CASE REPORT

Primary Renal Lymphoma: A Case Report and Literature Review

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ABSTRACT

Primary renal lymphoma is very rare. The prognosis is usually poor with median survival less than a year. We report a case in a 55-year old man with a mass in the left kidney detected by ultrasonography and computed tomography. Radical nephrectomy was performed under a preliminary diagnosis of renal cell carcinoma. After the surgery, four courses of chemotherapy were performed. Prognosis may have improved since those reports in the earlier medical literature.

Keywords: Diagnosis, kidney neoplasms, lymphoma, prognosis

WIMJ Open 2014; 1 (1): 6

INTRODUCTION

Although secondary renal involvement from systemic lymphoma is very frequent, primary renal lymphoma (PRL) is a rare entity (1). Primary renal lymphoma represents 0.7% of extra nodal lymphomas, 0.1% of all malignant lymphomas and about 1% of all kidney neoplasms (2, 3). There are approximately 70 cases described in the worldwide medical literature (4). In primary renal lymphoma, there is an absence of clinical manifestations of lymphatic disease in other organs. It is difficult to diagnose, and has a poor prognosis. We present a case of PRL with review of the literature.

CASE REPORT

A 55-year old man presented to our institution with a two-month history of dull ache in the left flank. There was no previous history of weight loss and weakness and no fever or night sweat. There was no macroscopic haematuria. He had no relevant medical history and no family history of neoplasms. There was no significant lymphadenopathy on physical examination but he had percussion pain at the left flank. No palpable abdominal masses were present.

Laboratory investigations were unremarkable. Routine blood test results were: white blood cell count 4.31 × 10^9/L (neutrophils 66.1%, lymphocytes 25.3%, monocytes 5.8%, basophils 0.0%, eosinophils 2.8%), red blood cell count 4.7 × 10^{12}/L, haemoglobin 125 g/L and haematocrit 44%. The platelet count was 154 × 10^9/L. Admission serum electrolytes, blood urea nitrogen (BUN) and creatinine values were within normal limits but erythrocyte sedimentation rate (ESR) was mildly elevated (42 mm/h); the liver function tests were normal. Findings on urinalysis were unremarkable. While the chest X-ray was normal, an abdominal ultrasound revealed a normal pancreas and spleen but a large hypoechoic mass was noted arising from the lower pole of the left kidney (Fig. 1). No hydronephrosis or ureteral obstruction was detected and the contralateral kidney was sonographically normal. The patient then underwent a contrast computed tomography (CT) of the abdomen (Fig. 2), which revealed a solid, low density, homogenous left renal mass (7.2 cm × 6.8 cm × 6.0 cm) with mild contrast enhancement. Retroperitoneal or mesenteric lymph nodes were not enlarged. Addi-

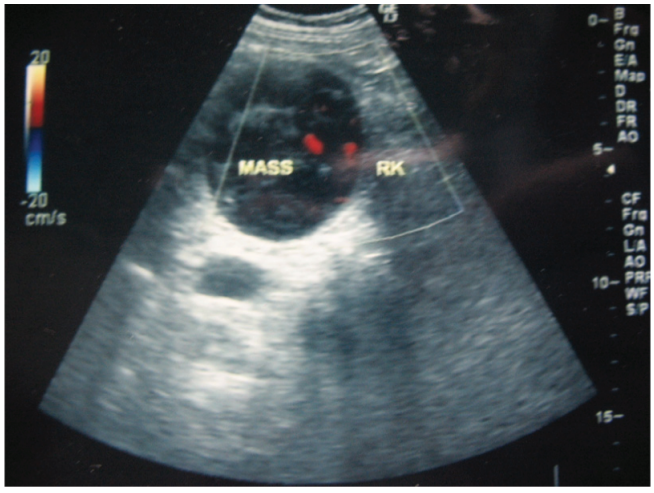

Fig. 1: Ultrasound reveals a relatively hypoechoic mass with uneven internal echo and blood flow signal.

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 DOI: 10.7727/wimjopen.2013.324

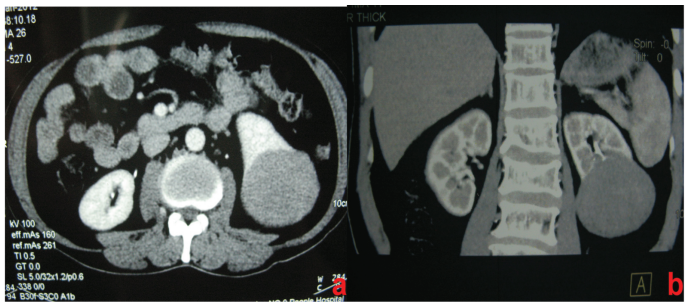

Fig. 2: Contrasted computed tomography (CT) reveals solid left renal mass with mild contrast enhancement. There is an absence of enlarged retroperitoneal or mesenteric lymph nodes.

tional imaging studies including CT scans of the thorax and brain as well as bone scintigraphy did not reveal metastatic lesions. Kidney neoplasm was suspected, but a preliminary characterization of CT-guided renal biopsy was not performed due to the risk of seeding in this malignant case. The patient underwent left radical nephrectomy for the presumed diagnosis of renal cell carcinoma.

Pathology biopsy of the left kidney specimen showed diffuse large lymphoid B-cells within the tumour (Fig. 3a). Immunohistochemical stains showed the following: the cells were positive for CD138 (++) [Fig. 3b], CD79α (++), Bcl-6

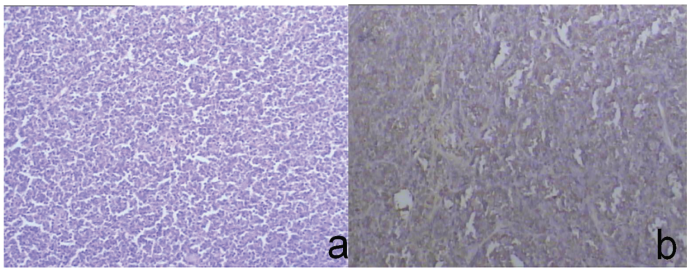

Fig. 3: (a) The renal mass specimen showing sheets of lymphoid cells with few scattered glomeruli within the tumour [haematoxylin and eosin stain × 100];
(b) Immunohistochemistry was positive for CD138 (++) [× 100].

(++), MUM-1 (++), MPO (++), CD43 (+), CD99 (+) and were negative for CD20 (-), CD5 (-), CD10 (-), PAX5 (-), ALK (-), RCC (-), CK (-), EMA (-) and S-100 (-), Ki67 was 30–40%. Based on these data, the favoured diagnosis was B-cell non-Hodgkin's lymphoma. Postoperatively, the patient was subjected to a bone marrow biopsy; all test results were negative. He received four courses of combination chemotherapy with cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisolone (CHOP). The patient has remained disease-free 23 months after surgery and 18 months after the end of the chemotherapy.

DISCUSSION

The existence of PRL has been questioned because the kidney does not contain lymphatic tissue. However, clinical reports support its existence (3, 5). Primary renal lymphoma most commonly affects older adults and the age of presentation is typically above 40 years. Primary renal lymphoma also seems to show a gender predisposition for males (6). Most cases are unilateral, although there have been reported instances of bilateral involvement. The clinical manifestations are similar to renal cell carcinoma: flank pain, haematuria, weight loss, abdominal mass and renal failure. Large B-cell lymphoma is the most common histologic type but small lymphocytic lymphoma is not unusual. The pathogenesis of PRL is not clear. The proposed pathogenetic mechanisms include: origin in the subcapsular lymphatics, seeding *via* the haematogenous route and lymphoid transformation of renal tissue subjected to chronic inflammation (7). As has been stated earlier, PRL is a rare disease and hence poses difficulty in diagnosis. In most reports, diagnosis was made after nephrectomy for suspected renal cell carcinoma. The application of modern imaging technology is the basis for the diagnosis of PRL. In particular, ultrasound and CT have certainly helped in the diagnosis. Diagnosis can be confirmed with the histology of the mass. Fine needle aspiration renal biopsy is the best method to establish diagnosis with high specificity and sensitivity (4). However, there are some risks of seeding in malignant tumour. The diagnosis of PRL can be made using the following criteria: (i) the presence of a renal mass confirmed as a lymphoma by pathology without extrarenal lymphomatous involvement, (ii) the absence of a leukaemic blood picture and abnormal bone marrow, and (iii) the absence of lymphadenopathy or hepatosplenomegaly (3). The index case achieved all the diagnostic criteria for identification of a PRL.

There is no standard treatment modality for PRL because of the small number of cases reported. The earlier limited literature reviews reported a poor prognosis. Survival was extremely low, less than one year (8). However, the recent case reports have suggested survival of more than one year (9). Our case has remained disease-free for 23 months. The preferred treatment for bilateral PRL is chemotherapy, with the CHOP regimen being the most effective. Surgery is preferred for unilateral PRL, and then supplemented by chemotherapy or radiotherapy (10)

Although PRL is a rare diagnosis in patients presenting with a renal mass, it must be taken into account in the differential diagnosis of any renal mass. We believe that delayed diagnosis is responsible for the poor prognosis. If diagnosed early, cure of PRL is possible.

In conclusion, primary renal lymphoma does exist. Improved recognition of the disease will help in earlier diagnosis and possibly improved prognosis.

REFERENCES

1. Ahmad AH, Maclennan GT, Listinsky C. Primary renal lymphoma: a rare neoplasm that may present as a primary renal mass. J Urol 2005; **173:** 239.
2. Tefekli A, Baykal M, Binbay M, Barut M, Muslumanoglu AY. Lymphoma of the kidney: primary or initial manifestation of rapidly progressive systemic disease? Int Urol Nephrol 2006; **38:** 775–8.

3. Stallone G, Infante B, Manno C, Campobasso N, Pannarale G, Schena FP. Primary renal lymphoma does exist: case report and review of the literature. J Nephrol 2000; **13:** 367–72.
4. Belbaraka R, Elyoubi MB, Boutayeb S, Errihani H. Primary renal non-Hodgkin lymphoma: an unusual diagnosis for a renal mass. Indian J Cancer 2011; **48:** 255–6.
5. Hart S, Ellimoottil C, Shafer D, Mehta V, Turk TM. A case of primary renal lymphoma. Urology 2012; **80:** 763–5.
6. Tuzel E, Mungan MU, Yorukoglu K, Basakci A, Kirkali Z. Primary renal lymphoma of mucosa-associated lymphoid tissue. Urology 2003; **61:** 463.
7. Omer HA, Hussein MR. Primary renal lymphoma. Nephrology 2007; **12:** 314–5.
8. Okuno SH, Hoyer JD, Ristow K, Witzig TE. Primary renal non-Hodgkin's lymphoma. An unusual extranodal site. Cancer 1995; **75:** 2258–61.
9. Pahwa M, Gupta N, Tyagi V, Chadha S. Primary renal lymphoma: is prognosis really that bad? Saudi J Kidney Dis Transpl 2013; **24:** 816–7.
10. Renaud J, Yartsev S, Dar AR, Van Dyk J. Successful treatment of primary renal lymphoma using image guided helical tomotherapy. Can J Urol 2009; **16:** 4639–47.

Submitted 09 Dec 2013
Accepted 21 Jan 2014
Published 15 May 2014
Online: http://myspot.mona.uwi.edu/wimjopen/67