A case report of a renal diffuse B-cell lymphoma

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Diffuse large B-cell lymphomas (DLBCL) are the most common lymphomas worldwide. They also appear to be the most common primary retroperitoneal lymphomas, but this presentation is relatively uncommon in the literature. Retroperitoneal masses, including lymphomas, often present with nonspecific symptoms and laboratory values, necessitating radiographic assessment and consideration of mass biopsy prior to the initiation of treatment. Here we present a case of a primary retroperitoneal DLBCL as well as a review of the clinical presentation, imaging findings, and differential diagnosis of such tumors.

Key Words: retroperitoneal mass, renal mass, lymphoma, computed tomography, biopsy

Introduction

Although the vast majority of retroperitoneal masses originate from the kidney, tumors of other retroperitoneal origin exist and may often resemble a renal mass. These tumors include multiple benign and malignant tumors, such as sarcomas, pheochromocytomas, germ cell tumors, and lymphomas. In evaluating renal/peri-renal tumors with an atypical radiographic appearance, it is important to remember this differential diagnosis. Multiple clinical clues can push physicians in the appropriate direction, specifically patient presentation and certain radiographic details of the mass. This case report describes a primary retroperitoneal diffuse large B-cell lymphoma (DLBCL) diagnosed with biopsy after imaging for lower back pain originally suggested a large left renal mass.

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Case report

The patient is a 53-year-old male with a past medical history of HIV and hepatitis B who presented with lower back pain with imaging from an outside facility revealing a large left-sided retroperitoneal mass.

Additional symptoms included mild nausea, decreased appetite, diarrhea, and weight loss over a 6 month period. His vital signs were stable and his physical exam was unremarkable except for a palpable left-sided retroperitoneal mass. The patient's CT scan of the abdomen and pelvis with contrast was reviewed, Figure 1 and 2. It showed a large homogenous retroperitoneal mass extending from the left kidney down to the pelvis without compression or significant displacement of the major vessels. The mass appeared to be displacing the kidney superiorly and anteriorly; its radiographic appearance was not typical for a mass emanating from the kidney itself. Of note, his laboratory work was normal except for a serum calcium of 13 and a CD4 count of 116. Due to the atypical radiographic appearance of the mass and his pertinent laboratory results, a CT-guided biopsy of the mass was performed. Cytology from the biopsy sample revealed DLBCL. Etoposide, Prednisone, Vincristine, Cyclophosphamide, Hydroxydaunomycin (EPOCH) therapy was therefore initiated. The patient was also restarted on HAART therapy with Abacavir, Epivir, and Dolutegravir. The patient is now receiving systemic chemotherapy for his DLBCL and is responding well.

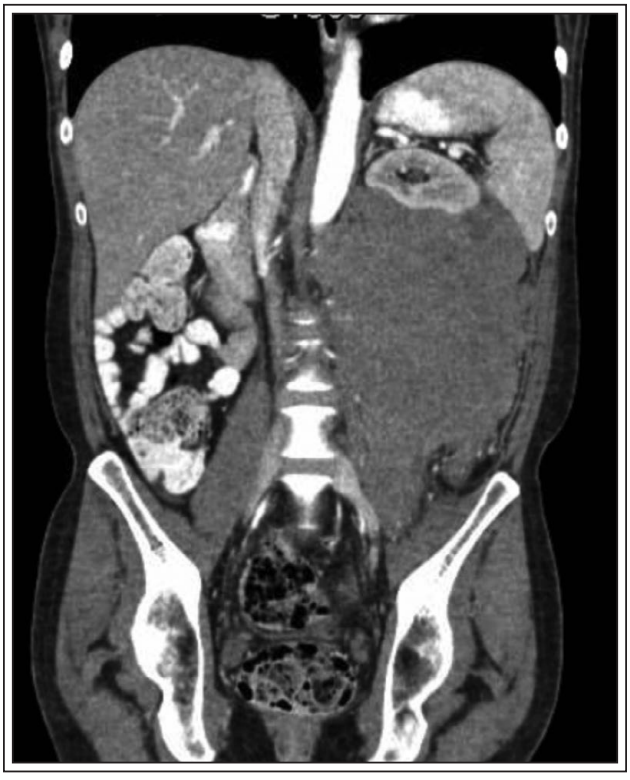

Figure 1. Coronal view of a contrast enhanced CT of the abdomen and pelvis showing a large left retroperitoneal mass that is homogenous and extending from the kidney to the pelvis and displacing the kidney superiorly.

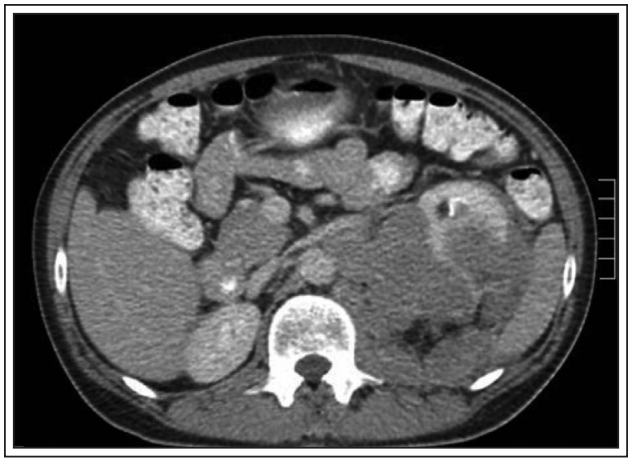

Figure 2. Transverse view of a contrast enhanced CT of the abdomen and pelvis showing a large left retroperitoneal mass displacing the left kidney anteriorly.

Discussion

In many cases, when imaging shows an enhancing large left-sided retroperitoneal mass, it is safe to assume a presumptive diagnosis of renal cell carcinoma (RCC) and to proceed with surgical resection of the tumor without the need for confirmatory testing (i.e. renal mass biopsy). In this case, based on the patient's clinical history and imaging findings, there was concern for a non-renal pathology, and a biopsy of the retroperitoneal mass was therefore performed, yielding a diagnosis of primary retroperitoneal DLBCL.

Multiple tumor types should be considered in the differential diagnosis of a retroperitoneal mass. Although symptomatology may vary with each individual tumor type, retroperitoneal tumors commonly present with vague symptoms or symptoms associated with other organ involvement because most retroperitoneal masses grow very large before they cause symptoms. Thus, differentiating between these diseases based on history and physical exam alone can be challenging. Laboratory findings often can be inconclusive with certain exceptions like non-seminomatous germ cell tumors. Thus, radiographic and histopathologic characteristics are often necessary to make a definitive diagnosis. Ultimately, the need to biopsy varies based on the clinical and radiographic presentations of the patient.

Certain radiographic features of a retroperitoneal mass may increase the likelihood of it being a lymphoma. The typical appearance of a retroperitoneal non-Hodgkin lymphoma (NHL) on CT is that of a homogeneous, poorly enhancing paraaortic or pelvic mass that extends between present structures without necessarily compressing or displacing them.[1] In addition, a factor that can allow

clinicians to differentiate from other retroperitoneal tumors include the fact that lymphomas will more than likely be homogenous on unenhanced and enhanced images. Necrosis and calcification are uncommon findings prior to the initiation of chemotherapy on CT, and involvement of mesenteric lymph nodes or non-nodal sites such as the liver, spleen, or bowel may also be noted on imaging.

Based on the patient's initial CT scans, there appeared to be a possibility that the patient in this case presentation had a primary renal lymphoma. Primary renal lymphomas are considered lymphoid malignancies originating in the renal parenchyma, not due to invasion from nearby masses of lymphoid origin. DLBCL appears to be the most common subtype of primary renal lymphoma.[2] The role of imaging is crucial in making the diagnosis of a renal lymphoma. Renal lymphomas most commonly present as multiple soft tissue masses, as opposed to a solitary renal mass, less than 3 cm in greatest diameter and have minimal enhancement compared to the adjacent renal parenchyma.[3] The frequency of solitary renal lymphomas has been reported to be less than 20%. They are usually homogenous and cause little to no mass effect on the renal parenchyma or collecting system. Minor compression may be noted with significantly larger masses; however, the reniform outline is mostly maintained. Some renal lymphomas may present as infiltrative tumors, resulting in an enlarged kidney with a normal renal contour. Contrast enhanced studies are especially important in these situations to identify areas of disease that exhibit poorly defined boundaries with the normal renal parenchyma. Associated retroperitoneal lymphadenopathy may also be seen in as much as 50% of cases. Renal lymphomas are not regularly associated with invasion of the renal hilum or sinus.

DLBCLs are the most common lymphomas worldwide, making up approximately 30% of all lymphoid malignancies.[4] Aggressive NHLs, including DLBCL, are classically known to present with systemic B symptoms such as unexplained fever, night sweats, fatigue, anorexia, weight loss, and lymphadenopathy. Abnormal laboratory findings may also be present, including cytopenias, hypercalcemia, hyperuricemia, elevated LDH levels, and elevated immunoglobulin levels, along with other variable laboratory findings more consistent with the specific organs involved in the spread of the disease. Of note, HIV is also a well-known risk factor for the development of lymphomas, including DLBCL.

The retroperitoneum is a fairly uncommon site of origin of lymphomas. In the largest study found, Nakayama et al published a case series regarding 50 retroperitoneal masses, 12 of which were ultimately diagnosed as being malignant lymphomas.[5] DLBCL also appears to be the most common subtype of retroperitoneal lymphoma. Chen et al described 32 cases of retroperitoneal hematologic malignancies, including 12 DLBCLs.[6] Of these 32 cases, the most commonly reported presentation was abdominal pain (n = 15), followed by back pain (n = 8). Retroperitoneal DLBCLs involving the genitourinary system have also been described in the literature. For example, Domazetovski et al described a case that presented with fever, malaise, and abdominal pain, ultimately culminating in acute renal failure.[7] Jaeger et al described a case that presented with flank pain and hematuria due to ureteral stricture,[8] while Cai et al described a similar case that presented with abdominal pain, renal colic, and hematuria despite relative sparing of the kidneys and ureters.[9] Of note, in the largest study found focusing specifically on primary retroperitoneal DLBCLs, Pileri et al reported nine cases but did not elaborate on each case's clinical presentation.[10]

In conclusion, a patient's past medical history, laboratory results, and imaging studies should all contribute to the evaluation/decision-making process in a patient with a retroperitoneal mass. When there is a high index of suspicion for lymphoma or the radiographic features are atypical, a biopsy of the mass should be performed prior to the initiation of definitive treatment (e.g. surgical resection). □

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