

## Primary mucinous adenocarcinoma of the renal pelvis and ureter

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### ABSTRACT

Mucinous adenocarcinomas of the renal pelvis and ureter are among the rarest upper urinary tract neoplasm. We report a case of multifocal primary mucinous adenocarcinoma of the renal pelvis and ureter occurring in association with a staghorn calculus and pyonephrosis. A 68 year old man had suffered from right flank pain and upper abdominal swelling for one year. After a series of investigation, a right staghorn stone with pyonephrosis leading to non-functioning kidney was found. Right nephrectomy was performed. The pathological report showed mucinous adenocarcinoma with ureteric margin positive for tumour deposits. Patient was re-operated; right ureterectomy with removal of bladder cuff was done. Although uncommon, the possibility of a tumor should be kept in mind especially in patients with a long standing urolithiasis accompanied by hydronephrosis and/or infection.

**Keywords:** mucinous adenocarcinoma, staghorn stone, renal pelvic tumor.

Most malignant renal pelvic tumors are of epithelial origin. Among them, about 90% are transitional cell carcinomas.<sup>1</sup> Primary mucinous adenocarcinoma of the renal pelvis is rare. We report a case of multifocal primary mucinous adenocarcinoma of the renal pelvis and ureter occurring in association with a staghorn calculus and pyonephrosis.

### CASE REPORT

A 68 year old nonsmoker male presented with pain and swelling in right upper side of abdomen for one year along with on and off mild fever. General physical examination revealed pallor. Abdominal examination revealed 20 x 15 cm smooth, firm, non-tender, ballotable lump in right lumbar area extending to the right hypochondrium. Routine hematological investigations showed hemoglobin of 6.7 gm %. Urine examination revealed packed pus cells with no red blood cells and the culture was sterile. Ultrasonography (USG) of abdomen showed right pyonephrosis with staghorn calculi. Intravenous urography (IVU) showed a right staghorn calculus with non visualized right kidney and normal functioning left kidney. Tc99m-Diethylenetriaminepentaacetic acid (DTPA) renal scintigraphy showed right non functioning kidney.

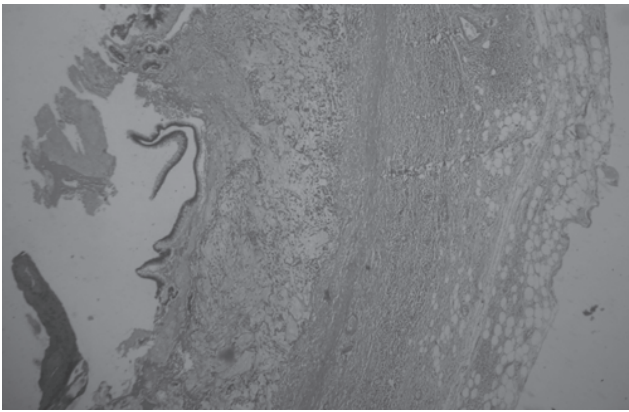
With the diagnosis of right nephrolithiasis with pyonephrosis leading to non functioning kidney, right nephrectomy was performed. Intraoperatively, kidney was markedly enlarged with perinephric adhesion and contained about 1500 ml of pus with staghorn calculus in the renal pelvis extending into lower calyx. On gross pathological examination, kidney measured 20 x 15 x 10 cm with an attached ureter of 9 x 2.5 cm in length. Cut section showed dilated calyx with loss of corticomedullary junction and a friable myxoid mass measuring 2 x 1.5 cm present near pelviureteral junction. Microscopic examination from pelviureteric mass showed a picture of mucinous adenocarcinoma

characterized by intestinal metaplasia with dysplasia, pools of extracellular mucin with numerous floating signet ring cells infiltrating lamina propria and muscularis layer (Fig. 1, 2). Ureteric resected margin was positive for tumour deposit. Lymphovascular invasion and perinephric fat invasion was not seen. Kidney showed features of xanthogranulomatous pyelonephritis. TNM stage of mucinous adenocarcinoma was T<sub>2</sub> N<sub>0</sub> M<sub>x</sub>.

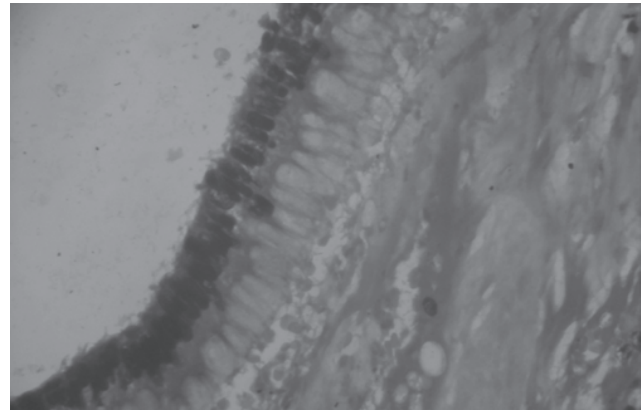
As ureteric margin was positive for tumour cells, contrast enhanced computed tomography (CECT) abdomen and pelvis was done which didn't show any obvious mass or lymph nodes. Cystoscopy was normal. Patient was re-operated and right ureterectomy with removal of bladder cuff was done. Histopathology report showed residual mucinous adenocarcinoma in segment of proximal ureter and marginal tissue from ureter and bladder cuff were free of tumour deposits. The postoperative course was uneventful. There is no evidence of recurrence in the 18 months of follow up with USG abdomen and cystoscopy with urine cytology.

### DISCUSSION

Malignancies originating within the renal pelvis and ureter are uncommon. Most of them are epithelial in origin and can be divided histologically into transitional cell carcinoma (90%), squamous cell carcinoma (10%) and adenocarcinoma (1%).<sup>1</sup> Primary adenocarcinoma are rare and subdivided into tubulovillous (71.5%), mucinous (21.5%), and papillary non-intestinal type (7%).<sup>2</sup> Reports of renal pelvic mucinous adenocarcinomas in the literature are limited to small series and isolated case reports. In 1946, Ackerman reported the first case<sup>3</sup> and till date, approximately 100 cases have been reported in the english medical literature.<sup>4</sup> The highest numbers of cases are reported from Japan (12 case) and India (10 case) while other parts of the world have reported occasional cases.<sup>5</sup> As far as our best knowledge, this is the first reported



**Fig. 1.** Intestinal columnar (tumour) cells at pelvoureteric junction (H&E, 40×)



**Fig. 2.** High-power microscopic examination of the myxoid mass near pelvoureteric junction revealing tall columnar epithelium (cytoplasm filled with mucin and nuclei pushed to one side) with pool of extracellular mucin. (H&E, 400×)

case from Nepal.

Most of the tumors are solitary either in renal pelvis or in the ureter. But Lauritzen *et al* observed multifocal tumors of both renal pelvis & ureter, in 4 out of 37 cases of their series.<sup>6</sup> We also have the same experience of multifocality in our case.

The pathogenesis of this tumor is not clear. The postulated pathogenesis is related to its frequent association with chronic irritation, inflammation, infection, hydronephrosis and urinary calculi. Glandular metaplasia of the urothelium that develops as a response to injury, may progress to dysplasia and adenocarcinoma.<sup>8</sup> Our patient had staghorn calculus leading to pyonephrosis, predisposing him to all the above conditions.

Most reported cases were identified in patients older than 60 years and there is no evidence of any sex difference. Patients are often asymptomatic. Hematuria is the most common presenting symptom while loin pain and palpable abdominal mass signifies a late stage. Over two thirds of cases are associated with urolithiasis and hydronephrosis.<sup>2</sup> However, there was no hematuria in our patient.

The radiological investigations to be done are USG, IVU, retrograde urography and CECT. CECT abdomen and pelvis scan is the investigation of choice and demonstrates the tumour with a total evaluation, e.g., appearance, extent, local spread and its effect on renal function.<sup>7</sup> There is no specific feature that differentiates mucous adenocarcinoma from transitional cell carcinoma (TCC) by clinical symptoms or imaging modalities.<sup>4</sup> The final diagnosis is confirmed by post-operative biopsy.

Lesions of mucinous adenocarcinoma are usually incidentally detected from surgically resected specimens. In a series of five cases by Chen *et al*, nearly all of their cases were undetected preoperatively, and these patients were thought to have some benign infections or inflammatory conditions. Subsequently an additional procedure of ureterectomy was performed in many cases. It is important to recognize this tumor intraoperatively as one of the predominant features intraoperatively is copious, thick, viscid mucous material (extracellular mucin) in the renal pelvis and calyces.<sup>4</sup> In our case, kidney was full of pus, hence we did not suspect

malignancy initially. We also had incidental diagnosis from surgically resected specimen by pathologist and as ureteric margins was positive for tumour cells, so the patient underwent additional procedure of ureterectomy with bladder cuff excision.

Radical nephrectomy and total ureterectomy including the ipsilateral bladder cuff excision is the treatment of choice.<sup>7</sup> Till date, no adjuvant radiotherapy and/or chemotherapy has been proven to be effective for mucinous adenocarcinoma of the renal pelvis.<sup>4</sup>

The overall prognosis of adenocarcinoma appears to be poor. One and five years survival rates are 75.7% and 21.1% respectively, about half of the patients die within 2 years of surgery.<sup>8</sup> But out of three subtypes of adenocarcinoma, mucinous adenocarcinoma have a better prognosis.<sup>2</sup>

Although primary mucinous adenocarcinoma is a very uncommon neoplasm, the possibility of a tumor should be kept especially in an elderly patient with long standing urolithiasis accompanied by hydronephrosis and/or infection.

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