

# Plasmablastic lymphoma presenting as a ureteral polypoid mass

Sohei Yamamoto\*, Yuma Yasuda, Yu Sakai

Department of Diagnostic Pathology, Anjo Kosei Hospital, Anjo, Japan

\*E-mail: soheiyamamoto@kosei.anjo.aichi.jp

## ABSTRACT

Plasmablastic lymphoma (PBL) is a rare and aggressive subtype of B-cell lymphoma, which occurs typically in the oral cavity of human immunodeficiency virus (HIV)-positive patients. We report a case of a 44-year-old HIV-positive patient with a solitary polypoid mass of the left ureteropelvic junction, causing unilateral hydronephrosis and clinically mimicking urothelial carcinoma. A laparoscopic nephroureterectomy was performed, and pathological examinations revealed the mass as PBL. PBL can present in various forms, even as a polypoid mass of the upper urinary tract, and it should be considered in the differential diagnosis of any mass detected in the HIV-positive patients.

## INTRODUCTION

Plasmablastic lymphoma (PBL) is a rare and aggressive B-cell lymphoma characterized by diffuse proliferation of neoplastic cells resembling B-immunoblasts or plasmablasts, with an immunophenotype of plasma cells.<sup>[1,2]</sup> Although first described as occurring in the oral cavity in the setting of human immunodeficiency virus (HIV) infection, PBL has since been identified in patients with other causes of immunodeficiency, in immunocompetent patients, and in a variety of anatomic locations.<sup>[1,2]</sup> Here, we present a case of PBL presenting as a polypoid mass of the ureteropelvic junction, causing unilateral hydronephrosis and clinically mimicking usual urothelial carcinoma.

## CASE REPORT

A 44-year-old HIV-positive male, on antiretroviral treatment, was referred to our hospital with asymptomatic unilateral hydronephrosis found on routine health examination. General physical examination findings were within normal limits. The last known CD4 count was 486 cells/ $\mu$ L, and all

other blood tests were normal or as expected. Urinalysis was remarkable for microscopic hematuria. Enhanced computed tomography showed a well-defined mass in the left ureteropelvic junction, and retrograde pyelography revealed a filling defect at the same site [Figure 1]. A left laparoscopic nephroureterectomy was performed due to the suspicion of urothelial carcinoma.

Gross examination revealed a hemorrhagic polypoid mass, 26 mm in size, in the left ureteropelvic junction [Figure 2a]. Histologically, the mass and subjacent structures were diffusely infiltrated by large lymphoid cells with abundant basophilic cytoplasm, eccentrically located enlarged nuclei with prominent nucleoli [Figure 2b and c]. Frequent mitotic figures (approximately 70 mitoses per 10 high-power fields), apoptotic bodies, and intermixed tingible body macrophages (a “starry-sky” appearance) were also documented. Although peripelvic adipose tissue was focally involved by neoplastic cells’ infiltration, all surgical margins were negative for malignant cells. On immunohistochemistry, the neoplastic cells were positive for CD138 [Figure 2d] and c-myc [Figure 2e]; only partly positive for CD79a, CD45 (leukocyte common antigen), and epithelial membrane antigen; and negative for

### Access this article online

#### Quick Response Code:



#### Website:

www.indianjurol.com

#### DOI:

10.4103/iju.IJU\_364\_17

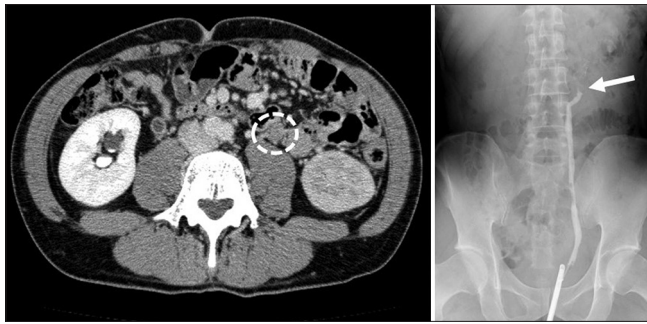
This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

**For reprints contact:** reprints@medknow.com

**Received:** 10.12.2017, **Accepted:** 27.01.2018

**Financial support and sponsorship:** Nil.

**Conflicts of interest:** There are no conflicts of interest.



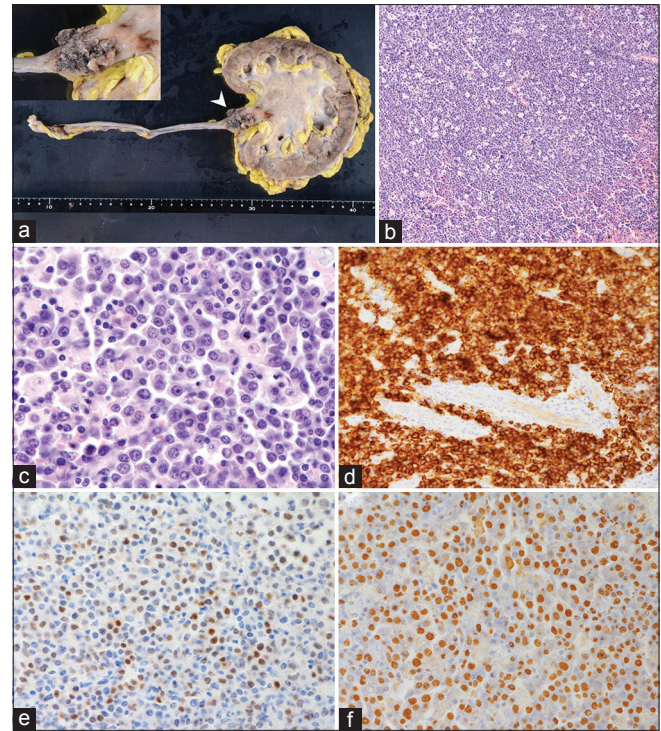
**Figure 1:** Left: Contrast-enhanced computed tomography of the abdomen shows a slightly enhanced tumor in the left ureteropelvic junction (marked by a broken line circle). Right: Retrograde pyelography shows filling defect (indicated by an arrow)

CD20, CD3, CD30, ALK-1, Bcl-2, Bcl-6, CD56, CD34, CD117, and pan-cytokeratin. Ki-67 labeling index was >90%. *In situ* hybridization for Epstein-Barr virus (EBV)-encoded RNA (EBER) was strongly positive in the neoplastic cells [Figure 2f]. After a staging workup including radiographic skeletal survey, positron emission tomography/computed tomography, and bone marrow biopsy, there was no evidence of other organ involvement, and serum and urine protein electrophoresis were normal. Therefore, the final diagnosis of PBL originating from the left ureteropelvic junction was made. The patient recovered uneventfully, and for 18 months following surgery, he was free of disease without receiving adjuvant therapy.

## DISCUSSION

Although PBL is characterized for its predilection of involving the oral cavity, a number of cases have been reported in extraoral sites such as the gastrointestinal tract, lymph nodes, and skin.<sup>[2]</sup> However, PBL originating from the urinary tract is extremely rare, and to date, there have been only two reported cases of ureteral PBL: one with a multifocal bladder tumor and another with a paraurethral mass.<sup>[3]</sup> To our knowledge, this is the first report of a case with PBL presented as a polypoid lesion of the ureteropelvic junction, clinically mimicking as usual type urothelial carcinoma, and resected by laparoscopic nephroureterectomy.

Distinction of PBL from plasma cell neoplasm with plasmablastic morphology may be challenging, and this is especially the case with anaplastic solitary extramedullary plasmacytoma.<sup>[4]</sup> Both PBL and plasma cell neoplasm with plasmablastic morphology may exhibit an identical immunophenotype, i.e., lacking B-cell specific markers and express plasma cell markers.<sup>[4]</sup> Features that favor PBL include association with HIV infection and EBER positivity in neoplastic cells. The presence of cells with morphologically obvious plasmacytic differentiation is not typical of PBL, but it is the defining feature of plasma cell neoplasm.<sup>[4]</sup> In the present case, plasmacytoid urothelial carcinoma (PUC) was also considered as a differential



**Figure 2:** (a) A dark reddish polypoid mass, 26 mm in diameter, is seen in the ureteropelvic junction (arrowhead, inset). (b) Diffuse infiltrate of large atypical lymphoid cells (H and E, stain, ×100). (c) The neoplastic cells have plasmablastic appearance (H and E, stain, ×600). The neoplastic cells are positive for (d) CD138 (immunoperoxidase stain, ×200), (e) c-myc (×400), and (f) Epstein-Barr virus-encoded RNA (*in situ* hybridization, ×400)

diagnosis, but immunohistochemical negativity for pan-cytokeratin ruled out a diagnosis of PUC.

Although the pathogenesis of PBL is poorly understood, recent studies have identified the presence of *MYC* gene rearrangements in addition to the association with EBV infection as important pathogenic mechanisms. As shown in the present case, EBER can be detected using an *in situ* hybridization technique in the PBL cells in 80% of HIV-positive and 46% of HIV-negative patients.<sup>[3]</sup> Rearrangement of the *MYC* gene at 8q24 can be detected in approximately half of PBL cases, with the immunoglobulin genes acting as the most frequent translocation partners.<sup>[5]</sup> In addition to *MYC* rearrangements, gains of *MYC* are also occasionally seen in PBL.<sup>[5]</sup> In the present case, the high nuclear expression of c-myc protein by immunohistochemistry (>50% of neoplastic cells) suggested its oncogenic potential.

PBL is usually associated with poor prognosis and has low sensitivity to chemotherapy.<sup>[2]</sup> Due to the advanced stage at diagnosis in most patients, curative surgical resection is rarely considered in the management of PBL. Fortunately, in this case, the patient has been free from relapse for 18 months without adjuvant therapies after nephroureterectomy, suggesting that an adequate resection with a safe margin could be a treatment option for early-stage PBL.

## CONCLUSION

PBL can present in various forms, even as a polypoid mass of the upper urinary tract, and it should be considered in the differential diagnosis of any mass detected in HIV-positive patients.

### *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. Delecluse HJ, Anagnostopoulos I, Dallenbach F, Hummel M, Marafioti T, Schneider U, *et al.* Plasmablastic lymphomas of the oral cavity: A new

entity associated with the human immunodeficiency virus infection. *Blood* 1997;89:1413-20.

2. Castillo JJ, Bibas M, Miranda RN. The biology and treatment of plasmablastic lymphoma. *Blood* 2015;125:2323-30.
3. Tille JC, Pelte MF, Schwartz J, Dietrich PY, McKee TA. Plasmablastic lymphoma clinically presenting in the urinary tract. *Ann Diagn Pathol* 2012;16:219-23.
4. Vega F, Chang CC, Medeiros LJ, Udden MM, Cho-Vega JH, Lau CC, *et al.* Plasmablastic lymphomas and plasmablastic plasma cell myelomas have nearly identical immunophenotypic profiles. *Mod Pathol* 2005;18:806-15.
5. Valera A, Balagué O, Colomo L, Martínez A, Delabie J, Taddesse-Heath L, *et al.* IG/MYC rearrangements are the main cytogenetic alteration in plasmablastic lymphomas. *Am J Surg Pathol* 2010;34:1686-94.

How to cite this article: Yamamoto S, Yasuda Y, Sakai Y. Plasmablastic lymphoma presenting as a ureteral polypoid mass. *Indian J Urol* 2018;34:152-4.