Case Report

Primary Lymphoma of the Kidney: Case Report and Review of Literature

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Clinical Practice Points

- Lymphomatous involvement of the kidney is often seen as a part of disseminated disease. Characteristics of disease are poor prognosis and survival of less than a year after diagnosis. The primary renal lymphoma is very rare, is usually part of general disease, is difficult to diagnostic, and has a poor prognosis.
- Noncharacteristic solitary renal masses need to be differentiated from renal cell carcinoma. We present a 73-year-old man with a solitary renal mass treated with partial nephrectomy and histologic diagnosed with a primary lymphoma of the kidney. In the preventive diagnosis, conventional radiology is unsuitable, contrast-enhanced renal ultrasound contributes to the diagnosis, and primary renal biopsy is useful.
- We reviewed the medical literature and discuss primary renal lymphoma, a rare disease with poor prognosis, whose treatment after postsurgical diagnosis based on chemotherapy with R-CHOP (rituximab plus cyclophosphamide/doxorubicin/vincristine/prednisone).

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Introduction

Incidence of non-Hodgkin lymphoma (NHL) has progressively increased to 150% compared with the past century.[1] The evolution of this disease shows a variation of incidences and a significant increase in patients older than the normal bimodal pattern in which the age group most affected is the young adult, predominantly male, population. The incidence of cases of renal involvement in NHL has been reported as between 2.7% and 6%.[2] Compared with a secondary involvement, primary renal lymphoma is exceptional and represents about 1% of the all renal tumors.[3]

The existence of a primary lymphatic renal disease (PRL) has been discussed, because the kidney does not contain lymphatic tissue and it is unclear how the disease may develop. Our hypothesis is the following: the presence of lymphatic tissue lymphatic vessels of the hilum or capsule or intensive recall of the B lymphocytes in the parenchyma in response to a persistent inflammation. There are approximately 100 cases described in the medical literature.[4] The term PRL is then applied to a renal localization in the absence of clinical manifestations of lymphatic disease in other organs and may also be defined as a NHL that starts. We present the case came to our observation and review of the literature.

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Case Report

An 73-year-old white male patient came to our attention for ultrasound documented bilateral renal cysts. Results of blood tests showed only a slight lymphocytopenia (15.8% of lymphocytes) and neutrophilic leukocytosis (76.8% of leukocytes). Hepatitis serologies, lactate dehydrogenase, and chest radiographs were negative. Due to the thickened wall cyst with a diagnostic doubt, the patient underwent contrast-enhanced renal ultrasound (Sonovue Bracco Diagnostics Inc, Princeton, NJ) that confirm, in the third lower right kidney with anterior development, a irregular appearance formation of 52 mm, partly solid. Administration of contrast agent showed enhancement of the wall, and remains avascular the central portion. The patient then underwent a contrast computed tomography (CT) of the abdomen, which confirmed the presence of lesion average 52 mm diameter, moderately thickened walls at baseline, irregular and hyperdense, and homogeneous content. After administration of contrast, there was no significant enhancement of the wall of lesion or parietal tokens are shown. CT of the chest, abdomen, and pelvis showed no further histologic localization of disease; also, a bone marrow biopsy showed a bone marrow free of significant histologic changes. Renal tumor was suspected and therefore we do not do a positron emission tomography, moreover, this examination cannot

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Figure 1 (A) Atrophic Renal Tubules. (B) Infiltration of Lymphoplasmacytic Cells. (C) Fibrous Capsule. (D) Tumor Cells. (E) Necrosis. Renal Lymphoma (Hematoxylin and Eosin) × 30

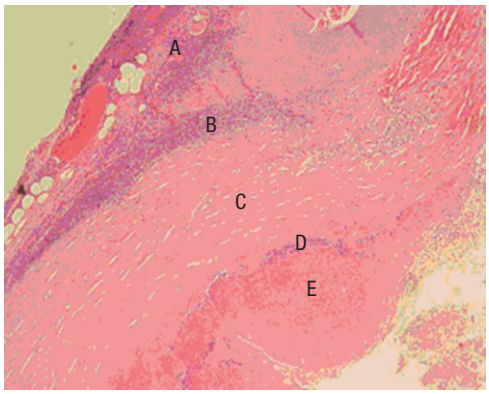
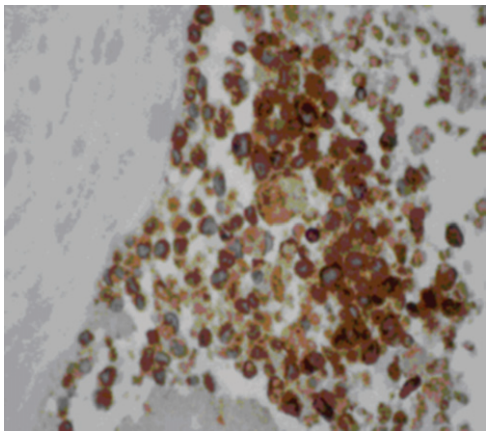


Figure 2 Immunohistochemistry of Renal Lymphoma: coloration for CD20^{+} Lymphocytes × 400



evaluate well the kidney because renal fluorine-18 fluorodeoxyglucose excretion interferes with kidney imaging.

A preliminary characterization of lesion biopsy was not performed due to the risk of seeding in this malignancy case. Due to the reduced renal function (Glomerular filtration rate 59 mL/min), we decided to perform neprhon-sparing surgery enucleoresection. The patient was treated with enucleoresection with renal warm ischemia for 22 minutes. On histologic examination, macroscopic formation had a maximum diameter of 6.8-cm, a thick fibrous wall that contained a large amount of necrotic dampish material, yellowish-gray in color. A microscopic examination documented non-Hodgkin anaplastic large cell B extensively necrotic and capsule (Figure 1). The cells were positive for CD 20^{+} (Figure 2), PAX 5^{+}, Bcl 2^{+}, CD 30$^{\pm}$, vimentin^{+}, and CD 45^{+}, and were negative for CD3^{-}, CKMNF116^{-}, CK7^{-}, CD10^{-}, EMA^{-}, CD56^{-}, desmin^{-}, -Myod1, Rod^{-}, and CD31^{-}.

Discussion

PRL is defined as a NHL that starts in the parenchyma and does not show invasion into adjacent lymph nodes. It is very rare and the diagnosis of PRL is difficult.[5] Lymphomas are malignant neoplasms characterized by the proliferation of cells in the lymphoid tissue. Lymphomatous involvement of the genitourinary tract occurs in 3%-6% of cases and is most common in the testis. The primary lymphoma of the kidney is a unique disease entity, because the kidney does not contain lymphatic tissue. There is no evidence that the disease is the primary or initial manifestation of systemic disease rapidly aggressive. It has been suggested that the lymphoma may originate from the renal sinus lymphatics or lymph vessels located near the renal capsule and form cords that enter into renal parenchyma, or follow chronic inflammation (such as chronic pyelonephritis, Sjogren's Syndrome, Systemic Lupus Erythematosus, Epstein Barr Virus of mononucleosis).[6] Many of the documented cases of renal lymphoma are secondary, and postmortem studies have found that approximately 50% of patients who died of NHL had renal involvement. PRL most commonly affects older male subjects, those older than 60 years (mean age at diagnosis, 64 years), frequently unilateral, rarely bilateral.[7,8] Large B-cell lymphoma is the most common histologic type, in up to approximately 30% of NHL, and is among the most clinically aggressive lymphoma. The clinical manifestations are similar to those of other renal neoplasms also may present with: proteinuria, nephrotic syndrome, kidney failure. It is very aggressive, with rapid systemic dissemination. Survival is extremely low, approximately 75% of patients die within the first year after surgery.[6] The final prognosis can be improved by early diagnosis and systemic chemotherapy. There are several options for the diagnostic evaluation of a possible involvement of the kidney, including renal ultrasonography, intravenous urography, CT, magnetic resonance imaging, and nuclear medicine.[7] The ultrasound contrast agent is the mode of choice, whereas normal CT with contrast techniques have not been a valuable diagnostic aid due to the extreme variability of findings that may be encountered in cases of renal lymphoma.

Conclusions

Overall, the prognosis of patients with renal lymphoma is less than 1 year.[5] The PRL is considered a systemic disease with renal manifestation, even without evidence of extrarenal locations. The total nephrectomy is not indicated, especially if it was possible to make a preoperative diagnosis.[6] Patients with atypical findings suspected for renal cell carcinoma should undergo a percutaneous renal biopsy, which has a sensitivity and a specificity of 70% -92% and 100%, respectively.[5,6] Although renal lymphoma is often part of a generalized process, our case appears to be limited to the kidney, so-called stage IE diffuse large B-cell lymphoma. This early-stage aggressive lymphoma, may be highly curable with CHOP (cyclophosphamide, doxorubicin, vincristine [Oncovin], and prednisone] plus rituximab, based on the revised International Prognostic Index of Sehn et al,[9] but only 41%-53% of patients with advanced disease are alive and disease free at a median follow-up of 3 years, especially when there is an overexpression of Bcl 2.[5-9]

Disclosure

The authors have stated that they have no conflicts of interest.

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