

Oncology

High Grade Mucinous Urothelial Carcinoma of the Renal Collecting System: A Case Report



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ABSTRACT

High grade mucinous urothelial carcinoma is a rare pathological variant. There is still controversy as to its nomenclature and classification. We report the case of a 64 year old female with history of pelvic pain who was incidentally discovered to have a left upper pole renal mass. Left nephroureterectomy was performed and histopathological examination revealed high grade mucinous urothelial carcinoma. Accurate diagnosis of this distinct pathological entity will allow for better understanding of phenotypic behavior and inform best treatment strategies.

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Introduction

In the majority of cases, urothelial carcinoma arises from the bladder, with only 7% of cases originating in the upper urinary tract.¹ It has numerous known histologic variants, many of which are recognized by the World Health Organization/International Society of Urologic Pathologists (WHO/ISUP) 2004 classification.^{2,3} Radiologically, as with most other transitional cell tumors of the upper tract, they are indistinguishable from renal parenchymal lesions. We present a case of high-grade mucinous upper tract urothelial carcinoma (UTUC) originating in the renal collecting system, a rare variant as yet to be classified by the WHO/ISUP.

Case presentation

Institutional review board approval was obtained for this case report. A 64 year old female with a past medical history of essential

hypertension, diabetes mellitus type 2, and obesity was initially referred by her primary care physician (PCP) to the urology clinic of an academic, tertiary care medical center for evaluation of an incidental left renal mass seen on abdominal ultrasound. Ultrasound, initially ordered by the PCP due to a complaint of pelvic pain, demonstrated a heterogeneous, hypoechoic structure in the upper pole of the left kidney measuring 4.8 × 3.9 × 4.6 centimeters (cm), with peripheral vascularity.

Upon presentation to the urology clinic, the patient reported intermittent “dark urine” for 2 months and occasional abdominal pain for 6 months. She denied weight loss, smoking and any family history of urological malignancy. Her urinalysis revealed 25–50 red blood cells, and laboratory investigations revealed a serum creatinine of 0.96 mg per deciliter.

Subsequent computed tomography of the abdomen and pelvis with intravenous contrast revealed a large, heterogeneously enhancing mass measuring up to 5.3 cm in coronal plane and located in the upper pole of the left kidney (Fig. 1). Based on the imaging, it was unclear if the mass originated from the collecting system or renal parenchyma. After discussion with the patient, the decision was made to proceed with cystoscopy, left ureteroscopy and possible biopsy.

Ureteroscopy revealed a macroscopically high-grade appearing papillary mass in the left upper calyx with associated bleeding and necrotic debris. The mass was biopsied and the pathology report revealed fragments of soft tissue with necrosis, acute and chronic

Abbreviations: UTUC, upper tract urothelial carcinoma; WHO/ISUP, World Health Organization/International Society of Urologic Pathologists; PCP, primary care physician; cm, centimeter.

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Figure 1. Coronal CT Image demonstrating a heterogeneous, hypoechoic mass in the upper pole of the left kidney.

inflammation and overlying benign urothelial epithelium. While the results of the biopsy did not demonstrate malignancy, given the suspicious imaging results and the visual appearance on ureteroscopy, the patient was counseled regarding her management options and elected to undergo definitive management in the form of a left nephroureterectomy.

An open, two incision approach for nephroureterectomy was undertaken. After excision of the ureteral orifice and bladder cuff, the remaining defect was repaired intravesically. The patient remained stable throughout the procedure and recovered without major complication. She was discharged on post-operative day

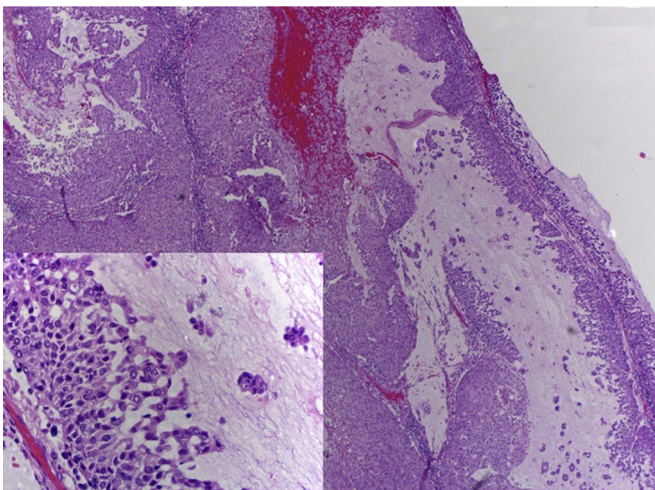


Figure 2. Invasive mucinous urothelial carcinoma of the kidney. The tumor is composed of invasive malignant urothelial cell groups localized in an abundant mucinous background. High power of the tumor (bottom left) shows neoplastic urothelial cells forming groups and cords within a mucin pool. The neoplastic cells have a characteristic eosinophilic cytoplasm (hematoxylin-eosin, original magnification $\times 100$, $\times 400$).

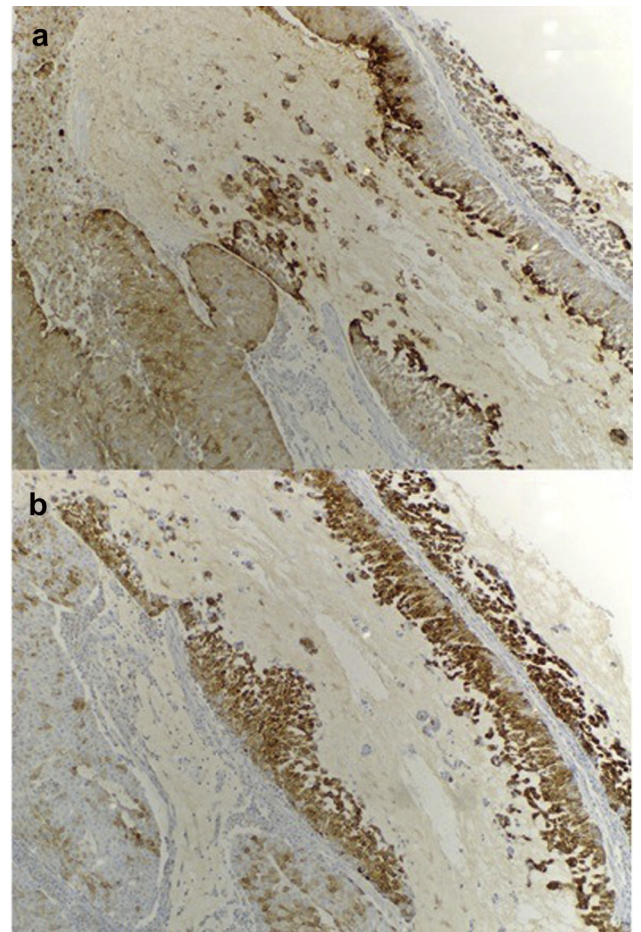


Figure 3. a) Neoplastic cells demonstrating positivity for MUC1 (immunohistochemistry, original magnification $\times 200$). b) Neoplastic cells demonstrating positivity for MUC2 (immunohistochemistry, original magnification $\times 200$).

(POD) four. A cystogram performed on POD 12 did not demonstrate any evidence of contrast leak and the Foley catheter was removed.

On gross pathology, a tan friable hemorrhagic mass with papillary fronds, measuring $5 \times 5 \times 4.5$ cm was found in the upper pole of the kidney with no evidence of infiltration of the renal capsule or perinephric fat. Microscopically, the tumor was found to invade the renal parenchyma (Fig. 2). Neoplastic cells were positive for CK7, GATA3, p63, MUC1, MUC2 (Fig. 3), with occasional MUC5AC-positive cells. Immunostains for CK20, PAX2 and PAX8 were negative. Additionally, mucicarmine and PAS stains highlighted extracellular mucin. Histopathological examination supported the diagnosis of high-grade mucinous urothelial carcinoma, pathological stage pT3NxMx.

Six months post-operatively, there has been no clinical or radiographic evidence of local recurrence or distant metastases. Surveillance cystoscopy at 5 months post-operatively revealed no suspicious bladder lesions.

Discussion

Although not yet formally recognized by the WHO/IUSP 2004 classification, mucinous urothelial carcinoma, also referred to as urothelial carcinoma with abundant myxoid stroma, is a distinct pathological entity.⁴ Although mucin production can be a hallmark of other urothelial carcinoma variants, the absence of true gland formation, gland-like lumina and atypical spindle cells distinguish

this particular variant from those included in the current classification.⁴

With few reported cases of mucinous urothelial carcinoma, only one of which originated in the kidney, the correlation between histopathology and disease course and progression is poorly understood.⁵ A case series of 13 patients with mucinous urothelial carcinoma of the bladder suggests that while this subtype exhibits variable aggressiveness, it may, overall, be less aggressive than the classically-defined urothelial carcinoma.⁴

It is important that surgical pathologists be aware of these rare subtypes, which may be associated with different outcomes from conventional urothelial carcinoma. Correct characterization of these distinct entities may have diagnostic, therapeutic or prognostic implications significantly impacting management.

Conclusion

Mucinous urothelial carcinoma of the renal pelvis is an unrecognized histologic variant of a relatively rare malignancy. Accurate

diagnosis as a distinct variant may allow for better understanding of its clinical course and guide treatment.

Conflicts of interest

None.

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