

# Ureteral spread of a primary cutaneous diffuse large B-cell lymphoma, leg type

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## ABSTRACT

We report a case of 76-year-old man, with a past medical history of primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT), who presented with ureteral tumor diagnosed as urothelial carcinoma on imaging investigations. Histological examination showed an unusual finding. The tumor was a ureteral localization of the PCDLBCL-LT previously diagnosed. To the best of our knowledge, this is the first case of PCDLBCL-LT with ureteral spread described in the literature.

**Key words:** Lymphoma, metastasis, ureter

## INTRODUCTION

Metastasis to the ureter may evolve by direct or indirect extension and invasion from the most common malignant tumors, such as breast, colon and lymphoma. Lymphoma involvement of the ureter is usually caused by contiguous spread from retroperitoneal lymph nodes and may cause stenosis or rupture of the ureter. Primary cutaneous B-cell lymphomas (PCBCL) have rare extracutaneous manifestations, especially cutaneous follicle-centre lymphomas or cutaneous marginal zone B-cell lymphomas. In contrast, cutaneous diffuse large B-cell lymphomas are characterized by an aggressive clinical course and frequent visceral involvement. We report a case of a primary cutaneous lymphoma that spread to the ureter.

## CASE REPORT

A 76-year-old male patient presented with 2 months history of progressing, non-specific, right

flank pain without any other accompanying symptoms. He had no history of urolithiasis, but he had presented 1 year earlier with a mass of the right knee measuring 5 cm in its greatest diameter. Histological examination showed lymphoid diffuse large cells proliferation, which stained diffusely with anti CD20, anti Bcl2 but was CD10 negative in the immunohistochemical study. A diagnosis of primary cutaneous diffuse large B-cell lymphoma, leg type (PCDLBCL-LT) was then made. No others lymphomas were found at the time of diagnosis. The patient had no others skin conditions, especially no history of infections.

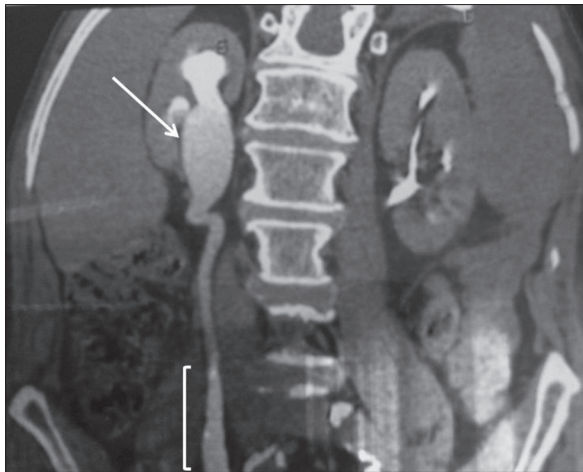
For the current symptoms, he underwent ultrasonography of the abdomen which revealed right hydronephrosis. Abdomino-pelvic computed tomography scan showed pyelo-calyceal dilatation with continuous obstruction, 8 cm in length, of the distal portion of the right ureter [Figure 1]. No retroperitoneal or mesenteric lymphadenopathy was found. The diagnosis of ureteral tumor of urothelial origin was then suspected and the patient underwent a right radical nephroureterectomy.

Macroscopic examination showed a kidney measuring 10 × 6 × 3 with a ureter measuring 12 cm of length. On sectioning the ureter, a white, firm, concentric thickness of the pelvic portion of the right ureteral wall was found. The abdominal portion of the ureter as well as the pelvi-calyceal system was dilated.

Histological examination showed infiltration of the ureteral wall by a lymphoid diffuse large cells proliferation that spared the urothelium [Figure 2a]. The tumoral cells were medium to large size centrocytes with a variable proportion of centroblasts and immunoblasts mixed with small reactive lymphocytes [Figure 2a]. On immunohistochemical study,

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**Figure 1:** Abdomino-pelvic computed tomography scan: A pyelo-calyceal dilatation (arrow) with continuous obstruction of the distal portion of the right ureter

the large cells stained diffusely and intensely with anti CD20, anti Bcl2 and were CD10 and Bcl6 negatives [Figure 2b-d].

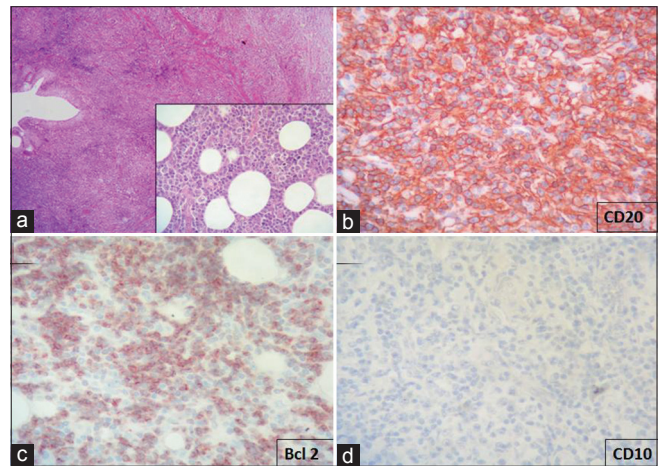
The diagnosis of ureteral spread of the cutaneous diffuse large B-cell lymphoma, leg type was then made.

## DISCUSSION

Metastases from a distant primary neoplasm to the ureter are encountered infrequently. The first case of distant ureteral metastasis was reported in 1909.<sup>[1]</sup> In the majority of cases, the presence of ureteral metastases is neither diagnosed nor suspected prior to death. Babaian *et al.* reported that only 0.3% of 11,689 patients with malignant disease had histologically proven metastatic lesions to the ureter in a large series of autopsies.<sup>[2]</sup> The primary sites include stomach, colon, uterine cervix, breast, skin, prostate, lung and adrenal gland.<sup>[2]</sup> Ureteral involvement by lymphoma is extremely rare and is usually caused by contiguous spread from retroperitoneal lymph nodes. A review of the literature showed that no cases of PCBCL involving the ureter have been reported.

PCBCL represent only 20-25% of all primary cutaneous lymphomas and are less frequent than cutaneous T-cell lymphomas.<sup>[3]</sup> They belong to a distinct group of B-cell lymphoproliferative disorders defined by their presentation in the skin, without evidence of extracutaneous spread at the time of diagnosis. The new 2008 World Health Organization-European Organization for Research and Treatment of Cancer classification for cutaneous lymphomas identifies 4 main subtypes of PCBCLs: Primary cutaneous marginal zone lymphoma, primary cutaneous follicular center lymphoma, PCDLBCL-LT, and primary cutaneous large B-cell lymphoma (PCLBCL), other.<sup>[3]</sup>

Normally, there are no extracutaneous manifestations for either primary cutaneous follicle-centre lymphomas



**Figure 2:** (a) Tumoral cells: Medium to large size centrocytes with a variable proportion of centroblasts and immunoblasts mixed with small reactive lymphocytes (b-d) Immunohistochemical study: Large cells stained diffusely and intensely with anti CD20, anti Bcl2 and were CD10 and Bcl6 negatives

or primary cutaneous marginal zone B-cell lymphomas. Consequently, mortality rates tend to be low.<sup>[4]</sup> In contrast, cutaneous diffuse large B-cell lymphomas are characterized by an aggressive clinical course and a higher mortality rate. In fact, disease-related 5-years survivals for PCDLBCL, LT range from 36% to 55% compared with over 95% for the other subtypes of primary cutaneous lymphomas<sup>[5]</sup> and meanly 40% of patients with PCBCL, LT develop extracutaneous disease, with a mean time to extracutaneous spread of 20 months.<sup>[6]</sup> The central nervous system (CNS) is the most common site of visceral dissemination with 27% of extracutaneous metastases occurring in the CNS. To the best of our knowledge, this is the first patient to be reported in the literature with PCDLBCL, LT involvement of the ureter.

Ureteral metastasis commonly causes no specific symptoms. Back pain occurs in approximately half of patients and a third of them will have hematuria. Early symptoms of ureteral metastases may be masked by the overbearing presence of the primary tumor or other metastases, or by the generally poor condition of the patient. In the present case, ureteral involvement by the lymphoma caused only a non-specific flank pain.

Radiographic findings are also variable. Extrinsic ureteral obstruction due to metastatic diseases depends on the pattern of the tumoral spread: Hematogenous submucosal/mucosal metastasis, hematogenous adventitia metastases spreading along periureteral vessels, scirrhous metastatic spread along periureteral vessels, or metastatic spread into lymph nodes with perinodal desmoplastic reaction.<sup>[7]</sup> Ureteral involvement by lymphoma presented in most reported cases as an obstruction and thickened wall.<sup>[8]</sup> If concentric thickness of the ureteral wall is found, physicians should be aware of the possibility of malignant lymphoma, especially if the patient had been diagnosed previously with malignant

lymphoma and should consider laparoscopic biopsy for diagnosis to avoid ineffective treatment as was the case of our patient. In fact, chemotherapy and radiotherapy are required for the treatment of PCLBCL. The prognosis is poor especially in cases of multiple skin lesions or extracutaneous manifestations.

To the best of our knowledge, this is the first case of PCBCL with ureteral spread described in the literature. PCBCL involvement of the ureter-or any other site of involvement-must be considered as a possibility in any patient with a history of cutaneous lymphoma, particularly when the lymphoma is morphologically of a large cell type, generates a large tumor burden in the skin and displays a non-germinal center immunophenotype (CD10 and BCL6 negative). In this particular case, evaluation of the ureteral mass including routine cytology and biopsy would have provided information essential to the pre-treatment management of the patient.

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