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Primary adenocarcinoma of the renal pelvis in spina bifida patient with history of simple cystectomy

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A B S T R A C T

Primary renal adenocarcinomas comprise of less than 1% of renal and ureteral epithelial tumors. We present a case of a 67-year-old male with a history of simple cystectomy who underwent left nephroureterectomy for primary enteric-type renal adenocarcinoma with cystitis glandularis features. Pathological examination confirmed grade 2 pT1N0MX primary enteric-type renal adenocarcinoma. The patient underwent left open radical nephroureterectomy, with an uneventful post-operative course. Surgical excision is the mainstay treatment, while chemotherapy and radiation are potential adjuncts. Prognosis remains poor, with a 50% overall survival rate within two years of surgery. Further research is needed to enhance treatment recommendations.

1. Introduction

The most prevalent primary malignant cancers of the renal pelvis are urothelial carcinoma and renal cell carcinoma, which collectively account for over 95% of cases.[1] In contrast, primary renal adenocarcinomas are exceptionally rare, constituting less than 1% of all renal and ureteral epithelial tumors. These tumors exhibit distinct histologic variants, namely enteric, mucinous, or signet ring cell types.[2,3] Notably, they can also present with additional features such as cystitis cystica or cystitis glandularis, which are proliferative disorders of mucus-producing glands of the urothelium, characterized as small cystic cavities with varying features.[4] In this report, we present a case of a patient with history of simple cystectomy with ileal conduit who subsequently underwent a left nephroureterectomy for primary enteric-type renal adenocarcinoma with features of cystitis glandularis.

2. Clinical course

Our patient is a 67-year-old male with a past medical history of spina bifida who had a simple cystectomy with ileal conduit in 1970 for neurogenic bladder. When he presented to our clinic, he reported a history of chronic left flank pain with recurrent urinary tract infections. Work up with a non-contrast CT scan was significant for a 7.5cm left renal mass with a markedly dilated left renal collecting system. Multiple colonic lesions were also identified, thus endoscopic evaluation was recommended. Colonoscopy revealed evidence of minor diverticulosis and internal hemorrhoids but was otherwise negative for malignancy. Renal nuclear medicine scan revealed a hydronephrotic left kidney that contributed to 27% of total renal function and hydroureter compatible with obstruction.

The patient was then brought to the operating room for diagnostic ureteroscopy/looposcopy. Looposcopy showed mild stenosis at the level of the fascia, however, no masses or abnormal lesions were noted within the conduit. Nephroscopy demonstrated a mass located in the renal pelvis that was biopsied, with pathology revealing an enteric-type adenocarcinoma. Staging scans of the chest, abdomen, and pelvis did not show evidence of metastatic disease. After careful consideration, the patient elected for a left open radical nephroureterectomy via a modified flank incision.

The surgery and postoperative course were uncomplicated, and the patient was discharged on post-op day 5. Final pathology confirmed grade 2 pT1N0MX primary enteric-type renal adenocarcinoma and features of cystitis glandularis.

3. Discussion

Primary renal adenocarcinoma is a rare finding that can present with nonspecific symptoms of flank/loin pain and hematuria.[5] This type of malignancy does not present with any characteristic radiologic features, making intraoperative cytology or frozen section an important initial step in confirming the presence of adenocarcinoma.[6] Case reports have demonstrated instances of patients with primary renal adenocarcinoma having viscous urine containing spherical clusters of cells with basophilic cytoplasm, though overall screening and confirmatory testing via

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Table 1
Summarized data reported in the literature for patients with primary adenocarcinoma of the renal pelvis.

Study	Patient Data and Significant History	Tumor Characteristics	Management	Outcome
Xiong et al. (2016)[2]	55-year-old male + hematuria + flank pain	Pathological examination: moderately differentiated adenocarcinoma of the renal pelvis, ureter, and urinary bladder	Right radical nephrectomy, ureterectomy, radical cystectomy and left ureterocutaneostomy	No evidence of tumor recurrence at 6-month follow-up
Lai et al. (2016)[3]	40-year-old male + flank pain	Pathological examination: primary enteric-type mucinous adenocarcinoma of the renal pelvisCEA: 31.4 ng/ml (Normal: 25 ng/ml)	Right radical nephrectomy	CEA returned to normal range by 1-month follow-upNo evidence of tumor recurrence at 14-month follow-up
Agrawal et al. (2021)[5]	60-year-old male + flank pain + hx of renal calculi	Pathological examination: grossly dilated pelvicalyceal system filled with gelatinous material and multiple brownish papillary nodulesFinal pathological stage: pT3aNx primary mucinous adenocarcinoma of the renal pelvis	Open right radical nephrectomy	No evidence of tumor recurrence at 18-month follow-up
Abbas et al. (2014)[6]	56-year-old male + flank pain	Histological examination: tumor with cysts, and papillae lined by pseudostratified columnar epithelium with hyperchromatic nuclei and vacuolated cytoplasm. Mucin pools with scattered poorly differentiated signet ring tumor cells were seen infiltrating into the renal cortexFinal pathological stage: pT3pN0 primary mucinous adenocarcinoma of the renal pelvis with in-situ adenocarcinoma of the ureter	Nephrectomy with partial ureterectomy	Metastasis involving the chest wall at 6-month follow-up
Gupta et al. (2020)[10]	51-year-old male + hx of renal calculi	Histopathological evaluation: well-differentiated tubular adenocarcinoma enteric type with muscularis involvementFinal pathological stage: pT2N0Mx papillary urothelial carcinoma with villoglandular differentiation	Robot-assisted laparoscopic radical nephroureterectomy, with bladder cuff excision and regional lymphadenectomy.	No evidence of tumor recurrence at 3-month follow-up
Ho et al. (2008)[8]	48-year-old asymptomatic female	Microscopic examination: adenocarcinoma with mucin production. Focal architectural and cellular atypia were notedCEA level 17.9 ng/mL (Normal: 25 ng/ml)	Hand-assisted laparoscopic radical nephrectomy	CEA returned to normal range by 1 month follow-upNo evidence of tumor recurrence at 9-month follow-up
Han et al. (2015)[11]	50-year-old male + smoker + flank pain + hx of renal calculi	Microscopic examination: renal pelvis composed of pools of mucus with clumps or strands of neoplastic glandular epithelium and the pelvic mucosa consisted of tall columnar cells that tended to stratify into two or more layers, with perfuse irregular infoldings and protrusions into the surrounding stroma	Radical nephrectomy	No evidence of tumor recurrence at 20-month follow-up
Li et al. (2020)[13]	66-year-old male + flank pain + fever + hx of renal calculi	Histological examination: tumor with intestinal metaplasia, glandular acini, and multiple extracellular mucins	Open radical nephrectomy	No evidence of tumor recurrence at 12-month follow-up
Raphael et al. (2011)[14]	56-year-old male + flank pain	Histological examination: tumor with glands, cysts, and papillae lined by pseudostratified columnar epithelium with hyperchromatic nuclei and vacuolated cytoplasm. Mucin pools with individual poorly differentiated signet ring tumor cells were seen infiltrating into the renal cortex	Right radical nephrectomy	Recurrence in the right renal fossa and metastasis involving the lumbar/cervical spine at 12-month follow-up
Megumi et al. (1998)[15]	16-year-old male + hematuria	Histopathological examination: tumor extending from the renal mucosa to the lumen showing papillary proliferation and mucin in some tumor cellsFinal pathological stage: pTl, INFcr, grade 2 papillary adenocarcinoma	Left nephrectomy	No evidence of tumor recurrence or metastasis at 12-month follow-up
Sisodia et al. (2012)[16]	45-year-old male + flank pain	Microscopic examination: tumor comprised of papillary and glandular pattern lined by a single layer of columnar epithelium with scattered goblet cells, which resembled an intestinal mucosa.	Radical nephrectomy	No evidence of tumor recurrence or metastasis at 1-month follow-up

urine analysis is not well-established within the literature.[7] Certain tumor markers such as CEA and CA19–9 may be elevated in patients with primary renal adenocarcinomas. As these markers are not typically expressed in a normal adult's serum, they can be helpful in the diagnosis of these cancers and can be utilized to assess treatment response.[8]

Due to the enteric features of our patient's adenocarcinoma and the colonic lesions present on the CT, performing a colonoscopy was imperative to rule out a possibility of a primary tumor within the colon. With a negative colonoscopy, we concluded this case presented a primary renal adenocarcinoma.

It is hypothesized that the pathogenesis of renal adenocarcinoma is due to persistent inflammation caused by chronic infection, urinary calculi, or hydronephrosis.[9] As a result of the long-standing injury caused by inflammation, glandular metaplasia of the urothelium ensues and eventually leads to dysplasia or the development of adenocarcinoma.[7] Our patient reported a history of recurrent UTIs, which could have contributed to the etiology of his disease. Those with urinary diversions may be at higher risk for renal adenocarcinoma, as urinary infections are potential long-term complications in this population, though correlative analyses investigating this hypothesis do not currently exist.[10]

The treatment protocol for primary renal adenocarcinoma remains unclear due to the rarity of the disease; as demonstrated in Table 1, various management approaches have been adopted for similar cases reported in the literature. Similar to other cases, our patient underwent a complete nephroureterectomy; scans at 3 months following surgery revealed no evidence of disease.[5] Nevertheless the prognosis of adenocarcinoma of the renal pelvis or ureter after surgical removal remains poor, with a 50% overall survival within two years of surgery.[11] Chemotherapy and radiation may play a role in select cases.[12] Further

research into identifying factors that contribute to prognosis are necessary to help direct both treatment recommendations and goals of care discussions.

4. Conclusion

Primary enteric adenocarcinomas of the renal pelvis/ureter are rare and can be associated with chronic inflammation and recurrent UTIs. As such, patients with urinary diversions may be at higher risk to develop these tumors. Management with surgical excision is most common, though other form of treatments have been reported, including radiation and chemotherapy.

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