

A Case of Primary Mucinous Adenocarcinoma of the Renal Pelvis: A Rare Occurrence

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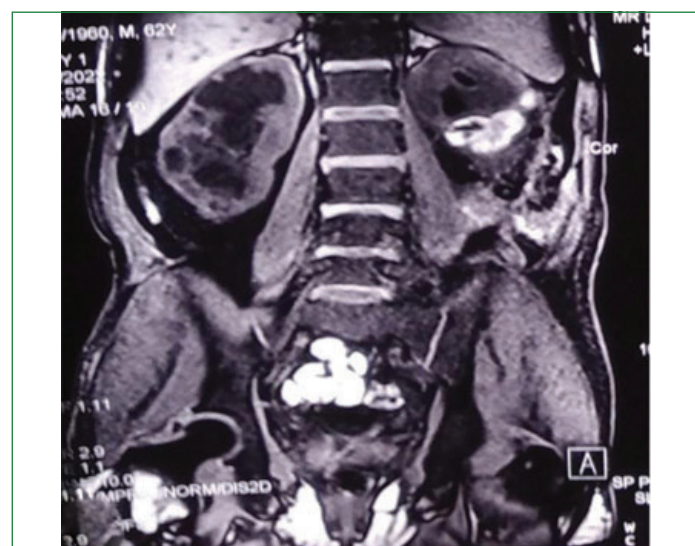
ABSTRACT

Adenocarcinoma represents only 1% of renal pelvis malignancies and can have three subtypes: tubulovillous, mucinous, and papillary non intestinal. Primary mucinous adenocarcinomas are very rare in the renal pelvis. We report a case of a 62-year-old male patient who presented with bilateral dull aching flank pain for 15 years. No flank tenderness was observed on clinical examination. Magnetic Resonance Imaging of the Kidney, Ureters, and Bladder (MRI KUB) showed marked dilation of the left kidney's pelvicalyceal system with irregular thinning of renal parenchyma, as well as a large filling defect. Percutaneous Nephrolithotomy (PCNL) obtained gelatinous material, and the Carcinoembryonic Antigen (CEA) level was found to be high. The patient underwent left radical nephrectomy, and gross examination of the kidney revealed staghorn calculi and multiple cavities filled with pus and gelatinous material. Microscopic examination showed atypical glands infiltrating the parenchyma with abundant extracellular mucin. Immunohistochemical (IHC) examination showed cytoplasmic positivity for CK7, CK20, and CEA, while PAX8 and GATA3 were negative. The Ki67 labeling index was more than 20%. The case was diagnosed as mucinous adenocarcinoma of the renal pelvis.

Keywords: Carcinoembryonic antigen, Immunohistochemistry, Kidney pelvis, Mucinous tumours, Urothelium

CASE REPORT

A 62-year-old male patient presented with bilateral dull aching flank pain for the last 15 years. He had no complaints of dysuria, haematuria, vomiting, etc. Six months ago, he developed severe pain with fever, chills, and rigors. He was admitted to a local hospital. His complete blood reports and peripheral blood revealed normocytic normochromic anaemia (Haemoglobin-10.1 g/dL). His blood urea nitrogen was increased (40 mg/dL), and creatinine levels were high (1.9 mg/dL). On ultrasonography, a calculus was found in the left renal pelvis with moderate hydronephrosis. Based on clinical, biochemical, and radiological examinations, he was diagnosed with chronic kidney disease due to nephrolithiasis with obstructive uropathy. No flank tenderness was present on clinical examination. The MRI KUB report showed moderate dilation of the right renal pelvicalyceal system with irregular thinning of renal parenchyma and marked dilation of the pelvicalyceal system of the left kidney with irregular thinning of renal parenchyma, as well as a large filling defect within [Table/Fig-1].



[Table/Fig-1]: MRI KUB showing large filling defect in left kidney.

The patient underwent right and left prone PCNL consecutively. A 500 mL thick gelatinous material was aspirated during left PCNL. The patient's CEA level was found to be >1000 ng/mL. The patient then underwent left radical nephrectomy. On gross examination, the left kidney measured 13×7×6 cm with an attached ureter measuring 6 cm in length. Staghorn calculi were identified upon cutting open. Multiple cavities were identified filled with pus and gelatinous material. The largest cavity measured 4 cm in diameter [Table/Fig-2].

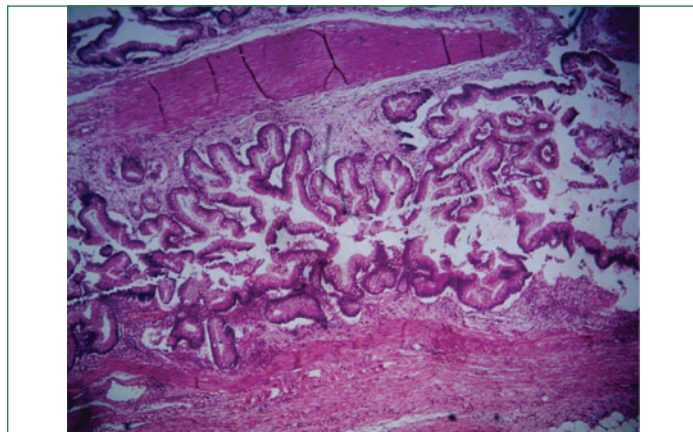


[Table/Fig-2]: Multiple cystic cavities in left kidney on gross examination.

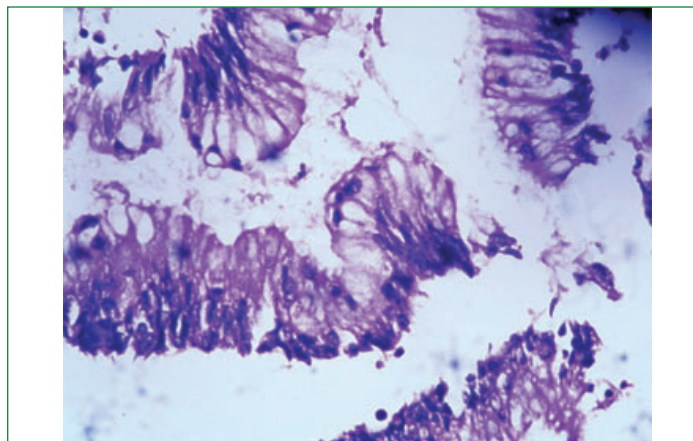
On microscopic examination, sections from different areas of the renal pelvis and cyst wall showed glands and papillary structures lined by atypical cells with pleomorphic, hyperchromatic nuclei and intracytoplasmic mucin. The glands and papillary structures infiltrate the renal parenchyma [Table/Fig-3,4]. Extracellular mucin was also observed (>50%). Histomorphological features were consistent with mucinous adenocarcinoma of the renal pelvis. TNM staging-T3NxMx.

On immunohistochemical examination, CK7, CK20, and CEA showed cytoplasmic positivity [Table/Fig-5-7]. PAX8, CDX2, and

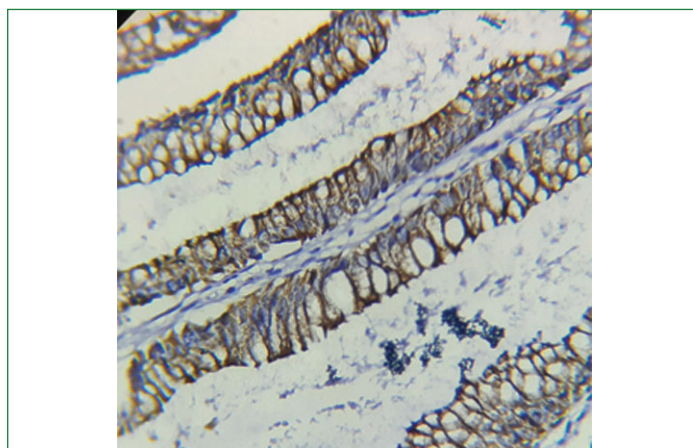
GATA3 were negative, and the Ki67 labeling index was more than 20% [Table/Fig-8].



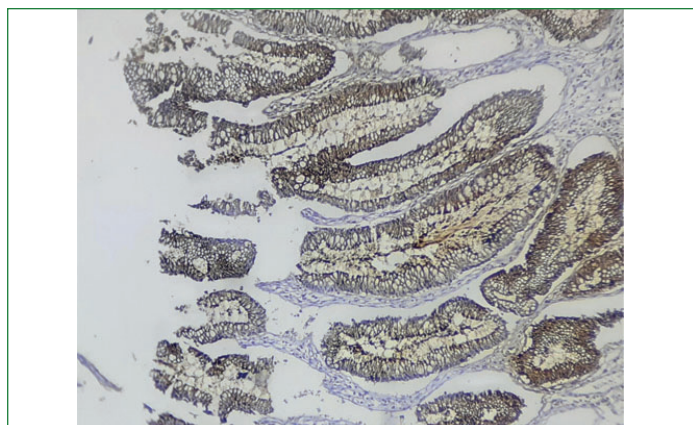
[Table/Fig-3]: Atypical glands infiltrating the renal parenchyma (H&E stain, 40X).



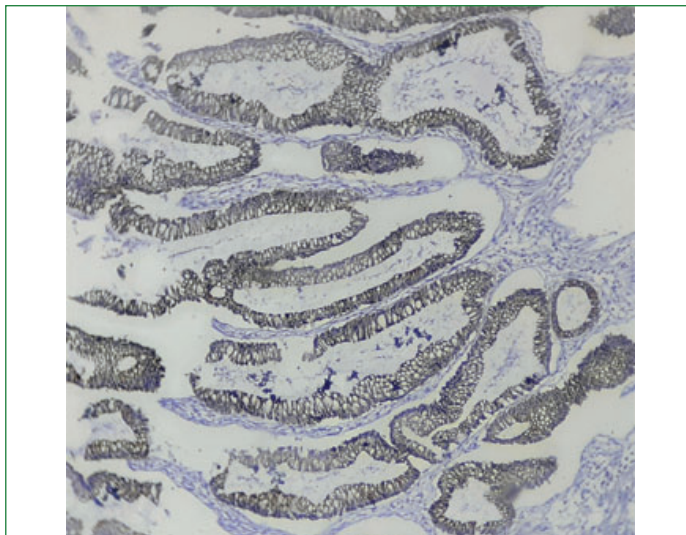
[Table/Fig-4]: Pleomorphic, hyperchromatic nuclei and intracellular mucin (H&E stain, 400X).



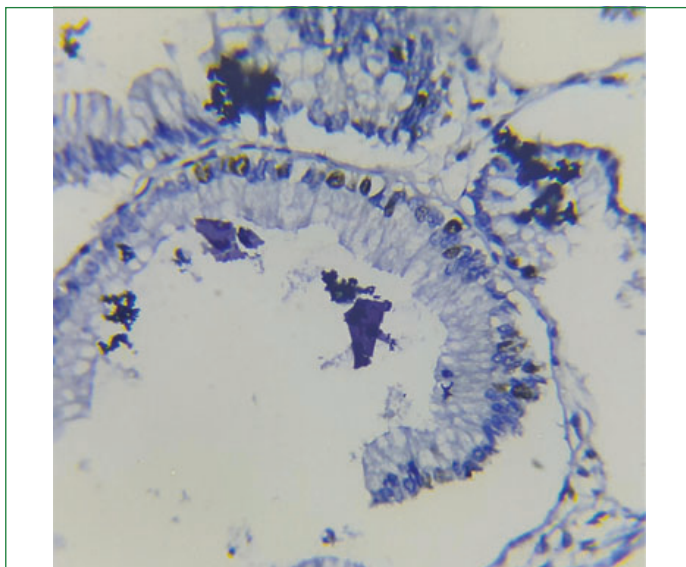
[Table/Fig-5]: Photomicrograph showing Ck7 positivity (400X).



[Table/Fig-6]: Photomicrograph showing Ck20 positivity (400X).



[Table/Fig-7]: Photomicrograph showing CEA positivity (100X).



[Table/Fig-8]: Photomicrograph showing Ki67 positivity (400X).

Postoperatively, the patient's CEA level slowly decreased to the normal range (4.8 ng/mL), and he was free from disease recurrence at the time of writing this case report.

DISCUSSION

In 1920, Hasebe M et al., reported the first case of primary mucinous adenocarcinoma of the renal pelvis [1]. Adenocarcinoma constitutes only 1% of renal pelvis malignancies and can be classified into three subtypes: tubulovillous, mucinous, and papillary non intestinal [2]. Tubulovillous and mucinous carcinomas, which account for 93% of cases, are representative of intestinal adenocarcinomas [3]. Primary mucinous adenocarcinomas are typically discovered incidentally after nephrectomy [4]. Until 2020, approximately 100 cases of primary mucinous adenocarcinomas have been reported [5]. The pathogenesis of this condition remains unclear, with two leading theories proposed. One theory suggests that the mucosal epithelium of the renal pelvis undergoes glandular metaplasia as a result of chronic irritation, such as urolithiasis, infections, and hydronephrosis, leading to abnormal hyperplasia and adenocarcinoma [6,7]. The other theory proposes that it can arise from sequestered renal epithelium due to maldevelopment, particularly in anomalous kidneys [2]. However, Takehara K et al., demonstrated that adenocarcinoma of the renal pelvis can originate from pre-existing transitional cell carcinoma [8].

Most cases of primary mucinous adenocarcinoma present after the age of 40, with a higher incidence in males. Smoking is a known risk

factor [9]. The tumour can be associated with ectopic kidneys [10] and horseshoe kidneys [11]. Common presenting features include flank pain, abdominal swelling, fever, and haematuria [12,13]. Radiological investigations reveal hydronephrosis and cortical thinning, and calculi may be present in the renal pelvis or ureter [5]. Gross examination of the kidney shows a complex heterogeneous mass with both solid and cystic components, along with gelatinous material [14]. Han B et al., reported a case with emboli in the left renal pelvic vein [15]. This condition is often misdiagnosed as calculus pyonephrosis [5,12]. Li H et al., reported a case that showed CDX2, CEA, Villin, and Ki-67 positivity on IHC examination [5].

On histopathological examination in present case, no areas showed irregular nests of urothelial cells with rounded or jagged contours. The tumour cells were negative for GATA3, indicating that this was not a case of transitional cell carcinoma or its variants. Tubular necrosis, interstitial neutrophils, patchy suppurative inflammation, and tubular thyroidisation were not present. These findings eliminated the diagnosis of pyelonephritis. The tumour was negative for PAX8, Oestrogen Receptor (ER) and Progesterone Receptor (PR), ruling out the possibility of endometrial or ovarian metastasis. The tumour cells were negative for CDX2, differentiating it from colonic adenocarcinoma metastasis. The tumour cells exhibited significant atypia and invasion into the renal parenchyma, indicating that this was not a case of intestinal metaplasia.

CONCLUSION(S)

Primary mucinous adenocarcinoma is a rare neoplasm of the renal pelvis. It lacks characteristic clinical and radiological features, and its rarity poses a significant challenge in establishing a standard treatment approach. Meticulous history-taking, radiological examinations, and additional assessments using tumour markers and preoperative cytological studies can improve the diagnostic accuracy of this condition.

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