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Plasmacytoid variant of urothelial carcinoma of the bladder manifesting as bilateral ureteral and small bowel obstruction

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ABSTRACT

Plasmacytoid urothelial carcinoma (PUC) is a rare variant of bladder cancer characterized by distinct histopathology and advanced stage at diagnosis. Multimodal treatment is usually indicated. We present a case of PUC causing bilateral ureteral obstruction with subsequent renal failure followed shortly by malignant small bowel obstruction, demonstrating the need for a high degree of clinical suspicion in diagnosis of this aggressive subtype. Moreover, the local invasiveness of the disease cannot be understated, given that it can rapidly spread with little radiologic evidence of progression until it is at an advanced stage.

1. Introduction

Plasmacytoid urothelial carcinoma (PUC) is a rare variant of bladder cancer estimated to comprise approximately 1–3% of cases.[1] First reported by Sahin and Zuckerberg in 1991, there have been approximately 275 cases described in the literature, with most cases presenting as advanced or late-stage disease portending a poor overall prognosis and median survival of 15 months.[2] Unlike other variants of urothelial carcinoma, PUC is further characterized by its tendency to be locally invasive with reports describing intraperitoneal and retroperitoneal spread.[2,3] Aggressive multimodal therapy for PUC is the mainstay of treatment. However, due to the nature of the disease, high clinical suspicion and early diagnosis are key to prolonging survival. We present a case of PUC involving a rather indolent lesion of the bladder with rapid progression to multiple clinical sequelae including both acute renal failure and bowel obstruction. The lack of definitive radiologic progression of the disease given initial findings and advanced stage at ultimate diagnosis illustrate the degree of clinical suspicion required to make a timely diagnosis and early multimodal treatment of this disease.

2. Case report

Our patient is a 71-year-old Caucasian male who was initially found to have a 5.7 cm infra-renal abdominal aortic aneurysm on non-contrasted CT abdomen/pelvis after presenting with vague abdominal pain, for which he underwent endovascular repair. He was found to have incidental asymmetric bladder wall thickening on imaging, for which urologic consultation was obtained. Patient underwent office cystoscopy 2 weeks later revealing sessile lesions on the dome and lateral walls with urine cytology suggesting high-grade urothelial carcinoma. He was scheduled for transurethral resection, however two days later the patient presented with acute renal failure requiring emergent hemodialysis. A non-contrast CT revealed new mild bilateral hydroureteronephrosis. He underwent bilateral nephrostomy tube placement with normalization of renal function, and etiology of this was initially thought to be related to endograft-induced aortitis causing bilateral ureteral obstruction. After a subsequent admission for Klebsiella septicemia 1.5 months following initial cystoscopy, patient ultimately underwent transurethral resection revealing multifocal sessile tumors and notably a contracted and fixed bladder. Bilateral antegrade nephrostograms revealed bilateral distal ureteral strictures approximately 4–6 cm in length. Bilateral ureteral wash cytology was obtained.

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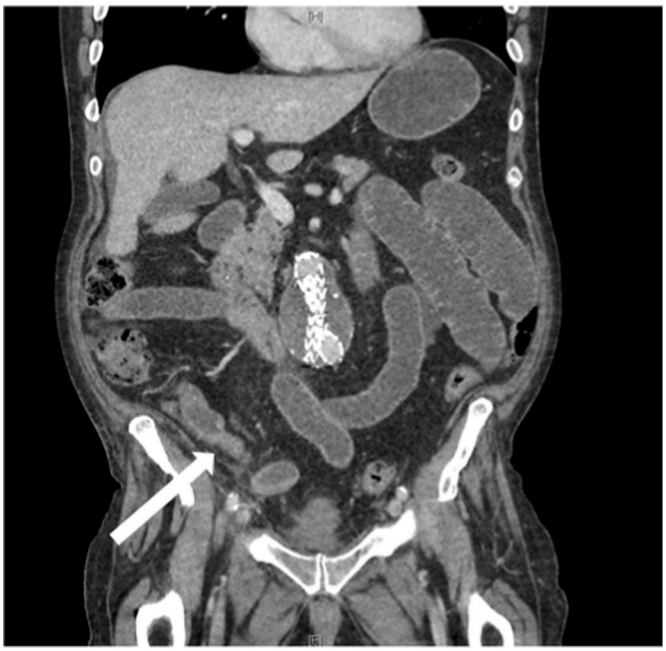

Fig. 1. CT scan of the abdomen/pelvis with IV contrast showing small bowel obstruction with evidence of a transition point at the terminal ileum.

Patient was discharged the following day, however he presented again two days following discharge with abdominal distension and pain. He was diagnosed with a distal small bowel obstruction on CT scan despite no previous abdominal surgery (Fig. 1). After failure of non-operative management, patient was taken to the operating room for exploratory laparotomy. Diffuse inflammation involving a strictured segment of terminal ileum and appendix was noted requiring resection of 10 cm of ileum with appendectomy. Intra-operative frozen sections revealed urothelial carcinoma. These correlated with findings on transurethral resection. In addition, urine cytology from the upper tracts was positive for high-grade urothelial carcinoma. The patient continued to deteriorate despite surgical intervention and was ultimately seen by oncology, but not considered a candidate for systemic therapy due to poor functional status. The decision was ultimately made to transfer the patient to hospice care, less than 3 months from initial evaluation. Final pathology of exploratory laparotomy revealed the presence of carcinoma in the muscularis propria of the ileum, the mesoappendix and serosal surface of the appendix, and mesenteric fat. Transurethral resection specimen revealed diffuse high-grade invasive urothelial carcinoma with plasmacytoid morphology and eccentrically-placed nuclei in discohesive clusters (Fig. 2a). Invasive cells with irregular nuclear profiles were observed clustering in the muscle of the terminal ileum (Fig. 2b). Immunohistochemistry (IHC) showed that specimens were positive for CK20, CK7, GATA 3, CD138 with loss of e-cadherin expression. Prognostic stage IVB: pT2,cN0, pM1b with no evidence of enlarged pelvic or retroperitoneal lymphadenopathy.

3. Discussion

PUC is a rare variant of bladder cancer that is particularly aggressive in nature and locally invasive, and has distinct behavior separating it from most other urothelial carcinoma. Oftentimes, lower urinary tract symptoms and circumferential bladder wall thickening with indurated mucosa are the only initial findings with hematuria only presenting in advanced stages.[4] In addition, the way in which this variant locally invades and behaves much like linitis plastica in certain GI malignancies provides some insight into its unique biology. The classical loss of e-cadherin expression exhibited in PUC contributes to its aggressive cellular invasiveness. In addition to the presence of CD138, the defect in e-cadherin expression is associated with a discohesive pattern of plasmacytoid differentiation. There have been a few cases of bowel obstruction and hydronephrosis as a results of locally invasive PUC,[3] however the timing and severity of clinical sequelae in this patient presents a unique case illustrating the need for timely diagnosis and high clinical suspicion in a patient with seemingly localized disease.

Management of PUC is poorly defined. A multimodal approach including surgery and perioperative chemoradiation is suggested.[5] There may be little prognostic difference with respect to muscle invasion and any plasmacytoid histology should be treated aggressively. There was no difference in overall survival in patients treated with neoadjuvant chemotherapy compared to upfront surgery with PUC.[5] Dayyani et al. described treatment in fifteen patients with metastatic PUC; 60% were treated with cisplatin-based chemotherapy with an overall response rate of 53%.[5] However, survival for those with metastatic disease was just over a year from the initiation of chemotherapy. Additionally, PUC has a strong predilection for recurrence in the peritoneum and poorer local recurrence-free survival than conventional urothelial carcinoma.[1] Because of this tendency for peritoneal metastasis, epithelial tumor markers such as carcinoembryonic antigen, cancer antigen (CA) 125, and CA19-9 are suggested to assess for early disease progression prior symptomatic or radiographic findings.[5]

4. Conclusion

Plasmacytoid urothelial carcinoma can present with locally advanced and metastatic disease resulting in acute renal failure and small bowel obstruction. Early diagnosis of PUC is challenging and arguably the most important prognostic factor in influencing survival. The invasive nature and dismal prognosis of PUC demands increased

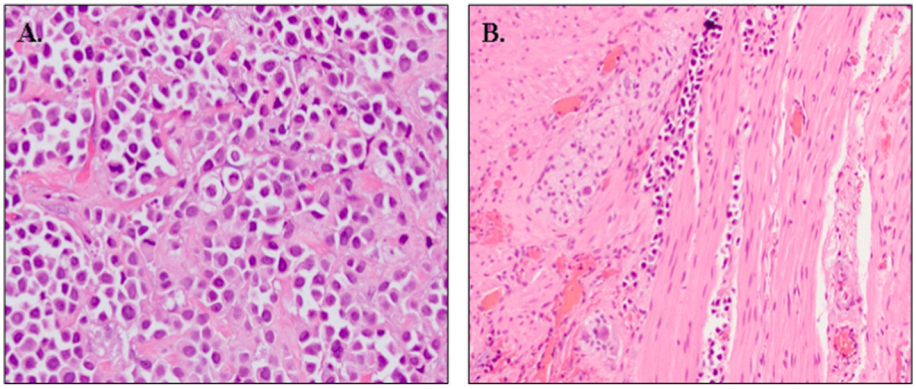

Fig. 2. High power H&E of malignant cells with plasmacytoid morphology from TURBT (**A**); Invasive urothelial carcinoma cells clustering within the muscle of the terminal ileum (**B**).

multidisciplinary awareness in recognizing the characteristic markers and rapidly evolving clinical symptoms that are consistent with this rare malignancy.

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Alex Carsel: original drafting, review and editing, methodology, data curation.

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Ashwin Raghavan: original drafting, review and editing, methodology.

Samer Altharthi: review and editing.

Cara Gatto-Weis: investigation, data curation.

Jorge Ortiz: supervision, review and editing, conceptualization.

Puneet Sindhwani: supervision, review and editing.

Firas Petros: supervision, review and editing, conceptualization.

Declaration of competing interest

None.

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