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CASE REPORT

Renal MALT lymphoma associated with Waldenström macroglobulinemia

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Mucosa associated lymphoid tissue lymphoma (MALT lymphoma) is mostly seen in the gastrointestinal tract; origin from the kidney is extremely rare. Waldenström macroglobulinemia (WM) is a clinicopathologic syndrome denoted by the presence of monoclonal gammopathy in the serum, typically caused by lymphoproliferative disorder. Literature review did not find any report of renal MALT lymphoma accompanied by WM. Herein, for the first time, we report a 72 year-old female patient with a history of chronic kidney disease, presenting with solitary renal mass; MALT lymphoma was confirmed by pathological examination. A serology study identified the presence of WM. No manifestation of hyperviscosity syndrome was noted. Bone marrow biopsy disclosed the concurrent systemic involvement. Her treatment response was uneventful and the renal mass responded with regressive change in size after chemotherapy. The renal function remained stable during follow-up. MALT lymphoma should be considered as an underlying pathology of isolated renal mass. Furthermore, patients with MALT lymphoma should be screened for Waldenström macroglobulinemia and hyperviscosity syndrome.

Conflicts of interest: The authors have no conflicts of interest relevant to this article.

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Introduction

Marginal zone B cell lymphoma, a distinctive group of non-Hodgkin's lymphoma, is divided into three subtypes, nodal, extranodal and splenic, depending on the involved location. In general, these lymphoma subtypes have an identical microscopic morphology. Among them, extranodal marginal zone B cell lymphoma, also named as mucosa associated lymphoid tissue (MALT) lymphoma, accounts for about 5% of non-Hodgkin's lymphoma, and usually involves many anatomic sites, mostly confined to stomach.[1] MALT lymphoma of renal origin is very rare, and only sporadic cases have been reported.[2,3]

Waldenström macroglobulinemia (WM) is characterized by the presence of lymphoplasmacytic lymphoma in the bone marrow, with the appearance of monoclonal gammopathy, mostly immunoglobulin (Ig) M in the serum.[4] WM often presents with nonspecific symptoms such as weakness, fatigue and weight loss. However, severe complications can be caused by WM-associated hyperviscosity syndrome. We encountered a female patient who presented with an incidental finding of renal mass. Pathological examination revealed MALT lymphoma. Further work-up revealed bone marrow involvement, and abnormal IgM was noted in her serum. She experienced an uneventful treatment course with chemotherapy only.

Case report

On February 2009, a routine renal echogram in a 72-year-old female patient with a medical history of chronic kidney disease for 5 years, found a 5.8 × 5.4 cm mix-echoic mass over the right middle kidney. The patient was asymptomatic and physical examination showed no specific finding. Her laboratory test was essentially normal, except for mild elevation of serum creatinine (Cr). Her results showed: Cr = 1.94 mg/dL, white blood cell count = 5800 cells/μL, hemoglobin = 12.2 g/dL, calcium = 9.4 mg/dL, uric acid 8.2 = mg/dL, lactate dehydrogenase = 187 u/L). Urine examination was unremarkable. Further study with abdominal computed tomography (CT) disclosed an ill-defined homogenous soft tissue mass, with mild enhancement, measuring 6.4 × 5.6 cm over the right kidney (Fig. 1). No definite enlargement of the retroperitoneal lymph node or pelvic side wall invasion was found. CT-guided biopsy was thus performed to obtain a pathological diagnosis.

Macroscopically, there was fibrovascular tissue diffusely infiltrated by small mature lymphoid cells (MALT lymphoma, Fig. 2). Microscopic examination found neoplastic cells composed of small lymphoid cells, with round or slightly irregular contours and abundant cytoplasm.

Immunohistochemical stain showed a positive result for CD20, and was negative for CD5, CD10, CD15, CD30, and CD23. Serum protein electrophoresis and immunofixation studies found the presence of monoclonal IgM kappa paraproteinemia. The patient's IgA, IgG, and IgM levels were 83.9 mg/dL, 1840 mg/dL, and 569 mg/dL, respectively. The total globulin level was 3.54 g/dL. Urine protein electrophoresis and immunofixation studies did not detect significant results. A subsequent bone marrow biopsy was performed, and a normal white blood cell count, with 10% plasmacytoid lymphocytes with CD-20 positive lymphoid cells, was noted. She then received chemotherapy a regimen of Leukeran and prednisolone for 8 months. No treatment-related complications developed. Gradual regressive change of renal mass, as evidenced by a series of CT scans, was noted. The IgM level declined gradually to as low as 145 mg/dL. Her renal function was rather stable during the follow-up period. Surgical interventions, such as nephrectomy, were not performed.

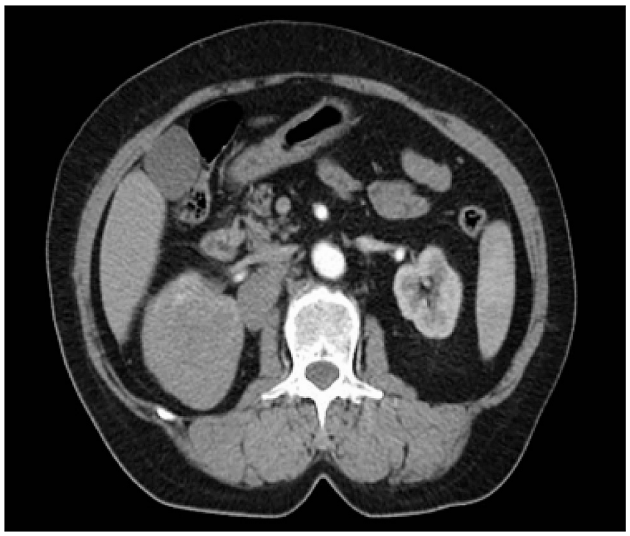

Figure 1 CT scan showed an ill-defined homogeneous mass over right kidney, measuring 6.4 × 5.6 cm. No definite enlargement of retroperitoneal lymph node or pelvic side wall invasion was found.

Discussion

Primary renal tumor can be benign or malignant; most solid masses involving kidneys are malignant. Renal cell carcinoma is responsible for 80% to 85% of all primary renal

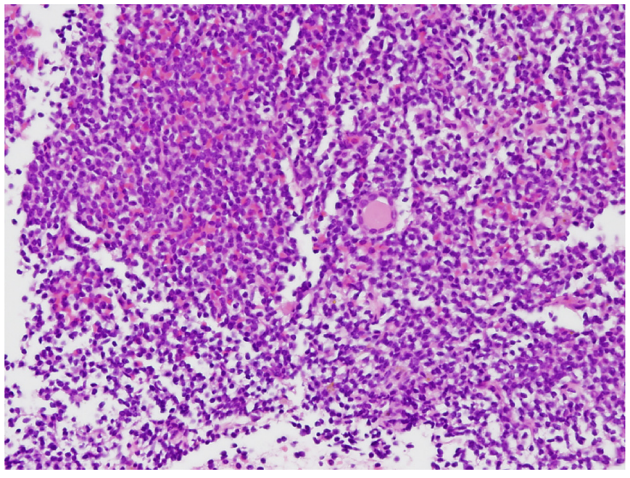

Figure 2 Histological picture of renal tissue showed fibrovascular tissue diffusely infiltrated by small mature lymphoid cells.

malignant tumors, followed by transitional cell carcinoma.[5] Renal lymphoma is rare, and is usually clinically silent. Flank pain, weight loss, hematuria or a palpable mass are the most common signs and symptoms. For most reported renal MALT lymphomas, clinical features varied widely and there was no specific presentation suggesting this rare renal tumor.[1] Although a variety of diagnostic tools for renal tumor evaluation are available, a computed tomography scan is the most sensitive, efficient and comprehensive method.[6] Renal lymphoma on CT scans, is typically homogenous, minimal contrast enhanced and rarely vascular invasive. For most of time, it is unilaterally involved and parenchymally located, which is different from renal cell carcinoma, transitional cell carcinoma, and metastatic renal tumors.[6,7] An image study of our patient exhibited these typical characters on a CT scan, and lymphoma was highly suspected prior to pathologic proof. For solitary renal tumors, surgical resection is indicated for definite treatment and accurate diagnosis. However, in the elderly or those at high risk for operations, percutaneous biopsy is also an alternative to confirm the diagnosis.[8]

The diagnosis of MALT lymphoma is usually made based on morphology, immunophenotype and genetic analysis. The morphology of MALT lymphoma is characterized with monocytoid B cells, small lymphocytes with abundant cytoplasm and lymphoepithelial lesions. Immunophenotype was positive for CD19, CD20, and CD22 and negative for CD5, CD10, and CD23.[1] The renal biopsy sample of our patient fulfilled the diagnosis of MALT lymphoma. Systemic involvement of her MALT lymphoma was documented later, with a positive result in a bone marrow biopsy. Garcia et al reviewed and summarized a total of 20 cases of MALT lymphoma involving the kidney. Half of the collected patients had simultaneous bone marrow or other organ involvement.[2] The treatment strategy included either nephrectomy or chemotherapy, and some patients underwent both treatments. Another report showed a good result in a rather short follow-up period, with all patients receiving nephrectomy.[3] There is currently no conclusive policy for the treatment of renal MALT lymphoma. Although the renal mass was rather large, our patient responded to chemotherapy well as evidenced by a continuous decrease in mass size. In our patient, renal function impairment had been noted for years. With the renal involvement of MALT lymphoma, renal function did not deteriorate. Contrast medium exposure during the image study did not cause kidney injury under prophylactic treatment and preparation. Previous studies did not provide information regarding the long term renal outcome of patients with and without nephrectomy.[2,3] Although data of patient survival is limited, preservation of renal function should be an important concern in these patients.

It has been estimated that approximately 15% patients with WM developed hyperviscosity syndrome and caused significant morbidity and mortality.[2] A literature review found that only 10 patients with MALT lymphoma had associated abnormal paraproteinemia, such as WM, and none of them was of renal origin.[4,9,10] Three of these patients had complications of hyperviscosity syndrome, and plasmapheresis or plasma exchange was thus performed. The prognosis of these patients was not favorable; disease relapse was frequent, despite the combination of systemic chemotherapy and tumor resection. There was no study to investigate the reason why MALT lymphoma had a higher incidence of hyperviscosity syndrome than other lymphomas. In an uncertain period of time from disease onset to diagnosis, our patient did not have any clinical features related to hyperviscosity syndrome. Although a high level of paraproteinemia was not directly correlated with the development of hyperviscosity syndrome, serum IgM levels of our patient were not as high as in previous reports.[4,9,10] The clinical implication and prognostic value of the development of WM in association with hyperviscosity syndrome in renal MALT lymphoma, however, is not yet clear. More patient collection and clinical studies are required to clarify this issue.

In conclusion, for the first time, we report a patient of renal MALT lymphoma presenting with asymptomatic solitary renal mass. In addition to bone marrow involvement, WM was detected but hyperviscosity syndrome was not evident. The renal lymphoma demonstrated a regressive change after systemic chemotherapy and thereby nephrectomy was not indicated.

References

1. Good DJ, Gascoyne RD. Classification of non Hodgkin's lymphoma. *Hematol Oncol Clin N Am* 2008;**22**:781—805.
2. Garcia M, Konoplev S, Morosan C. MALT Lymphoma involving the kidney. A report of 10 cases and review of the literature. *Am J Clin Pathol* 2007;**128**:464—73.
3. Kato Y, Hasegawa M, Numasato S. Primary mucosa-associated lymphoid tissue-type lymphoma arising in the kidney. *Int J Urol* 2008;**15**:90—2.
4. Valdez R, Finn WG, Ross CW. Waldenström macroglobulinemia caused by extranodal marginal zone B-cell lymphoma. A report of six cases. *Am J Clin Pathol* 2001;**116**:683—90.
5. Kovacs G, Akhtar M, Beckwith BJ. The Heidelberg classification of renal cell tumours. *J Pathol* 1997;**183**:131—3.
6. Urban BA, Fishman EK. Renal lymphoma: CT patterns with emphasis on helical CT. *Radiographics* 2000;**20**:197—212.
7. Zhang J, Lefkowitz RA, Bach A. Image of kidney cancer. *Radiol Clin North Am* 2007;**45**:119—47.
8. Volpe A, Kachura JR, Geddie WR. Techniques, safety and accuracy of sampling of renal tumors by fine needle aspiration and core biopsy. *J Urol* 2007;**178**:379—86.
9. Yamasaki S, Matsushita H, Tanimura S. B-cell lymphoma of mucosa-associated lymphoid tissue of the thymus: a report of two cases with a background of Sjögren's syndrome and monoclonal gammopathy. *Hum Pathol* 1998;**29**:1021—4.
10. Allez M, Mariette X, Linares G. Low-grade MALT lymphoma mimicking Waldenström's macroglobulinemia. *Leukemia* 1999; **13**:484—5.