

# Renal mucosa-associated lymphoid tissue lymphoma: a case report and literature review

## Linfoma de tejido linfoide asociado a mucosa renal: reporte de un caso y revisión de la literatura

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### Abstract

**Background:** Renal MALT lymphoma is a rare entity with few reports in the literature. These neoplasms arise at extranodal sites, usually related with chronic inflammation due to infection or autoimmune disorder and share histologic and immunophenotypic features. To date, 10 cases of renal MALT lymphoma have been described.

**Case presentation:** We present a case of a MALT renal lymphoma in a 69-year-old woman with suspected preoperative diagnosis of renal cell carcinoma (papillary vs chromophobe type), RENAL SCORE 9p. We performed a retroperitoneal laparoscopic left partial nephrectomy guided by intraoperative ultrasound.

**Conclusion:** There are few reports in the literature regarding renal MALT lymphoma. We consider that this might be considered as a differential diagnosis of renal mass.

Keywords:

Renal lymphoma, mucosa-associated lymphoid tissue (MALT), nephrectomy

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#### Resumen

**Antecedentes:** El linfoma MALT renal es una entidad rara con pocos reportes en la literatura. Estas neoplasias surgen en sitios extraganglionares, generalmente relacionadas con inflamación crónica debida a infección o trastorno autoinmunitario, y comparten características histológicas e inmunofenotípicas. Hasta la fecha se han descrito 10 casos de linfoma MALT renal.

**Presentación del caso:** Presentamos un caso de linfoma renal MALT en una mujer de 69 años con sospecha de diagnóstico preoperatorio de carcinoma de células renales (tipo papilar vs cromófobo), RENAL SCORE 9p. Se realizó nefrectomía parcial izquierda laparoscópica retroperitoneal guiada por ecografía intraoperatoria.

#### Palabras clave:

Linfoma renal, tejido linfoide asociado a mucosas (MALT), nefrectomía

**Conclusión:** Existen pocos informes en la literatura sobre el linfoma MALT renal. Consideramos que esto podría plantearse como diagnóstico diferencial de masa renal.

#### Background

Mucosa-associated lymphoid tissue low-grade B-cell lymphoma (MALT), first described by Isaacson and Wright in 1983,<sup>(1)</sup> is recognized as a distinct subtype in the non-Hodgkin lymphoma classification and is a different group from B-cell lymphomas specified in the World Health Organization (WHO) classification.<sup>(1,2)</sup> Renal involvement represents less than 1% of prevalence of MALT lymphomas.<sup>(3-6)</sup>

Pelstring *et al.* reported the first case of renal MALT lymphoma in 1991 and Colovic first described primary renal MALT lymphoma in 1999.<sup>(7)</sup> They are one of the less aggressive lymphomas and often present as an indolent, localized disease and the treatment of these cases was heterogeneous, from surgical, chemotherapy, immunotherapy, to radiotherapy.<sup>(7,8)</sup>

Clinically, patients with renal lymphoma are usually asymptomatic or may refer nonspe-

cific symptoms such as low back pain and/or haematuria. Diverse radiographic presentations have been reported, including single or multiple retroperitoneal masses and, in some cases, hydronephrosis or diffuse nephromegaly.<sup>(9)</sup>

#### **Case Presentation**

69-year-old woman with a history of IA endometrial cancer treated with adnexa-hysterectomy in March 2021. In a follow up CT scan a completely endophytic hypo-vascular tumor on the left kidney was evidenced, poorly delimited at the level of the upper polar line, approximately 30 mm in the axial plane, suggesting a papillary or chromophobe tumor. RENAL SCORE: 1+3+3+p+2=9p (Figure 1). Figure 1. Retropyelic branch entering the tumor. Cross-section



A retroperitoneal laparoscopic left partial nephrectomy with intraoperative ultrasound guidance was performed.

Pathological report showed microscopically alteration of the histoarchitecture, whith lymphoid proliferation that infiltrates adipose tissue and whose intraparenchymal borders are delimited from the preserved renal parenchyma. This lymphoid proliferation replaces some glomerular and ductal structures, leaving others preserved. Morphologically, no clear epithelial infiltration was observed. The cell population was predominantly made up of small lymphoid cells, some plasma cells, and very few and isolated large cells. There were few and isolated lymphoid follicles with reactive-appearing germinal centers.

Immunohistochemistry (IHC) was positive for: (Figure 2: A, B, C, D, E). CD20, CD3, CD5, CD23; cytokeratin's: bcl-2, MIB-1. and negative for: CD10, bcl-6. Labeling CD138, Kappa light chains and Lambda light chains was also observed: indicating of isolated plasma cells remains. The final diagnosis was renal MALT type lymphoma. Figure 2. Infiltration by non-Hodgkin's lymphoma b, extranodal MALT type



A: Infiltrated renal parenchyma. **B**: Preserved renal parenchyma. **C**: oval glomerulus (indicated by arrow). **D**: Black arrow indicates epithelial tubule, red arrow conglomerate lymphocytes. **E**: IHC CD20 positive, arrow points to glomerulus.

The staging with PET CT, bone marrow biopsy and flow cytometry did not show distant disease.

Considered as primary renal MALT type lymphoma stage IE, no further treatment was indicated and the patient remains disease free in the follow up.

#### Discussion

In 2020 the French LyKID study published the largest non-autopsy lymphoma series with renal involvement, out of 87 cases only five were MALT type.<sup>(10)</sup>

MALT lymphomas can affect several anatomical sites and the appearance is usually preceded by an inflammatory process, mainly observed in the gastrointestinal tract, respiratory system, salivary glands, head and neck, thyroid, breast, gallbladder, cervix, and ocular adnexa.<sup>(3-5,7-9,11,12)</sup> Renal mucosa-associated lymphoid tissue lymphoma: a case report and literature... Felipe Córdoba A., et al.

The etiology of renal MALT lymphoma is not well established. However, repetitive injury to renal lymphatics due to a chronic inflammatory process, such as chronic pyelonephritis, usually precedes the onset of the disease. Usually debut as a localized disease with slow clinical progression. However, transformation to high-grade in the late course of the disease was reported in 8% of the patients.<sup>(5,13)</sup>

Radiographic presentations may vary, as well as their clinical presentation.<sup>(14)</sup> There are no differences in terms of imaging diagnosis since it debuts as a renal mass. The role of PET- FDG (18F-fluorodeoxyglucose positron emission tomography computed tomography) in this scenario remains debatable due to the low avidity of MALT lymphoma extranodal lesions, and their tendency to be located in tissues with physiologic [18F]FDG-uptake can make image interpretation difficult.<sup>(15)</sup>

In case of nodal and extranodal involvement, a diagnostic percutaneous biopsy can be considered to carry out systemic treatment. In our case it was not necessary since it was a single presentation in the kidney.

The prognosis for patients with renal MALT lymphoma is good with chemotherapy, surgical intervention, or radiation. The 5-year overall survival for MALT lymphoma in the genitourinary tract is 75.6 %, even though for those with different anatomical origins ranges from 69.1 to 87.9 % .<sup>(10)</sup>

Chemotherapy takes place when there is a multicentric or systemic involvement. In the literature, most patients diagnosed with B-cell lymphoma underwent cytotoxic chemotherapy with rituximab and intratecal methotrexate. Patients with MALT lymphomas have better prognosis compared with those with high grade lymphomas.<sup>(12)</sup> In most cases, the diagnosis is made after nephrectomy, generally due to suspicion of another oncological lineage, and most of these patients do not require additional treatment.

#### Conclusions

This is a rare disease and should be considered as a differential diagnosis in the case of an atypical renal mass. Early diagnosis and timely treatment have positive results.

#### **CRediT taxonomy**

- CAF: Conception, design, supervision, data collection, writing.
- VBRF: Data collection, analysis and interpretation, literature review.
- Camean Juan: Analysis and interpretation, literature review.
- Chemi Joaquin: Analysis and interpretation, literature review
- JJ: Supervision, writing
- FC: Conception, design, critical review
- VGM: Conception, design, critical review

#### Ethics approval and consent to participate

We obtained the ethics approval.

#### **Consent for publication**

We obtained written consent for publication from the patients.

#### Availability of data and material

The information was obtained from patients medical records.

#### **Competing interest**

Authors declare that there is no conflict of interest regarding the publication of this article.

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