CLINICAL CASE

Primary ureteral lymphoma


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Abstract

Background: Lymphomas affecting the urinary tract are a relatively rare pathology.

Clinical case: We present herein the case of a 54-year-old man diagnosed with a lesion of the right renal pelvis and proximal ureter associated with 2 hepatic lesions. Biopsy of said lesions revealed B cell (CD20+) non-Hodgkin’s lymphoma. Treatment was begun with rituximab resulting in good clinical progression.

Discussion: A total of 2.5% of lymphomas affect the urinary tract. The treatment of choice is systemic chemotherapy for 30 weeks with intermittent cycles of bleomycin, cyclophosphamide, vincristine, and dexamethasone. Rituximab may be used in cases of CD20+ lymphomas. Urinary system lymphomas have a 5-year survival rate of 50%.

Conclusions: Urinary tract lymphomas are rare and their treatment is based on chemotherapy.

Keywords: non-Hodgkin’s lymphoma; B cell lymphoma; Ureteral

Linfoma ureteral primario

Resumen

Introducción: Los linfomas que afectan al aparato urinario constituyen una enfermedad relativamente rara.

Caso clínico: Presentamos el caso de un varón de 54 años que es diagnosticado de una lesión en la pelvis renal y un uréter proximal derecho, asociado a 2 lesiones hepáticas. Se realiza biopsia de dichas lesiones objetivándose un linfoma no Hodgkin de células B (CD20+). Se inició tratamiento con rituximab con muy buena evolución clínica.

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Discusión: La afectación de la vía urinaria por linfomas representa el 2,5% de los linfomas. El tratamiento de elección es la quimioterapia sistémica durante 30 semanas con ciclos intermitentes de bleomicina, ciclofosfamida, vincristina y dexametasona. En el caso de los linfomas CD20+ puede utilizarse rituximab. El pronóstico de los linfomas del sistema urinario es del 50% a los 5 años.

Conclusiones: Los linfomas del aparato urinario son raras entidades, cuyos tratamientos son a base de quimioterapia.

Introduction

Malignant lymphomas are a relatively frequent pathology (the seventh most common cause of death by cancer in the U.S.) and favored today by the immunodeficiency that is characteristic of transplants and AIDS. Primary lymphomas that affect the urinary tract are a relatively rare pathology.

Ten percent of the cases with urinary tract involvement have clinical manifestations and there is a preference for the adult patient.\(^1\)

Case presentation

A 54-year-old man was an outpatient referral from the digestive service where he was being evaluated for liver disease from the hepatitis C virus due to a polypoid lesion in the right renal pelvis identified in a control abdominal CAT scan.

The CAT scan revealed grade II/IV right pyeloureteral ectasia produced by a polypoid lesion in the renal pelvis that extended to the proximal ureter, as well as hepatic lesions (fig. 1-3). Urinary cytology was negative for neoplastic cells and cystoscopy did not show the presence of concomitant bladder lesion.

Liver puncture biopsy was performed that resulted in the anatomic pathology of CD20+ B cell non-Hodgkin’s lymphoma (figs. 4 and 5), which was also demonstrated in the biopsy carried out \textit{a posteriori} at the kidney level. Afterwards, therapy with rituximab was begun.

After one year of treatment the patient has good treatment response, presenting with minimal remnants of the disease at the level of the proximal ureter (fig. 6).
Primary ureteral lymphoma

Discussion

Lymphomas make up a group of diseases arising from the lymphoid organs. Fifty percent of necropsies show urinary tract involvement, but its clinical involvement is demonstrated in only 10% of the cases, being located at the renal, vesical, ureteral, prostatic, testicular, and penile levels. Of the lymphomas affecting the urinary system, 2.5% affect the urinary tract.

Lymphomas usually affect the urinary tract in middle-to-advanced age patients and the majority of case series show a predominance in men. A greater incidence of lymphoid neoplasias have been described in the last few years in patients with AIDS and in transplantation patients, as a consequence of their immunodeficiency.

The clinical presentation of lymphomas affecting the urinary tract tends to be nonspecific, with a constitutional syndrome, weight loss, flank pain, hematuria, and kidney failure.

It can be asymptomatic and diagnosed incidentally. Presentation can be unilateral or bilateral.

Radiologic images may be indistinguishable from other types of neoplasias, and therefore when this pathology is suspected, diagnostic biopsy should be performed.

Staging can be considered complete once clinical examination, blood analysis, chest x-rays, lung and abdominopelvic CAT scans, and bone marrow biopsy are carried out. A lymphoma can be considered primary only when the results from all these techniques are negative.

Histologic study and immunohistochemical techniques are tests that confirm non-Hodgkin’s lymphoma.

Figure 4  Hepatic lesion biopsy showing extensive lymphocytic infiltrate marked with CD20.

Figure 5  Core biopsy of the renal pelvic lesion showing extensive lymphocytic infiltrate marked with CD20.

Figure 6  Abdominal CAT scan reconstruction image after completing treatment.
Surgery can never be regarded as curative for these tumors and its role should be considered diagnostic for the purpose of establishing treatment.

The treatment of choice is systemic chemotherapy. An intensive treatment has been designed that entails 30 weeks of intermittent cycles of bleomycin, cyclophosphamide, vincristine, and dexamethasone with high doses of methotrexate. There has been complete remission in 78% of the cases that has been maintained for 2 years in 80% of the patients treated. Even though the customay alternative chemotherapy treatment appears to be the CHOP pattern (up to 9 cycles with cyclophosphamide, doxorubicin, vincristine, and prednisolone), there are other alternative patterns such as VIM-Bleo that combines ifosfamide, methotrexate, etoposide, and bleomycin.¹ There are new therapeutic modalities for lymphomas that affect the genitourinary system that are based on immunophenotypic analysis; for example rituximab has shown excellent results in CD20+ lymphomas.² Rituximab is a monoclonal antibody that targets the CD20 receptors of both benign and malignant B cells. Even though its metabolism has not yet been established, it can be used in patients with kidney failure.³

The prognosis of primary lymphomas of the urinary system is better that that of secondary lymphomas, because the former are organ-confined. There is a 50% 5-year survival rate.¹

Conclusions

Ureteral lymphoma is a rare entity that can be indistinguishable from other urothelial tumors. Prognosis of the primary forms is relatively good, compared with other disseminated forms. Treatment is based on chemotherapy.

Ethical responsibilities

Protection of persons and animals. The authors declare that the procedures followed conformed to the ethical standards of the responsible committee on human experimentation and were in accordance with the World Medical Association and the Declaration of Helsinki.

Data confidentiality. The authors declare that they have followed the protocols of their work center in relation to the publication of patient data.

Right to privacy and informed consent. The authors have obtained the informed consent of the patients and/or subjects referred to in the article. This document is in the possession of the corresponding author.

Financial disclosure

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Conflict of interest

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