**Case Report**

# Primary mucinous adenocarcinoma of renal pelvis: A case report

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**Summary**

Primary mucinous adenocarcinoma of the renal pelvis is a very rare tumour which is usually associated with urolithiasis and hydronephrotic changes due to chronic irritation and infection, leading to glandular metaplasia of lining urothelial cells. We present a 62- year old male with a history of ureteric calculi 7 years back now presenting with pain in the left flank and a palpable mass at left loin who was diagnosed clinically and radiologically to have hydronephrosis with a non functioning kidney. He underwent radical nephrectomy. On histopathological examination he was diagnosed to have mucinous adenocarcinoma of the renal pelvis stage PT4N1M0. Immunohistochemically the tumour cells were positive for CK7 and negative for CK20 and focally positive for CDX2.

Key words: Mucinous adenocarcinoma, metaplasia, hydronephrosis

## Introduction

Primary mucinous adenocarcinoma arising from the renal pelvis is very rare and account for less than 1% of renal pelvic neoplasms. The first case was reported by Ackerman in 1946[1]. Adenocarcinomas of the renal pelvis are usually subdivided into tubulovillous, mucinous, and papillary non-intestinal categories. Tubulovillous and mucinous group represent intestinal adenocarcinomas constituting 93% of the cases[2]. Mucinous adenocarcinomas are presumed to originate from the transitional epithelium that has undergone intestinal metaplasia[3]. We report a case of mucinous adenocarcinoma of the renal pelvis associated with hydronephrotic changes. This is the first case we have encountered at our institute.

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## Case Report

A 62-year-old man presented with a dull aching pain in the left flank for one year which had gradually increased in intensity in the last three

months. He had a past history of removal of a left sided ureteric stone seven years ago. He also complained of loss of weight and occasional fever. On general examination he was anaemic. The abdomen revealed a firm fixed mass in the left loin, extending to the left hypochondrium. On abdominal ultrasonography an enlarged left kidney with gross hydronephrosis and parenchymal thinning was found (Figure 1). Abdominal CT scan revealed gross hydronephrotic changes of left kidney due to pelvi-ureteric junction obstruction (Figure 2). DTPA (diethylenetriaminepentacetate) renogram showed gross parenchymal insufficiency of the left kidney with a split function of 8.417%. With a provisional diagnosis of hydronephrosis and a non functioning kidney exploration with or without nephrectomy was planned. During surgery the kidney was seen as a loculated cystic cavity filled with anchovy sauce coloured fluid with thinned out cortex. There was adhesion of the left kidney to the ureter, psoas muscle and aorta. Patient underwent left radical nephrectomy.

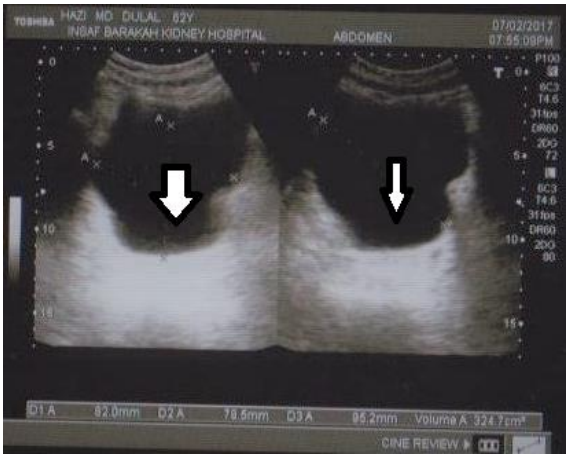


**Figure 1**: Ultrasonography of left kidney(arrow) reported as hydronephrosis

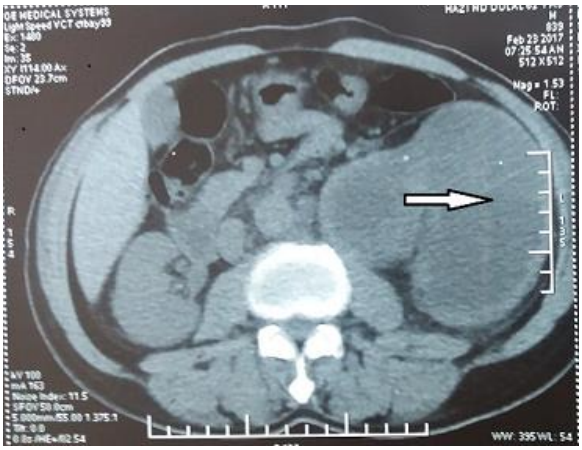


**Figure 2**: CT scan of abdomen shows gross hydronephrotic change in left kidney (arrow) with pelvi-ureteric junction obstruction.

On gross examination the kidney was found enlarged, cystic, bosselated and measured 14.5x9.5x9.0 cm. The capsule was intact and adherent. Cut section revealed slightly reddish copious mucinous material coming out from a widely dilated pelvis. The pelvi-ureteric junction revealed a velvety surface and a few ulcerated and papillary structures. The cortex was markedly thinned out. Multiple tiny grayish white nodules were seen in the thinned out pelvic wall. Perinephric fat showed a single lymph node. (Figure 3)

**Figure 3**: Cut section of left kidney showing dilated pelvicalyceal system.

Microscopically the renal pelvis was lined partly by transitional epithelium with transition to a tall columnar epithelium resembling gastric mucosa with delicate papillary folds and ulceration (Figure 4). The ulcerated area showed extensive mucin pools with numerous poorly differentiated signet ring cells infiltrating the wall of the renal pelvis extending into the pelviureteric junction (Figure 5). Foci of pleomorphic tumour cells with hyperchromatic nuclei in attempted gland formation were seen. The tumour was also seen infiltrating into the surrounding renal parenchyma and perinephric fat with preserved glomeruli as well as tubules (Figure 6). The isolated lymph node showed tumour deposits. The tumour cells as well as mucin pools were positive for Periodic acid Schiff (PAS) staining (Figure 7)

Immunohistochemistry revealed the tumour cells to be CK7 positive, CK20 negative and CDX2 focally positive (Figure 8). The histopathological diagnosis was mucinous adenocarcinoma of renal pelvis, stage PT4N1Mx.

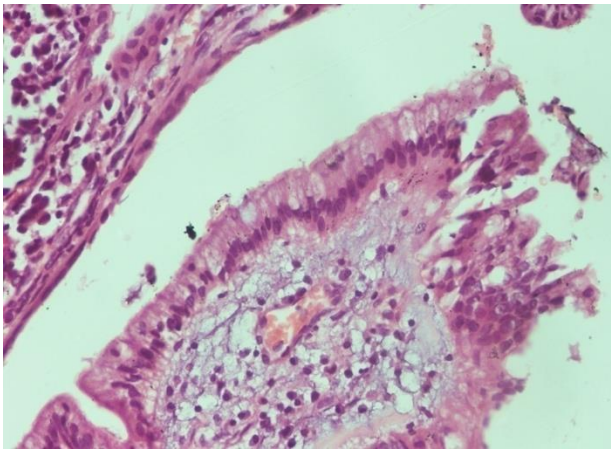

**Figure 4:** mucinous metaplasia of urothelial lining. (H&E x40)

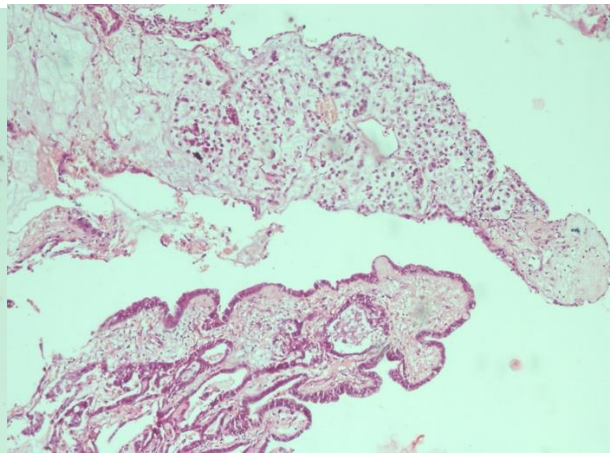

**Figure 5**: Neoplastic papillary structures along with poorly differentiated mucinous

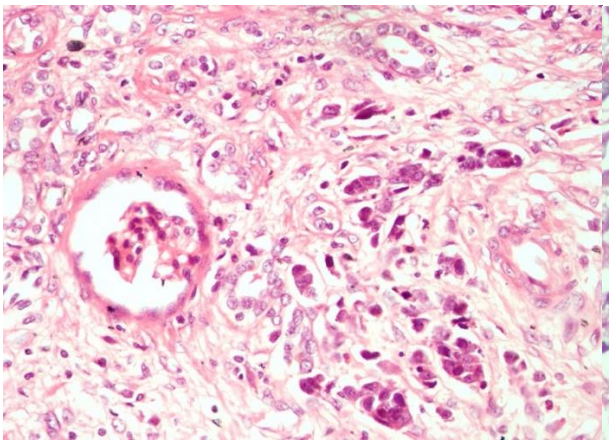

**Figure 6**: The tumour cells infiltrating the renal parenchyma (H&E x40)

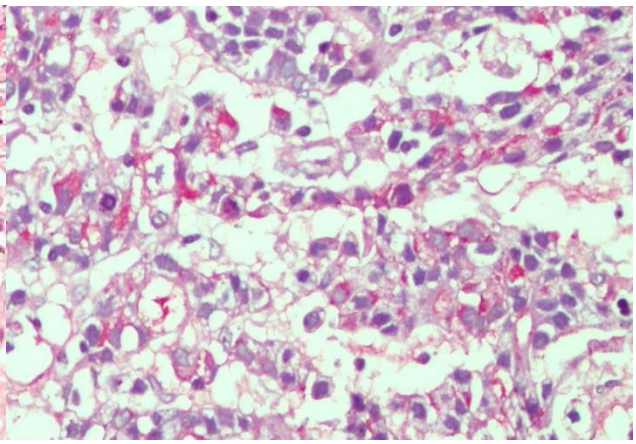

**Figure 7**: The tumour cells are positive for PAS staining. (PAS x40)

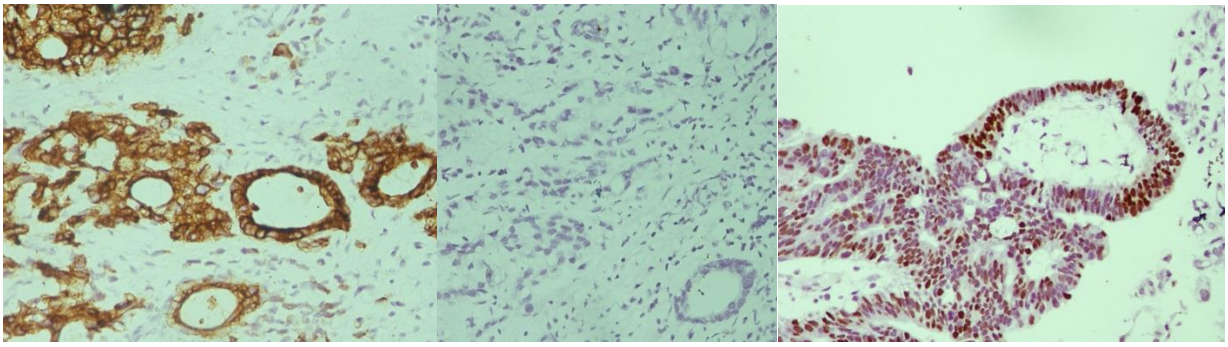

Figure 8: Immunohistochemistry shows CK-7 positive, CK-20 negative and CDX2 focally positive tumour cells.

## Discussion

The most frequently observed tumour of urothelial origin is urothelial carcinoma which accounts for 90% of all renal pelvic and ureteric tumours, which is followed by squamous cell carcinoma, adenocarcinoma and leiomyosarcoma[2]. Mucinous adenocarcinoma of the renal pelvis is a rare condition, and they are often reported as isolated cases. Only a small number of individual cases are reported in the English medical literature which accounts for about 100 cases to date[4]. This is the first reported case of a mucinous adenocarcinoma of renal pelvis from Bangladesh.

These tumours are often associated with chronic inflammation, hydronephrosis and urinary renal calculi[5]. The suggested pathogenesis considers that chronic irritation, inflammation and infection may lead to glandular metaplasia of the urothelium which progress to dysplasia and adenocarcinoma[6]. Aufderheide and Streitz reviewed 28 cases of mucinous adenocarcinoma of renal pelvis in 1974. They found that majority of the cases occurred in patients older than middle age and were usually associated with a long history of infection, stones, or hydronephrosis[7]. Our patient was 62-years of age and also had a history of ureteric calculus removal seven years back and he presented with gross hydronephrotic change of left kidney detected radiologically.

A pre-operative diagnosis is rarely made for this type of tumour, and most cases are diagnosed from resected specimens by a pathologist. The symptoms of these patients are often nonspecific, and haematuria is the most common sign in the majority of cases. In addition, flank pain and/or a palpable abdominal mass may also be clinical symptoms in late-stage patients[8]. Our patient presented with flank pain followed by palpable mass in the loin and history of gradual weight loss.

A histopathological diagnosis of mucinous adenocarcinoma requires some strict criteria to differentiate it from mere mucinous metaplasia of urothelial lining due to chronic irritation, (1) histological evidence of architectural or cellular atypia; (2) microscopic evidence of invasion of the renal pelvic wall and renal parenchyma, or of nodal or distant metastasis; and (3) evidence of overt invasion or recurrence, or of nodal or distant metastasis[7]. In our case criteria for malignancy were histologically obvious diffuse infiltration of tumour cells including many signet ring cells in the renal parenchyma,

perirenal fat tissue as well as tumour deposits in isolated lymph node. The signet ring cells were containing PAS positive mucin with in the cytoplasm. On immunohistochemistry the tumour cells were CK7 positive but CK 20 negative and CDX2 focally positive in our patient which is dissimilar to immunohistochemistry characteristics of mucinous carcinoma arising from large gut[9]. More over in our case there is an area of transition from urothelial lining epithelium of renal pelvis to metaplastic glandular lining close to the invasive component of mucinous adenocarcinoma which is a proof of primary origin of the tumour from renal pelvis.

Local recurrence due to spillage of tumour cells during surgical manipulation and downward seeding in the distal ureter have been reported. Hence radical nephrectomy and complete removal of ureter followed by chemotherapy is the preferred management plan for this type of tumour. Furthermore, these tumours carry a poor prognosis with about 50% mortality within 2 years after surgical removal[10]. In our case the radical nephrectomy was performed for hydronephrosis with non functioning kidney. As the surgeons were unaware about the actual pathology, ureterectomy was not done. The case was diagnosed histopathologically as mucinous adenocarcinoma of renal pelvis stage pT4N1Mx. During postsurgical chemotherapy the patient died 3 months after the diagnosis.

## Conclusion

This is the first reported case in Bangladesh of mucinous adenocarcinoma of renal pelvis associated with hydronephrosis and history of urolithiasis, which was diagnosed incidentally during histopathological examination. Thus, awareness of the urologists of such entity with knowledge of the nature of the disease at the time of surgery may help him to take a decision to plan for adequate extent of surgical removal of the tumour.

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