Extranodal Richter’s syndrome of the urinary bladder

Almudena Carrión-Valencia*, Jonathan Rodríguez-Talavera, Begoña Ballesta-Martínez

Urology Service, University Hospital Nuestra Señora La Candelaria, Santa Cruz De Tenerife, Spain

*E-mail: almudena088@hotmail.com

INTRODUCTION

Bladder tumors are the second-most frequent urological neoplasms. Nonurothelial bladder tumors are rare (10%–25%).[1] One of these is lymphomas of the urinary bladder (0.2–0.5%). Lymphomas can be classified into primary tumors localized to the bladder or secondary tumors in patients with a history of previous hematological disease. One of the forms is called Richter’s Syndrome, which is the transformation of a low grade chronic lymphocytic leukemia into a high-grade lymphoma. Usually, it happens in peripheral lymph nodes (64%), but it can manifest with extranodal involvement (41%): gastrointestinal, central nervous system, ocular, skin face, bone, and bronchus.[2] The importance of our case is that, despite an extensive review, we have not found any reports of extranodal Richter’s Syndrome in the bladder.

CASE REPORT

An 84-years-old male with a history of hyperlipidemia, former smoker, and chronic lymphatic leukemia (diagnosed by flow cytometry of peripheral blood: low-grade CD 19/5+; CD 20+; CD 23+; classical form) diagnosed in 2009 was treated with chemotherapy (chlorambucil-prednisone) which resulted in full remission. In 2015, the patient was evaluated for loss of weight, appetite, and fever. The hematologist consulted us because of hematuria with coexistent bilateral obstructive uropathy. We performed transurethral resection of the bladder which revealed secondary lymphoma in the bladder, probably the result of a high-grade transformation from chronic leukemia, a very rare location of this transformation.
was taken up for evaluation of hematuria and the findings were a ten centimeters solid hypovascularized bladder tumor on the floor of the bladder, the ureteral meatuses were not found. Transurethral resection of bladder (TURB) tumor was performed in the same sitting, removing all the exophytic part of the tumor. Blood tests revealed elevation of creatinine after surgery, so bilateral urinary diversion with 8 F nephrostomy tubes was done which resulted in the improvement of renal function.

The biopsy showed extensive muscle infiltration of high Grade B lymphoproliferative process, a diffuse large B-cell lymphoma (DLBCL) with immunohistochemical marking CD 20 + and Ki 67 + at least in the 80% of the cell nuclei, a proliferative index which indicates the aggressiveness of the illness [Figure 2]. Given this result, the patient was referred to hematology, who began treatment with R-COP with the improvement of the general status.

New CT-scan confirmed the absence of abdominal and retroperitoneal lymph nodes [Figure 3], but persistence and progression of the bladder tumor, that was infiltrating the prostate, the right seminal vesicle, and the right lateral edge of the rectum.

It was decided to start second-line chemotherapy with rituximab, gemcitabine, and oxaliplatin. The bladder tumor was refractory to treatment and a review of the biopsy confirmed the diagnosis. The patient’s condition deteriorated due to progression of the disease, despite second-line treatment, and it was decided to start palliative treatment.

**DISCUSSION**

Bladder cancer is the seventh-most common tumor and the second-most frequent urological neoplasm. The majority (90%) have an urothelial origin. Nonurothelial bladder tumors are rare. They can be classified as nonurothelial epithelial (squamous carcinoma and adenocarcinoma) and nonurothelial nonepithelial (melanoma, sarcomas, carcinosarcomas, and lymphomas), the latter are uncommon.[1]

Lymphoma of the urinary bladder is an uncommon lesion. It can be either primary or secondary and the most frequent histological subtype is DLBCL. It occurs in approximately 5%, with a transformation index about 0.5%–1%. Even after an extensive literature search, the authors could not find any reports of extranodal Richter’s syndrome in the bladder.

The most frequent symptom of lymphoma of the urinary bladder is hematuria. Other symptoms include dysuria, frequency, and abdominal or lumbar pain. The cystoscopy findings in 70% of the cases are that of a single solid bladder mass, in 20% there are multiple masses, and in 10% a diffuse thickening of the bladder wall. The most common location is the bladder dome.[3] Pathological diagnosis, with immunophenotypic analysis and an extension study, is essential to specialized medical treatment.

The prognosis of the nonurothelial variants of the bladder cancer is worse than that of urothelial malignancies.[1] In the case of secondary lymphomas, it depends on the extent...
of the illness, health status and age of patient, tumoral mass, and clinical manifestations (fever and weight loss). The prognosis of Richter’s syndrome is generally unfavorable.\textsuperscript{4} The most important prognostic factor is the clonal relationship between the chronic lymphocytic leukemia and the aggressive lymphoma clones.\textsuperscript{5}

Chemotherapy and monoclonal antibodies are the treatment of choice, which is determined by the histologic type and the disease extension. The most frequent treatment schedule is based on cyclophosphamide, doxorubicin, vincristine, prednisone, and rituximab.\textsuperscript{5}

**CONCLUSION**

Lymphoma of the urinary bladder can be a challenge for the urologist, due to the low incidence and an unusual clinical presentation. Richter’s syndrome should be taken into consideration in a patient with a history of lymphoproliferative syndrome. TURB is indicated for histopathological diagnosis and for confirming the transformation to DLBCL as also alleviating hematuria and obstructive uropathy.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

**REFERENCES**

1. Chalasani V, Chin JL, Izawa JL. Histologic variants of urothelial bladder cancer and nonurothelial histology in bladder cancer. Can Urol Assoc J 2009;3:S193-8.
2. Omoti CE, Omoti AE. Richter syndrome: A review of clinical, ocular, neurological and other manifestations. Br J Haematol 2008;142:709-16.
3. Venyo AK. Lymphoma of the urinary bladder. Adv Urol 2014;2014:1-19.
4. Rossi D, Gaidano G. Richter syndrome: Pathogenesis and management. Semin Oncol 2016;43:311-19.
5. Parikh SA, Kay NE, Shanafelt TD. How we treat richter syndrome. Blood 2014;123:1647-57.

How to cite this article: Carrión-Valencia A, Rodríguez-Talavera J, Ballesta-Martinez B. Extranodal Richter’s syndrome of the urinary bladder. Indian J Urol 2018;34:297-9.