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Primary Renal Lymphoma

Abdullah Ladha and Ghulam Haider

ABSTRACT

A 62-year-old male patient presented with right flank pain and right renal mass on CT scan. Patient left against medical advice and had nephrectomy done elsewhere. Histopathology revealed a diffuse large cell B-cell lymphoma. Patient presented again, with disseminated disease and was started on chemotherapy. Although a rare disorder, missing primary renal lymphoma as one of the causes of renal mass can lead to disseminated disease and unnecessary nephrectomy, in spite of chemotherapy being standard management.

Key words: Renal mass. Primary renal lymphoma. Chemotherapy. Nephrectomy.

INTRODUCTION

Approximately one-third of Non-Hodgkin Lymphomas (NHL) arise in tissues other than lymph node, spleen, Waldeyer ring and thymus, and are referred as primary extra-nodal NHL.¹ Primary Renal Lymphoma (PRL) is an extremely rare disease, it is defined as NHL arising in the renal parenchyma, not resulting from invasion of an adjacent lymphomatous mass.¹ Renal tissue usually lacks lymphoid tissue, therefore, it has been postulated that lymphatics in renal capsule might be the source of lymphoma, which subsequently invades renal parenchyma.² It accounts for 0.7% of all extra-nodal lymphomas in North America and 0.1% of all malignant lymphomas in Japan.^{3,4}

CASE REPORT

A 62-year-old male patient from Quetta presented with complaints of low-grade fever and dull, non-radiating right flank pain of three months duration. Fever was associated with chills and was continuous in nature. Past medical history was significant for diabetes mellitus, controlled on insulin. Patient had a previous history of Hepatitis B Virus (HBV) infection, completely treated by interferon. At presentation, the patient was sero-negative for HBV surface antigen and had negative PCR for HBV. Prior to admission, patient had received different medications from general practitioners based on ultrasound findings suggestive of paranephric fluid collection, but with no relief.

On physical examination, he appeared lethargic with blood pressure 125/75 mmHg and pulse rate of

112/minutes. Abdomen was prominently distended on right side with a large non-tender palpable mass in right lumbar region, which moved down on deep breathing. Rest of examination was unremarkable.

Initial laboratory investigations showed anemia with hemoglobin level of 10.2 gm/dl and hematocrit of 32.2. Lactic dehydrogenase level was 1840 u/l, renal and liver function tests were within normal limits; urine detailed report was unremarkable. Chest X-ray showed left hilar prominence. A CT scan of chest, abdomen and pelvis revealed enlarged right kidney with hypodense mass in it, loss of cortico-medullary differentiation, fluid collection in posterior para-renal area. It also revealed multiple left cortical cysts. The findings of CT scan suggested a neoplastic lesion and possibility of tuberculosis (Figure 1). A CT guided fine needle biopsy was planned but patient refused and went home against medical advice.



Figure 1: Abdominal CT scan images showing hypodense renal mass involving the right kidney with perinephric extension and collection.

A few weeks later, patient's complaints worsen and he was taken to another hospital where right nephrectomy was performed. Subsequent histopathology revealed a Diffuse Large B-cell Lymphoma (DLBCL) of the kidney.

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Patient was referred to our centre for further staging and chemotherapy. A repeat CT of chest, abdomen and pelvis revealed newly developed multiple soft tissue nodules in both lungs, largest nodule measuring 3 x 3 cm in right lung. It also revealed bilateral loculated pleural effusion and evidence of right nephrectomy. MRI of brain, CSF examination and bone marrow examination revealed no involvement by lymphoma at that time. Patient was subsequently staged as III-B and was started on R CHOP chemotherapy. Initially, the patient responded well with stable pulmonary disease and no evidence of disease in other regions on CT scans. After about one year, the patient presented with the complaints of altered mental status. At that time, MRI of brain revealed a brain lesion for which patient was started on radiotherapy to brain.

DISCUSSION

The overall percentage of NHL originating in extra-nodal site is between 25% and 35% in most countries, most common primary extra-nodal sites are represented by stomach, skin, small intestine, breast, thyroid, lung, heart, liver, genitourinary tract, adrenals and bone.⁵ Previous reports have described flank pain to be most common presenting symptom,^{6,7} which was also present in this patient. Other reported presenting complaints include abdominal mass, hematuria and systemic complaints such as weight loss, fever and fatigue.⁷ In previous studies, parenchymal involvement of bilateral kidneys has been associated with renal insufficiency.⁶ This patient had one kidney affected and did not present with renal insufficiency.

Diffuse Large B-cell Lymphoma (DLBCL) is the largest sub-type of NHL, comprising 30% of all lymphomas in the West.⁸ A large population-based study from the registry of Danish lymphoma group found that 40% of DLBCL had extra-nodal presentation.⁹ This patient also had DLBCL, which has been frequently reported as most common of PRL in other reports.^{6,7} In this patient, the primary renal DLBCL rapidly metastasized to lung and brain within a few months, resulting in poor prognosis, likewise investigators from all over world have reported poor prognosis in primary renal DLBCL.

Cases from Japan reported less than 2 years survival inspite of treatment and localized disease.⁷ Similarly, cases from the United States have reported a median time of survival of 8 months only.¹ Some investigators have also reported a comparatively favourable prognosis of more than 3 years with early diagnosis and systemic chemotherapy.¹⁰

Standard management of a renal mass is nephrectomy. However, primary renal lymphoma is an exception in which patient can be treated with systemic chemotherapy like R-CHOP. Therefore, in spite of its uncommon occurrence, it is of immense importance to distinguish primary renal lymphoma from renal carcinoma and an infectious disease as a cause of renal mass. An early diagnosis and management can help to improve outcome in patients with primary renal lymphoma.

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