Mucosa-associated lymphoid tissue lymphoma involving the kidney: a case report and review of the literature

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Abstract Mucosa-associated lymphoid tissue (MALT) lymphomas arise from various anatomic sites, mostly observed in the gastrointestinal tract; however, involvement of the kidney is extremely rare. We report a case of MALT lymphoma involving the kidney in a 70-year-old man. Radical nephrectomy was performed under the diagnosis of renal cell carcinoma preoperatively. However, the renal specimen showed that the diffuse small- to medium-sized lymphocytes were scattered and formed colonies, and the neoplastic lymphoid cells were positive for CD20, CD79 α , and Bcl-2, but negative for CD10 and Cyclin D1; therefore, the final histological diagnosis was MALT lymphoma involving the renal pelvis. Although he was referred to the department of hematology, no additional treatment was given postoperatively, and he has survived for 4 months without evidence of a recurrence of lymphoma at the last follow-up. To the best of our knowledge, to date 37 reports of MALT lymphoma involving the kidney have been published in the literature. We report a case of MALT lymphoma involving the kidney, and review the existing literature.

Keywords MALT lymphoma · Kidney · Nephrectomy

Introduction

Low-grade B-cell lymphoma of mucosa-associated lymphoid tissue (MALT) was first reported by Isaacson [1]. MALT lymphomas arise from various anatomic sites, but the involvement of the kidney is extremely rare [2]. Pelstring et al. reported the first case of renal MALT lymphoma in 1991 [3]. To the best of our knowledge, to date 37 reports of MALT lymphoma involving the kidney have been published in the literature. MALT lymphoma is such a low grade of B-cell lymphoma that the prognosis is relatively good [4, 19], but the management of MALT lymphoma arising from the kidney has not been established.

We report a case of MALT lymphoma involving the kidney and describe the clinical feature reported in existing literature.

Case report

A 70-year-old man was referred to our department of urology because a left renal mass was detected by computed tomography (CT) scan. His medical history was unremarkable, and he had no notable physical findings except for left back pain. Laboratory findings revealed only elevated levels of serum creatinine (1.10 mg/dl). The urine cytological examination result was negative (classII). A contrast-enhanced CT scan showed an infiltrative hypovascular mass measuring 3.0 × 2.5 cm arising from the left renal pelvis with para-aortic lymph node enlargement (Fig. 1). Left retrograde pyelography showed normal findings and cytology of the ureteropelvic urine was negative (classII) (Fig. 2). Positron emission tomography (PET) scan revealed intense fluorine-18-fluorodeoxyglucose (FDG) uptake [maximum

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Fig. 1 A contrast-enhanced CT scan of the abdomen reveals an enlarged hypodense soft infiltrative mass from the renal pelvis to the parenchyma of the left kidney (*arrow*) and an enlarged lymph node in the para-aortic (*arrowhead*)

Fig. 2 Left retrograde pyelography shows normal findings and cytology of the ureteropelvic urine was negative (class II)

standardized uptake value (SUVmax) = 2.9] in the para-aortic lymph node (Fig. 3).

At this point, we could not diagnose whether the patient had renal carcinoma or another renal malignancy such as atypical renal cell carcinoma (RCC) including the papillary and chromophobe subtypes, or renal pelvis cancer, and so on. However, we suspected atypical RCC because of the point that the urine cytological examination result was negative, and retrograde pyelography showed normal findings, and PET/CT did not show intense FDG uptake in renal mass, and thus, we performed radical nephrectomy and para-aortic lymph node dissection extraperitoneally.

Fig. 3 These are images of an 18-FDG PET-CT scan. An 18-FDG uptakes in the para-aortic lymph node (SUVmax = 2.9)

Grossly, the specimen contained a border indistinctive mass infiltrating the renal pelvis and the para-aortic lymph node was swollen. On histological examination, diffuse small- to medium-sized lymphocytes were scattered and formed colonies showing plasmacytic differentiation partially in the renal pelvis, parenchyma, and the lymph node (Fig. 4). The neoplastic lymphoid cells were positive for CD20, CD79 α , and Bcl-2, but negative for CD10 and Cyclin D1 (Fig. 5). These findings supported a diagnosis of MALT lymphoma arising in the kidney. No additional treatment was given postoperatively by the department of hematology.

Discussion

Primary renal lymphoma (PRL) is defined as lymphoma arising in the renal parenchyma, not resulting from the invasion of an adjacent lymphomatous mass [5]; however, in other studies, concurrent lymph node involvement alongside a solitary renal mass is accepted as PRL [6]. Thus, the diagnosis of PRL is challenging. Most PRLs are classified as intermediate- and high-grade B-cell lymphomas, such as diffuse large B-cell lymphoma [7], but low-grade PRLs such as MALT lymphoma are extremely rare. MALT lymphoma arises from various anatomic sites, such as the gastrointestinal tract, ocular adnexa, breast, respiratory tract, salivary gland, thymus, thyroid, skin, gall

Fig. 4 The tumor is composed of diffuse small- to medium-sized lymphocytes scattered and forming colonies showing plasmacytic differentiation partially in the renal pelvis and lymph node. Hematoxylin-eosin (H&E) staining, magnifications: **a** ($\times 10$), **b** ($\times 40$), **c**, **d** ($\times 400$)

bladder, and uterine cervix, but primary MALT lymphoma of the kidney is extremely rare [2, 17, 18, 21, 26]. MALT lymphoma is usually associated with chronic inflammation, such as *Helicobacter pylori* infection, or with autoimmune disease such as Sjogren's syndrome or Hashimoto thyroiditis [7, 8, 20]. The etiology of PRL is not yet clear, because the kidney does not have lymphoid tissue. In contrast, it is reported that renal MALT lymphoma coexists with nephritis, sarcoidosis, or renal actinomycosis [7, 24]; thus, these chronic inflammations have been suggested as possible etiological factors [9]. Although the diagnosis of PRL is extremely challenging, it is essential to distinguish PRLs from other malignancies such as RCC and renal pelvic carcinoma. In their imaging patterns, PRLs are often large infiltrative renal tumors that extend into the perinephric space and retroperitoneum. Further, PRLs are typically hypovascular and do not invade the vessels. Careful evaluation to ensure that the epicenter of the tumor is not within the collecting system is helpful in

differentiating PRL from urothelial carcinoma [10]. PET/CT may be useful in the diagnosis because renal lymphoma is often intensely FDG-avid, but MALT lymphoma is still a matter of debate, and conflicting results have been reported [11]. In contrast, RCC including the papillary and chromophobe subtypes may not show intense FDG uptake [12]. In brief, the use of imaging alone is not sufficient to distinguish between lymphoma and other renal malignancies. Therefore, a definitive diagnosis usually requires biopsy, wherever possible, to prevent an unnecessary nephrectomy. The pathological feature of MALT lymphoma is that diffuse small- to medium-sized lymphocytes are scattered and shows nodules-like structure with positive for Bcl-2, but negative for CD10 and Cyclin D1 in immunohistochemical analysis [13].

In general, the therapeutic strategies for MALT lymphoma vary depending on extension of disease. For localized disease, local treatment (either radiotherapy or surgery) or single agent such as rituximab is suggested. For

◀**Fig. 5** Immunohistochemistry analysis shows that the neoplastic lymphoid cells were positive for CD20, CD79 α , and Bcl-2, but negative for CD10 and Cyclin D1

disseminated disease, single agent such as rituximab or polychemotherapy such as R-CHOP (rituximab, vincristine, doxorubicin, cyclophosphamide, prednisolone) or

Table 1 Summary of the clinical features of MALT lymphoma involving the kidney

References	Sex/ age (years)	Location	Associated disease	Specimen	Treatment	Follow-up
Pelstring et al. [3]	62/F	Kidney, parotid gland	Sjogren syndrome	NA	Chemotherapy	AWD, 15 months
Parveen et al. [2]	69/F	Kidney, perirenal LN	None	Nephrectomy	Nephrectomy, irradiation	ANED, 17 months
Imahori [17]	56/M	Kidney, orbit	None	NA	Nephrectomy	Died with lymphoma, 13 years
Mak et al. [18]	62/M	Kidney, GI tract, tonsils	IgA nephropathy	Biopsy	Chemotherapy	ANED, 20 months
Kazuhiro et al. [19]	68/M	Kidney, prostate, salivary glands	None	NA	No	AWD, 5 years
Colovic et al. [20]	50/M	Kidney	HP gastritis	Nephrectomy	Nephrectomy	NA
Tao and Khan [21]	9/M	Kidney, axillary LN, lung	Epstein-Barr virus infection	Biopsy	Chemotherapy	Died with lymphoma, 4 years
Mhawech et al. [22]	76/F	Kidney	None	Nephrectomy	Nephrectomy	NA
Jindal et al. [23]	45/M	Kidney	None	Nephrectomy	Nephrectomy, chemotherapy	Alive, 13 years with multiple relapses
Stokes et al. [24]						
Case 1	68/F	Kidney	MPGN	Biopsy	Prednisone	NA
Case 2	72/F	Kidney	MPGN	Biopsy	Prednisone	AWD, 6 months
Mita et al. [25]	77/M	Kidney	None	NA	Nephrectomy	NA
Tuzel et al. [26]	43/M	Kidney	None	Nephrectomy	Nephrectomy	ANED, 28 months
Laven et al. [27]	51/M	Kidney, bone marrow	Renal cysts	Partial nephrectomy	Partial nephrectomy	ANED, 12 months
Mortlock et al. [28]	84/F	Kidney, para-aortic LN	Systemic lupus erythematosus	NA	Chemotherapy	NA
Qiu et al. [7]						
Case 1	83/F	Kidney, perirenal fat	None	Biopsy	Chemotherapy	ANED, 8 months
Case 2	53/M	Kidney	Sarcoidosis	Partial nephrectomy	Partial nephrectomy	ANED, 10 months
Case 3	72/M	Kidney, orbit	None	Nephrectomy	Nephrectomy	NA
Garcia et al. [16]						
Case 1	77/F	Kidney	None	Biopsy	NA	ANED, 36 months
Case 2	65/M	Kidney	Renal actinomycosis	Biopsy	Antibiotics	ANED, 14 months
Case 3	54/M	Kidney	None	Biopsy	Chemotherapy	ANED, 36 months
Case 4	75/F	Kidney	TCC, bladder	Nephrectomy	Nephrectomy, chemotherapy	ANED, 12 mo

Table 1 continued

References	Sex/ age (years)	Location	Associated disease	Specimen	Treatment	Follow-up
Case 5	66/M	Kidney	Subsequent RCC, contralateral kidney	Nephrectomy	Nephrectomy	ANED, 53 months
Case 6	83/M	Kidney	None	Biopsy	Chemotherapy	ANED, 9 months
Case 7	65/F	Kidney, retroperitoneal LN	HP gastritis	Nephrectomy	Nephrectomy	AWD, 6 months
Case 8	73/M	Kidney, bone marrow	Adenocarcinoma, colon	Biopsy	Chemotherapy	AWD, 9 months
Case 9	47/M	Kidney, soft tissue of flank and thigh	None	Biopsy	Chemotherapy; ASCT	ANED, 42 months
Case 10	18/F	Kidney	HP gastritis	Biopsy	Chemotherapy	ANED, 10 months
Kato et al. [29]	30/M	Kidney	Nephrotic syndrome	Nephrectomy	Nephrectomy	ANED, 21 months
Li et al. [30]						
Case 1	48/F	Kidney	Chronic pyelonephritis	NA	Nephrectomy	NA
Case 2	55/F	Kidney	Chronic pyelonephritis	NA	Nephrectomy	NA
Peces et al. [31]	77/M	Kidney, bone marrow	Peptic ulcer, esophagitis	Nephrectomy	Nephrectomy, chemotherapy	Died with lymphoma, 37 months
Dadekam et al. [32]	77/M	Kidney	None	Biopsy	Radiation	ANED, 3 years
Otsuki et al. [33]	69/M	Kidney, upper ureter	None	Nephroureterectomy	Nephroureterectomy, chemotherapy	ANED, 78 months
Chi et al. [34]	72/F	Kidney, bone marrow	Waldenstrom macroglobulinemia, chronic kidney disease	Biopsy	Chemotherapy	NA
Asgari et al. [35]	74/F	Kidney, upper ureter	Renal calculus	Nephrectomy	Nephrectomy	NA
Niwa et al. [36]	86/F	Kidney, renal pelvis	None	Nephroureterectomy	Nephroureterectomy	ANED, 4 months
This case	70/M	Kidney, para- aortic LN	None	Nephrectomy	Nephrectomy	ANED, 4 months

ANED alive with no evidence of disease, ASCT autologous stem cell transplantation, AWD alive with disease, GI gastrointestinal, HP helicobacter pylori, LN lymph node, MPGN membranoproliferative glomerulonephritis, NA not available, RCC renal cell carcinoma, TCC transitional cell carcinoma

R-FC (rituximab, fludarabine, cyclophosphamide) is suggested [14]. In contrast, the therapeutic strategies for MALT lymphoma arising from the kidney are controversial. Therefore, nephrectomy or chemotherapy seems to be performed taking the existing reports into account.

The prognosis for patients with renal MALT lymphoma is reported to be good with chemotherapy, surgical intervention, or radiation [15, 17, 23]. There were variations in survival with regard to MALT lymphomas depending on their anatomical sites. The 5-year overall survival for MALT lymphoma arising in the genitourinary tract was 75.6 %, whereas that for those with different anatomical origins ranged from 69.1 to 87.9 % [4].

To the best of our knowledge, 37 cases of MALT lymphoma involving the kidney have been reported, and the clinical findings are summarized in Table 1. Although Garcia reported a series of 10 cases, which was the literature with the largest sample [16], the majority were case reports. The mean patient age was 62.6 years (9–86 years) and the mean follow-up duration was 33.6 months (3–156 months). There were 22 men and 16 women. Nineteen patients had lymphoma localized only to the kidney (PRL) and 19 patients had evidence of systemic disease. As a location except the kidney, it was relatively frequent involved in bone marrow and para-aortic lymph node. Twenty patients had other diseases in addition to

MALT lymphoma, such as *Helicobacter pylori* gastritis and sarcoidosis. A biopsy was performed for 8 patients, who had atypical mass localized only to the kidney (no. 10, 11, 19, 20, 21, 24, 28, 33 from the top in the Table 1). Of 8 patients, 3 patients underwent chemotherapy, and 5 patients underwent various therapies without nephrectomy. In regards with treatment, 15 patients underwent only nephrectomy, 10 patients received only chemotherapy, and three patients received nephrectomy followed by chemotherapy. The other 10 patients underwent various treatments such as the administration of antibiotics, prednisone, radiation, or nephroureterectomy. Ten of 38 patients were alive without evidence of disease and 6 patients were alive with disease at the end of the observation duration, and 3 patients died with lymphoma. Of 3 patients, they died after diagnosis at 3, 4, and 13 years, respectively. The follow-up data of other 10 patients were not available.

As mentioned above, in most cases, diagnosis is made after preoperative radical nephrectomy for suspected RCC. When nephrectomy is performed earlier, most patients do not receive additional treatment. On the other hand, when diagnosed after a biopsy of renal mass with another lesion other than kidney, patients are often treated by chemotherapy alone. Therefore, diagnosis can be confirmed with a biopsy, which prevents an unnecessary nephrectomy. It may be that nephrectomy is overtreatment considering the outcome between the patients receiving nephrectomy and chemotherapy. In our case, the infiltrative mass was located from the renal pelvis to the parenchyma and covered the renal artery. Therefore, we were technically hampered in performing the biopsy, and performed radical nephrectomy and para-aortic lymph node dissection for suspected atypical RCC preoperatively, and no additional treatment was given by our department of hematology. I wonder if there are methods that a lymphoma is diagnosed besides renal biopsy? A bone marrow biopsy may be useful, because there are a few patients whose bone marrow examination shows atypical cells [20, 29, 32, 35].

In summary, we have described a case report and reviewed the literature. Although MALT lymphoma is extremely rare in patients with a renal mass, early diagnosis and proper treatment result in a favorable outcome. Therefore, in the case of the atypical renal mass, a renal biopsy may be useful when taking a renal lymphoma into account; it may prevent an unnecessary nephrectomy and also avoid delay in starting the appropriate treatment.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Research involving human participants and/or animals For this type of study formal consent is not required.

Informed consent Informed consent was obtained from all individual participants included in the study.

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