

Rare Plasmacytoid Urothelial Carcinoma of the Bladder: Two Case Reports

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Plasmacytoid urothelial carcinoma is a rare and aggressive form of urothelial carcinoma characterized with delayed presentation and poor prognosis. Very few cases of this carcinoma have been reported in the literature. Here, we report and discuss two cases of bladder plasmacytoid urothelial carcinoma of a 57-year-old male presented with renal colic, and a 33-year-old female presented with macroscopic hematuria. Pathologic examinations of the transurethral biopsies revealed urothelial carcinoma with plasmacytoid appearance. Subsequently, immunohistochemical evaluation showed positive expression of epithelial markers and CD138. Additionally, losing of the membranous expression of E-cadherin verified the diagnosis of plasmacytoid urothelial carcinoma.

Keywords: plasmacytoid; urothelial carcinoma; CD138; E-cadherin.

INTRODUCTION

Plasmacytoid urothelial cancer (PUC) is an aggressive and quite uncommon form of urothelial carcinoma (UC).⁽¹⁾ It is characterized with late presentation and poor prognosis.⁽²⁾ Microscopically, PUC reveals a plasmacytoid morphology appearing with eosinophilic cytoplasm and eccentric nuclei. It also shows a discohesive growth and often extends in the perivesical adipose tissue.⁽³⁾ Additionally it may morphologically resemble plasmacytoma, lymphoma, or carcinoma variants, therefore the appropriate diagnosis in small biopsy samples is very important.⁽⁴⁾

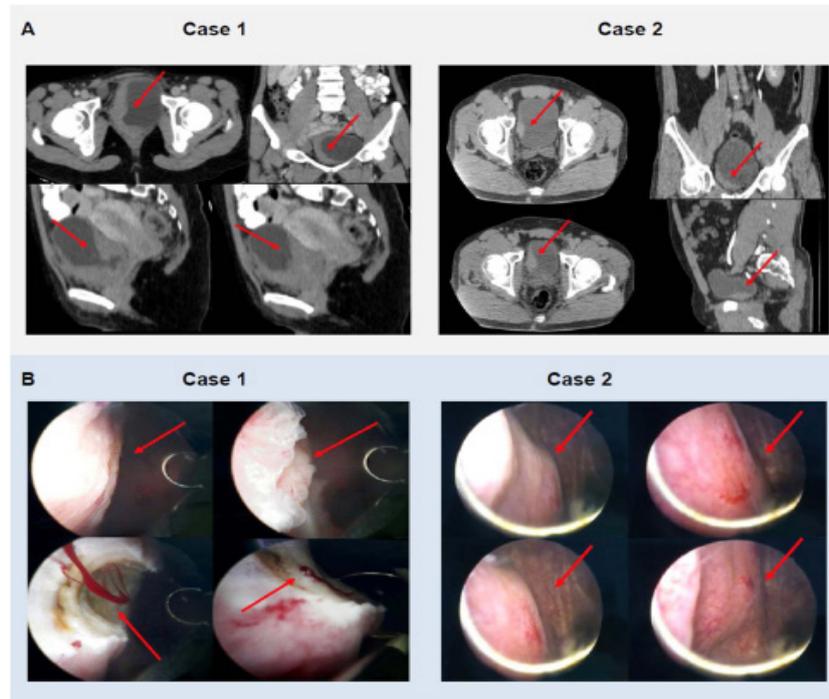


Figure 1. preoperative appearance of the Mass abdomen. A. abdominopelvic tomography. B. Cystoscopic appearance. Arrows indicate the mass located on the right side of the bladder Wall in both cases (Case 1: female, Case 2: Male).

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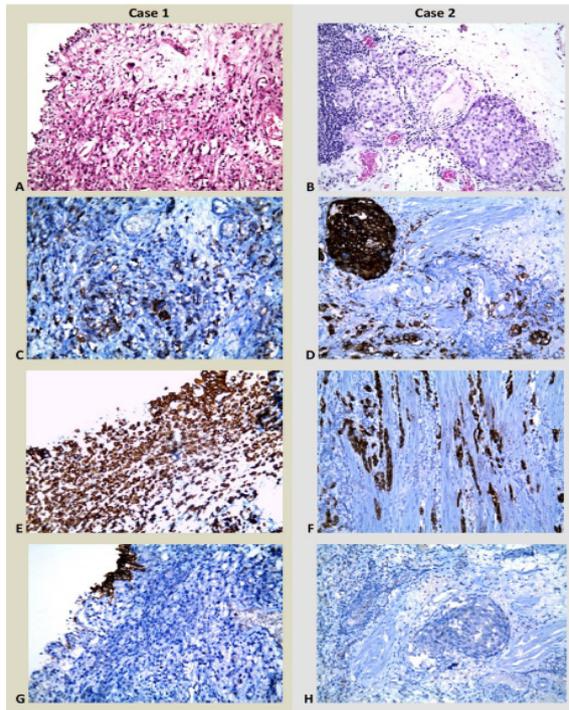


Figure 2. Microscopic views of the tumor. Hematoxylin-eosin staining: A, B: the appearance of PUC as discohesive (A) or groups (B). Immunohistochemical stainings: C, D: CD138 positive staining indicating PUC. E, F: CK7 positive staining in surface epithelium (E) and in PUC (E, F). G, H: E-cadherin positive staining in surface epithelium (G) and lost of membranous staining in PUC (G, H).

In the present study, we aimed to report and discuss two uncommon cases of bladder PUC of both sexes, which presented with renal colic or macroscopic hematuria to our hospital surprisingly at similar times.

CASE REPORT

Case 1: A 33-year-old female presented to our hospital with macroscopic hematuria.

Case 2: A 57-year-old male patient presented to the urology outpatient department with a history of left renal colic.

A solid lesion was observed in the right side wall of the bladder of the both cases in cystoscopy examination (**Figure 1**) and incomplete transurethral resections (TUR) were performed. Therewithal, abdomino-pelvic tomography revealed a mass which constituted a marked wall thickening and extension to the bladder perivesical adipose tissue (**Figure 1**). In both cases, microscopic evaluation of the TUR materials showed a high-grade tumor, arranged in discohesive cords and plasmacytoid features; in both cases penetrating the muscle. Immunohistochemically, tumors of both cases showed positivity for PANCK, CK7 and CD138, besides negativity for E-cadherin (**Figure 2**). In the light of these results, radical cystectomy and ileal loop operations were performed. In the pathological evaluations of radical materials, the tumors morphologically resembled the previous TUR samples. Also, tumors of these materials infiltrated the perivesical fatty tissue. The conditions of the patients were consulted with the oncology department and chemotherapies were planned.

After radical cystectomy the male patient was evaluated for distant metastasis by thorax CT and bone scintigraphy. However distant metastasis was not detected. The preoperative evaluation of the female patient for distant metastasis was performed by thoracic CT and eventually no pathological formation was determined. Due to the right iliac LAP metastasis and perivesical spread determined in the male patient and the tumor infiltrating perivesical fatty tissue and vaginal anterior wall of the female patient, gemcitabine and cisplatin-based combination chemotherapy was administered to both patients. The male patient was given 3 cycles of chemotherapy and there was no pathological involvement in the MR examination of the lower and upper abdomen. He was started to drug-free follow-up after taking the 4th cure chemotherapy. The female patient's 2nd cure chemotherapy is completed. After 3 cycles of systemic chemotherapy, upper and lower abdomen will be reevaluated with MR. If pathological formation is not found, treatment with 4 cycles of chemotherapy will be terminated and the patient will be followed up.

DISCUSSION

PUC of urinary bladder is an uncommon malignant neoplasm declared by recent World Health Organization (WHO) classification.⁽⁵⁾ Chung et al. (2017) reported an 8-year retrospective search of the archive at their institution identifying 22 patients with PUC. The median age of the cases were stated as 74 years (range 51-86) and only three of them were reported as female. Uncommonly, the 33-year-old female case presented in this study was a rare case both in terms of age and sex. Also, the researchers reported the percentage of muscle-invasive disease as 86%, and the distant metastases as 9% among the presented PUC cases.⁽⁶⁾ Muscle and perivesical adipose tissue invasion were present in the both cases of the present study. In the male case lymphovascular and perineural invasions of PUC were common and significant. Moreover, tumor metastasis was detected in 7 out of 22 lymph nodes detected. In another study of retrospective case series the rate of PUC was reported as 2.4%, which is the lowest rate among the UC variants. Furthermore, PUC was accounted for 7% of all deaths from UC at 1 year period.⁽⁷⁾ In accordance with the literature, PUC cases of the present study were also late presented and unfortunately advanced pathological tumor stage was detected in the radical materials.

The morphological similarity of PUC to other benign and malign lesions such as cystitis with plasma cell infiltration, plasma cell-derived neoplasms, lymphomas, lymphoepitheliomas and metastatic carcinoma of the breast and stomach is the main cause of misdiagnosis.⁽⁸⁾ PUC as the rare variant of UC should be considered in order to avoid misdiagnosis. In this regard, immunohistochemistry is an essential technique to make a correct diagnosis on small biopsies.

CD138 expression is reported as an important marker for PUC. It can be observed in other malignant tumors, such as plasmacytomas, melanomas, rhabdomyosarcomas, and other carcinomas as well as it can be positively staining in all variants of UCs and various other epithelial tumors of the urinary tract.⁽⁹⁾ Therefore, the pathological diagnosis should be based on the morphological aspect and the differential diagnosis should be considered widely.

Radical treatment must be provided to patients diagnosed with PUC. This view is also supported by Ohtaka et al. (2016) treated PUC successfully with a combination of radical cystectomy and adjuvant chemotherapy. Median overall survival of the PUC patients is lower than conventional urothelial carcinomas.⁽¹¹⁾ In this regard, current studies that express survival rates in wide series are limited. Cockerill et al. (2017) reported 46 patients with plasmacytoid carcinoma, 27 (59%) died of bladder cancer with a median survival of 1.7 years, and 10 patients died of other causes with a median follow up of 2.2 years. Of the 9 surviving patients, the median follow-up was reported as 10.3 years.

In conclusion, consideration of this rare variant by the pathologist is important for correct diagnosis and determining prognosis. Thus, the radical resection of this aggressive tumor variant, which is not predicted clinically and radiologically, provides a relatively prolonged survival of the patient.

CONFLICT OF INTEREST

There is no conflict of interest among the authors.

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