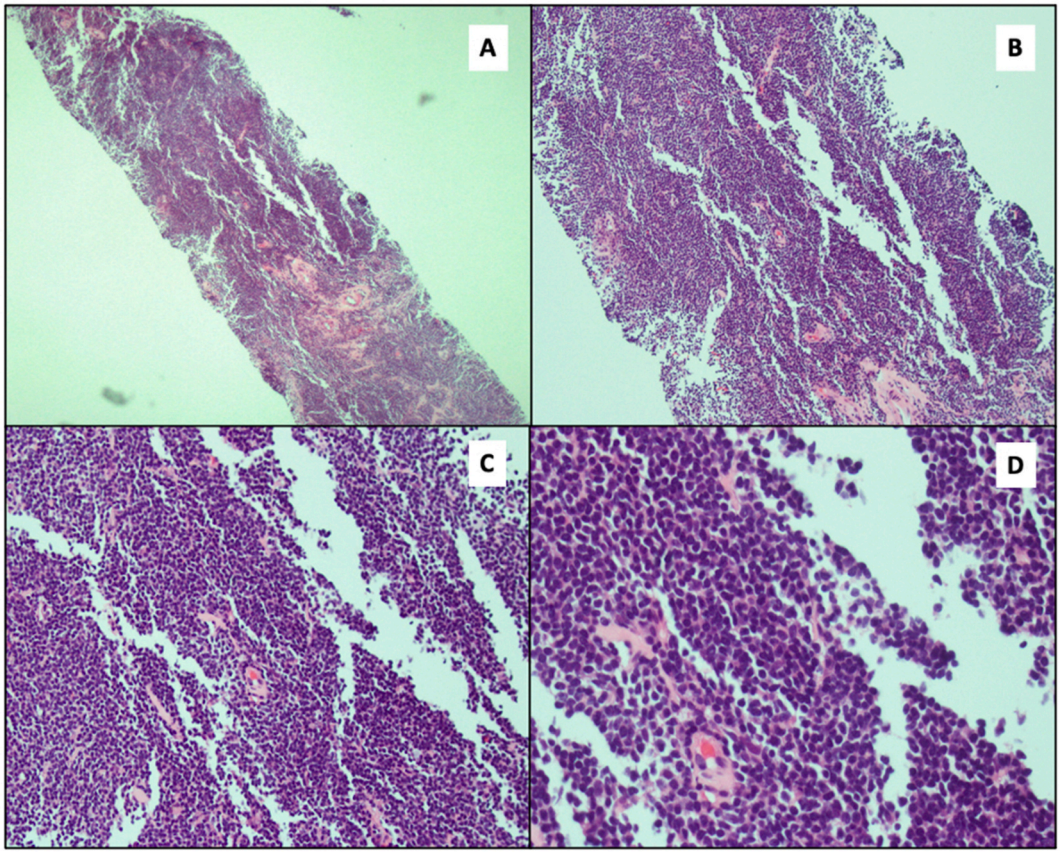

Fig. 2. Retroperitoneum mass biopsy (H&E sections): A. Low power magnification at 4x showing effaced lymph node architecture. B. Low power magnification at 10x showing diffuse proliferation of atypical small lymphoid cells. C. Low power magnification at 20x showing no necrosis or mitosis. D. High power magnification at 40x showing lymphoid cells with slightly irregular nuclear contour, moderate amount of cytoplasm and inconspicuous nucleoli.

local radiation, and observation.

Given the patient's flank pain had resolved and he was otherwise asymptomatic, no evidence of systemic disease, and stable mass size, chemotherapy was not pursued. Furthermore, the risks of immunosuppression secondary to chemotherapy in the context of the COVID-19 pandemic were considered. Radiation therapy was not advised due to lack of data supporting its use to treat perirenal lymphoma. Radical nephrectomy was not considered due to unimpaired renal function. The patient decided on conservative management with observation and serial imaging. At 6-month follow-up, the patient underwent repeat PET-CT which revealed no change in the size or FDG uptake of the perinephric mass and no evidence of systemic disease; therefore, he continues with conservative management.

3. Discussion

Management of perinephric NHL is largely unexplored due to its exceptional rarity, as supported by the paucity of case reports. A case report from 2000 described a 73-year-old man with nonspecific symptoms of fever, malaise, abdominal fullness and hepatomegaly who was

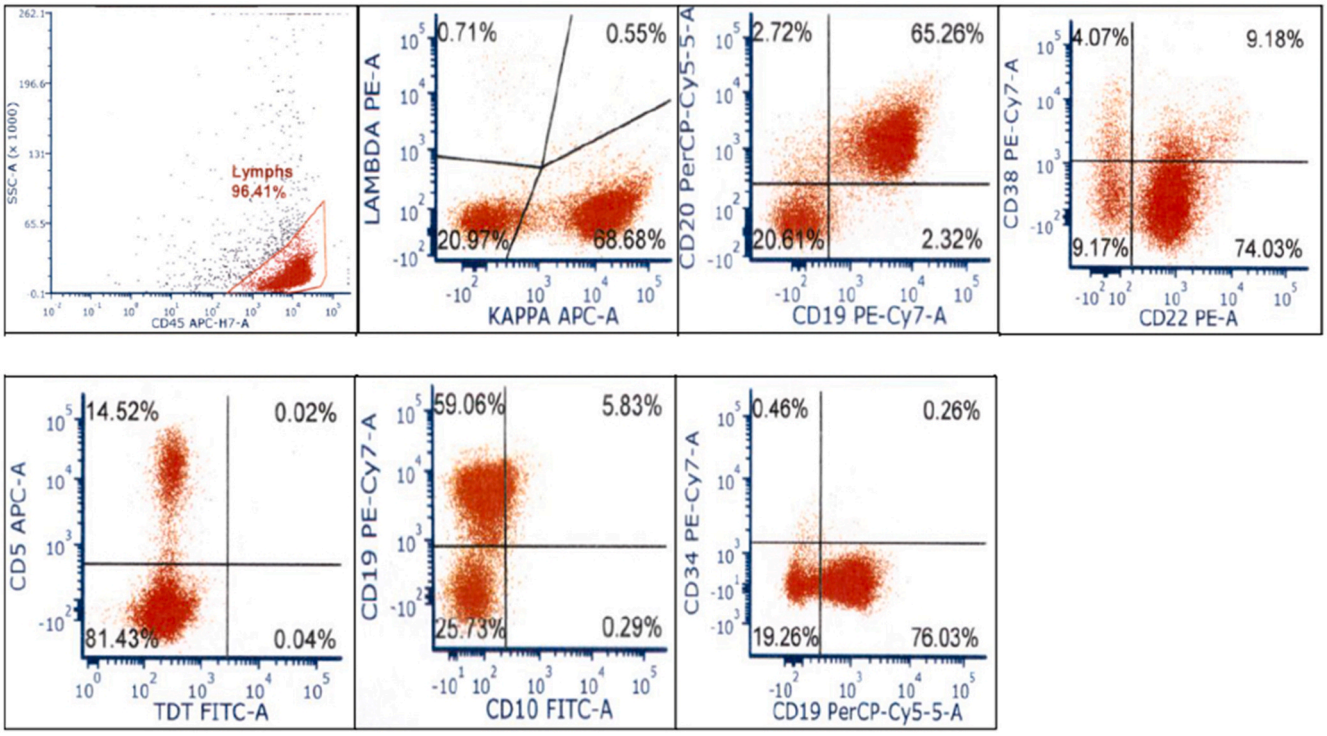

Fig. 3. Cytogram analysis of CD45 positive lymphoid gate shows surface kappa light chain restricted B cell population. Positive markers: CD19, CD20, CD22, CD38 (dim) Negative markers: CD5, CD10, CD34, TdT.

ultimately diagnosed with perinephric NHL. Following treatment with combination chemotherapy for 6 months (regimen not specified), the patient's tumor had not responded and subsequent bone marrow biopsy revealed infiltrative disease.[4] This suggests the need to pursue a full work up, including bone marrow biopsy, upon initial diagnosis to evaluate for systemic disease. The bone marrow biopsy results in this patient also shed doubt on whether the perinephric mass was a primary lymphoma or secondary to unidentified systemic disease. Additionally, this case emphasized the importance of maintaining a high level of suspicion for lymphoma when presented with an isolated perirenal mass. This conclusion is further supported by a case report from 2009, which described a 72-year-old man who also presented with nonspecific symptoms of flank pain, fever and fatigue. The patient underwent a CT scan, Magnetic Resonance Imaging (MRI) scan and an ultrasound before ultimately having a biopsy revealing perinephric NHL.[2] This patient was managed with the CHOP regimen.

Unlike prior case reports, we utilized PET-CT as an early diagnostic tool to assess for systemic disease, as PET-CT has 97% specificity and sensitivity for renal lymphoma.[5] Our use of PET-CT in the work up of perinephric lymphoma provides novel anecdotal evidence of its influence on management decisions for this patient.

Ultimately, management of our patient was guided by the patient's symptoms, biopsy results, unremarkable laboratory results, unremarkable physical exam, lack of renal impairment, lack of systemic disease, the stable size of the mass and discussion with the patient. Although further follow-up is warranted, close observation avoids the toxicity of systemic therapy and was corroborated by a PET-CT performed 6 months later revealing no change in mass and no evidence of metastatic disease. While we cannot definitively say close observation is the best management option, it appears a justifiable option and was the patient's preference. Further follow-up is necessary to reveal how optimal the decision ultimately was. A conservative approach is a relatively novel option for the management of perinephric lymphoma. The multidisciplinary workup for our patient was done swiftly so that the proper clinical teams could be involved in the management decision. The team maintained a high level of suspicion for lymphoma and biopsied the lesion early in the work up.

4. Conclusion

This case provides a framework for the work-up and management of isolated perinephric lymphoma, an otherwise poorly elucidated manifestation of lymphoma. Our report highlights the importance of an expeditious work up including a PET-CT while also revealing that careful observation may ultimately be a justifiable course of action.

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Declaration of competing interest

None of the contributing authors have any conflict of interest, including specific financial interests or relationships and affiliations relevant to the subject matter or materials discussed in the manuscript.

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