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A Rare Case of Mucinous Adenocarcinoma of the Renal Pelvis Masquerading as a Non-Functioning Pyonephrotic Kidney

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ABSTRACT

Background: Primary mucinous adenocarcinoma of the renal pelvis is an exceptionally rare malignancy, with fewer than 100 cases reported in the literature [1]. The absence of specific clinical and radiological features often leads to delayed or incorrect preoperative diagnosis [2].

Case Presentation: We report a 54-year-old male who presented with long-standing flank pain, cloudy urine, and a non-functioning kidney, clinically and radiologically suggestive of pyonephrosis. The patient underwent laparoscopic nephrectomy. Histopathological examination revealed moderately differentiated mucinous adenocarcinoma arising from the renal pelvis. Despite surgical management, the disease showed aggressive behavior with early metastatic progression.

Conclusion: Mucinous adenocarcinoma of the renal pelvis should be considered in the differential diagnosis of long-standing non-functioning kidneys, particularly in the setting of chronic inflammation [1]. Early recognition remains challenging, and prognosis is poor despite surgical intervention [2].

Keywords: Renal pelvis; Mucinous adenocarcinoma; Pyonephrosis; Non-functioning kidney; Rare malignancy

INTRODUCTION

Primary adenocarcinoma of the renal pelvis is rare, accounting for less than 1% of upper tract malignancies [2]. Among these, the mucinous subtype is exceedingly uncommon.[1] Chronic irritation of the urothelium due to infection, obstruction, or renal calculi is believed to induce glandular metaplasia, which may progress to adenocarcinoma.[3] Due to nonspecific symptoms and lack of characteristic imaging features, most cases are diagnosed only after nephrectomy performed for presumed benign pathology [1,2]. We present a rare case of mucinous adenocarcinoma of the renal pelvis masquerading as a non-functioning pyonephrotic kidney.

CASE REPORT

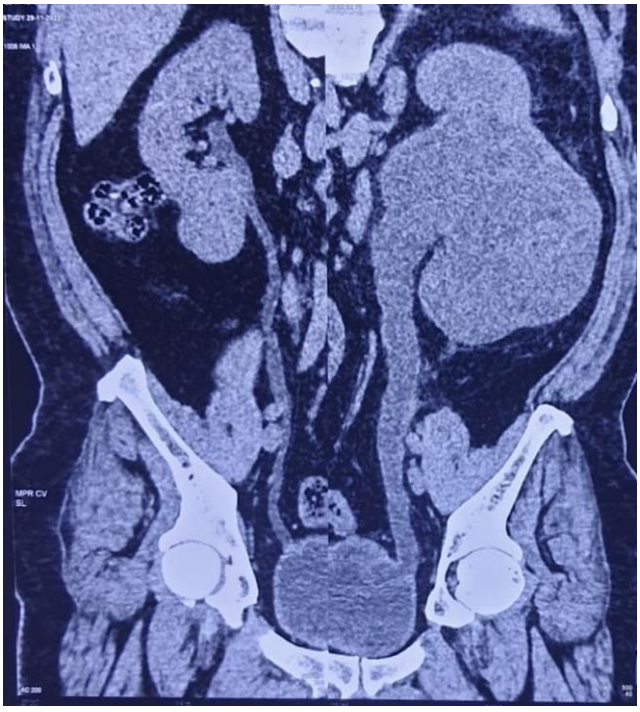
A 54-year-old DM, HTN male presented with complaints of cloudy, turbid urine, dull aching pain in the left loin, and generalized weakness for one year. There was a prior history of left percutaneous nephrolithotomy with double-J stenting performed four years earlier.

On physical examination, a vague, ill-defined lump was palpable in the left lumbar region. A healed scar of prior PCNL was noted. Laboratory investigations revealed normal renal function with a serum creatinine of 1.02 mg/dL. Urinalysis showed 2–5 pus cells per high-power field.

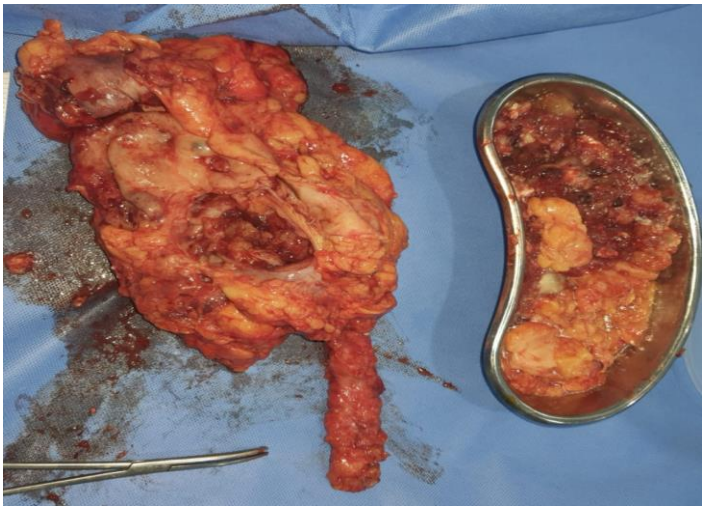
Contrast-enhanced CT of the kidneys, ureters, and bladder demonstrated a grossly hydronephrotic left kidney with cortical thinning and low-attenuation contents (HU ~40), suggestive of pyonephrosis [1]. A DTPA renogram confirmed a non-functioning left kidney.

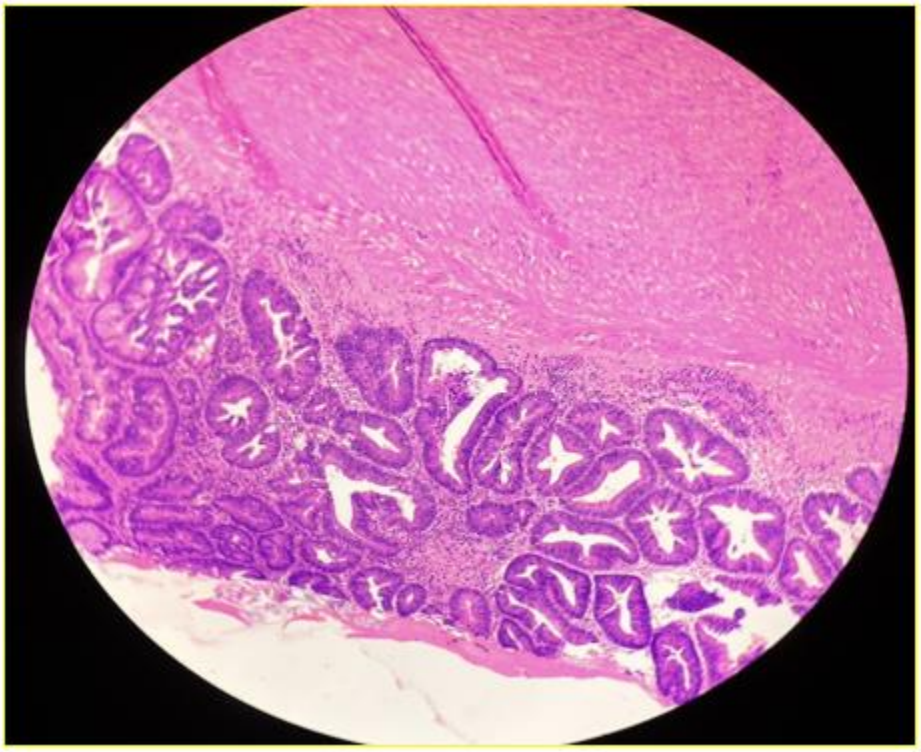
In view of a non-functioning, infected kidney, the patient underwent transperitoneal laparoscopic left nephrectomy. Intraoperatively, the kidney was grossly enlarged and adherent to surrounding tissues. On the cut section, the pelvicalyceal system was filled with abundant gelatinous material.

Gross pathological examination revealed a 10 × 7.5 × 2.5 cm kidney specimen with mucinous contents occupying the renal pelvis. Microscopic evaluation showed moderately differentiated adenocarcinoma with mixed enteric and mucinous patterns arising from the urothelium of the pelvicalyceal system. Tumour cells were observed floating within large extracellular mucin pools.



The postoperative course was uneventful. Subsequent PET-CT revealed axial and appendicular skeletal metastases. The patient received palliative radiotherapy and chemotherapy with gemcitabine and oxaliplatin. However, he tolerated only one cycle of chemotherapy. One month later, he developed lung and skull base metastases and succumbed to the disease within six months of diagnosis [1].





DISCUSSION

Primary mucinous adenocarcinoma of the renal pelvis is a rare and aggressive malignancy. Chronic inflammation and long-standing obstruction are considered key etiological factors, leading to glandular metaplasia of transitional epithelium [3]. The majority of reported cases originate from Asian countries, particularly India [1].

Preoperative diagnosis is challenging due to nonspecific symptoms and imaging findings that often mimic pyonephrosis or xanthogranulomatous pyelonephritis [1,2]. Consequently, diagnosis is frequently established only on histopathological examination following nephrectomy.

Radical nephrectomy remains the mainstay of treatment for localized disease [2]. The role of adjuvant chemotherapy or radiotherapy is not well defined due to the rarity of the condition and lack of standardized treatment protocols. Prognosis is generally poor, with most patients developing early metastatic disease and limited survival [1].

CONCLUSION

Primary mucinous adenocarcinoma of the renal pelvis is an exceptionally rare entity that may clinically and radiologically mimic benign inflammatory conditions [1]. High clinical suspicion is warranted in patients with long-standing non-functioning kidneys and a history of chronic inflammation [3]. Surgical excision remains the primary treatment modality, though overall prognosis remains unfavorable [2].

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