Case Report

Primary Renal MALT Lymphoma: A Conservative Approach to a Rare Disease

Phuong T. Ngo*, Harsh Parmar, and Arthur Topilow
Department of Medicine, Jersey Shore University Medical Center, USA

Abstract

Primary renal MALT lymphoma, a rarity, is an important diagnostic consideration for renal masses. It is an indolent disease that presents with nonspecific symptoms and is grossly similar to renal cell carcinoma on imaging. Management of the reported cases has been variable. There are currently no standardized treatment guidelines. We present a case of an 80-year-old man who was found to have a primary renal MALT lymphoma who underwent a partial nephrectomy without subsequent immunotherapy, chemotherapy or radiation therapy. A number of previous case reports also took a more conservative approach with surgery only and our patient, like theirs, is currently doing well without recurrence of disease. We present this case, therefore, to suggest approaching these low grade lymphomas with surgery only, without subjecting patients to further systemic treatments.

ABBREVIATIONS

MALT: Mucosa-Associated Lymphoid Tissue; CT: Computed Tomography; CHOP: Cyclophosphamide,Hydroxydoxorubicin, Oncovine and Prednisone; R-CHOP: Rituximab, Cyclophosphamide,Hydroxydoxorubicin,Oncovine and Prednisone

INTRODUCTION

Low grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT), first described by Isaacson and Wright in 1983, is now recognized as a distinct subtype in the classification of Non-Hodgkin's lymphoma [1]. They are further subdivided into those arising in the stomach and those arising in non-gastric areas, commonly the small intestine, salivary glands, head and neck, lung, thyroid, breast, gallbladder, cervix and ocular adnexa [2,3]. The kidneys are commonly involved in systemic disease but rarely are they a primary site of lymphoma. Primary renal MALT lymphoma was first described by Colovic in 1999 and since then, to the best of our knowledge, there have been 26 other reported cases. Treatment has varied amongst these cases, including partial and radical nephrectomy, chemotherapy, immunotherapy and radiation. We present a patient who was incidentally found to have a primary renal MALT lymphoma that then underwent a left partial nephrectomy without further systemic treatment and is clinically well two years post surgery. Therefore, in addition to being an important differential for renal masses, primary renal MALT lymphomas can perhaps be treated with surgery alone.

CASE PRESENTATION

An 80-year-old male with recent resection of a malignant carcinoid in the colon presented for a routine follow up with report of decreased appetite, nocturia, weight loss and changes in his bowel habits since his colon resection. He denied flank pain, dysuria, hematuria, incontinence and hesitancy. Physical exam showed abdominal scars from his colon resection and hernia repairs but was otherwise unremarkable without palpable masses. Laboratory data showed chronic microcytic anemia and worsening acute kidney injury with creatinine of 1.76mg/dL from a baseline of 0.9-1.0mg/dL 6 months prior. He had a non-contrast computed tomography (CT) of the abdomen and pelvis for follow up of his colonic resection and was found to have a 36 mm exophytic lesion in the lower pole of the left kidney with soft tissue attenuation suspicious for renal cell carcinoma (Figure 1).

He then underwent a laparoscopic left partial nephrectomy. The specimen was 3.5 x 2.5 x 2.5cm unencapsulated solid mass with a pale pink to grey-white smooth, rubbery cut surface. There was a small secondary nodule that was loosely attached to the surface of the larger nodule. Histologic exam revealed a small cell, B-cell lymphoma with extensive plasmacytic differentiation, which was consistent with extranodal marginal zone lymphoma of mucosal associated lymphoid tissue (MALT lymphoma). Flow cytometry showed monoclonal B-cell population with a large cell component with light chain restriction expressing CD19, CD20, CD23 and CD11c (Figure 2). He then underwent a bone marrow biopsy that was negative for lymphoma, confirming the MALT lymphoma was isolated to the left kidney. He received no further systemic treatment and is doing well 2 years after surgical treatment.
DISCUSSION

MALT lymphomas are a subtype of Non-Hodgkin’s lymphoma that arise in a variety of epithelial tissues but very rarely do they arise in the kidneys primarily. These marginal zone lymphomas typically occur in the setting of chronic stimulation of the immune system by reactive B-cell lymphoid tissue. For example, lymphomas in the stomach are associated with Helicobacter pylori resulting in chronic gastritis, lymphomas in the salivary gland are associated with Sjogren’s syndrome, and those of the thymus are associated with chronic thyroiditis [3]. Renal MALT lymphomas are again an anomaly in this regard, having no specific predisposing inflammatory condition. Khalil et al., conducted a population-based study on the incidence of marginal zone lymphomas in the United States from 2001-2009 and showed the overall rate of lymphomas increased with age [4]. Extrapolating from that, renal MALT lymphomas are perhaps due in part to the overall chronic inflammation and immune senescence that come with aging [5]. However, though the majority of reported cases involved elderly patients, Kato, et al reported a case of primary renal lymphomas arising in a 30-year-old male who presented with frequent urination and right lower abdominal pain [6].

The symptoms of renal MALT lymphomas are also seemingly nonspecific. The first case reported by Colovic et al., involved a 50-year-old man being treated for resistant hypertension who was incidentally found to have a right renal mass on routine ultrasound, and since then, cases have ranged from asymptomatic patients to those with flank pain, weight loss and hematuria. Our patient had weight loss with decreased appetite and nocturia as well as changes in bowel habits since his colon resection. His symptoms may have been due to the lymphoma, but they also could have been the result of his colon resection. The diagnosis of renal MALT lymphomas, therefore, has relied on imaging. Once

Table 1: Summary of previously reported cases.

<table>
<thead>
<tr>
<th>Author</th>
<th>Patient</th>
<th>Symptoms</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Parveen et al [3]</td>
<td>69F</td>
<td>right upper quadrant abdominal pain</td>
<td>nephrectomy, local radiation</td>
<td>N/A</td>
</tr>
<tr>
<td>Colovic et al [7]</td>
<td>50M</td>
<td>hypertension follow up</td>
<td>nephrectomy</td>
<td>N/A</td>
</tr>
<tr>
<td>Mhawech et al [8]</td>
<td>76F</td>
<td>right flank pain, anorexia, weight loss and hypertension</td>
<td>radical nephrectomy</td>
<td>N/A</td>
</tr>
<tr>
<td>Jindal et al [9]</td>
<td>45M</td>
<td>fever and abdominal pain</td>
<td>radical nephrectomy and chemotherapy</td>
<td>multiple relapses in 13 yrs</td>
</tr>
<tr>
<td>Tuzel et al [10]</td>
<td>43M</td>
<td>N/A</td>
<td>radical nephrectomy</td>
<td>no recurrence after 28 mos</td>
</tr>
<tr>
<td>Mortlock et al [11]</td>
<td>84F</td>
<td>lower extremity edema, shortness of breath, weight loss</td>
<td>chemotherapy, not specified</td>
<td>N/A</td>
</tr>
<tr>
<td>Qiu et al [12]</td>
<td>83F</td>
<td>back pain</td>
<td>rituximab</td>
<td>no recurrence after 8 mos</td>
</tr>
<tr>
<td>Qiu et al [12]</td>
<td>53M</td>
<td>asymptomatic</td>
<td>partial nephrectomy</td>
<td>no recurrence after 10 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>77F</td>
<td>N/A</td>
<td>N/A</td>
<td>no recurrence at 36 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>65M</td>
<td>N/A</td>
<td>antibiotics for renal actinomycosis</td>
<td>no recurrence at 14 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>54M</td>
<td>N/A</td>
<td>chemotherapy</td>
<td>no recurrence at 36 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>75M</td>
<td>N/A</td>
<td>nephrectomy, chemotherapy</td>
<td>no recurrence at 12 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>66M</td>
<td>N/A</td>
<td>nephrectomy</td>
<td>no recurrence at 53 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>83M</td>
<td>N/A</td>
<td>chemotherapy</td>
<td>no recurrence at 9 mos</td>
</tr>
<tr>
<td>Garcia et al [13]</td>
<td>65F</td>
<td>N/A</td>
<td>nephrectomy</td>
<td>N/A</td>
</tr>
</tbody>
</table>
diagnosis has been established, the treatment plans for these renal MALT lymphoma patients have differed. In cases of gastric MALT lymphomas, therapy is aimed at eradicating *H. pylori*. Those who fail this initial treatment are then treated with localized radiation. Non-gastric MALT lymphomas are also similarly treated with surgery and local radiation with or without chemotherapy and immunotherapy. For primary renal MALT lymphomas, however, standardized therapeutic strategies have not yet been established. Of the 26 reported cases of renal MALT lymphomas, 15 had nephrectomy, 9 received chemotherapy, 1 was treated with only radiation and 1 with only rituximab. While the majority of patients who received chemotherapy and radiation had no recurrence, those who only had nephrectomy also have not had recurrence, bringing to question what the treatment of renal MALT lymphomas should be (Table 1). From the cases reviewed, only one patient had multiple recurrences, and he was initially treated with both nephrectomy and chemotherapy, showing nephrectomy alone may not be enough and adjuvant chemotherapy does not guarantee definitive treatment. Furthermore, our follow up of two years may not be sufficient time to see recurrence, let alone the few months used in the reviewed cases. However, the majority of patients who were treated conservatively with surgery alone appear to have fared as well as those who received additional systemic treatment.

**CONCLUSION**

Several cases of primary renal MALT lymphomas seem to have been treated successfully with surgery alone. Our patient similarly is doing well two years status post nephrectomy without further treatment, suggesting that these low grade lymphomas might be treated without additional systemic therapy.

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**REFERENCES**


