

Mucinous adenocarcinoma of the renal pelvis with adenocarcinoma *in situ* of the ureter

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Abstract

Primary epithelial tumor of the renal pelvis is rare, and adenocarcinomas account for less than 1% of malignancies arising from the epithelium of the renal pelvis. We describe in this study a case of a 56-year old male patient who presented with an abdominal mass and dull aching pain for one year. A diagnosis of hydronephrosis of the right kidney was made based on imaging studies. Grossly, the entire kidney was converted into a cystic mass measuring 16 × 12 × 10 cm and filled with gelatinous material along with staghorn calculi in the pelvis measuring 7 × 4 × 3 cm. Histological examination of the tumor showed glands, cysts, and papillae lined by pseudostratified columnar epithelium with hyperchromatic nuclei. Scattered signet ring-type cells were also seen floating in large pools of extracellular mucin. Sections from the ureter showed a component of adenocarcinoma *in situ*. No invasive tumor was identified in ureteric tissue. Thus, a diagnosis of mucinous adenocarcinoma of the renal pelvis with *in situ* adenocarcinoma of the ureter was made.

Key Words: Adenocarcinoma, adenocarcinoma *in situ*, mucinous adenocarcinoma, renal pelvis

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INTRODUCTION

The commonest malignant tumor of the renal pelvis is transitional cell carcinoma, accounting for 7% of the primary renal tumors followed by squamous cell carcinoma. Adenocarcinomas of the renal pelvis are rare. They are usually subdivided into tubulovillous, mucinous, and papillary nonintestinal categories. Tubulovillous and mucinous group represent intestinal adenocarcinomas constituting 93% of the cases.^[1,2] Mucinous adenocarcinomas are presumed to be originating from the intestinal metaplasia of

the transitional epithelium.^[2-4] There are no documented cases showing adenocarcinoma *in situ* in the ureter or renal pelvis. We report a case of mucinous adenocarcinoma of renal pelvis with *in situ* adenocarcinoma of the ureter.

CASE REPORT

A 56-year-old male patient presented with swelling in the right side of the abdomen for a period of one year which was gradually increasing in size. He complained of continuous dull aching pain. There was no history of referred pain, dysuria, or hematuria. Ultrasonography of the abdomen revealed a right renal mass in favor of hydronephrosis. Intravenous urography showed a right nonvisualized kidney with a staghorn stone and multiple secondary calculi. Computed tomography scan showed gross hydronephrosis of the right kidney, two calculi and the ureter was dilated up to the level of iliac vessels. With the provisional diagnosis of hydronephrosis secondary to urolithiasis, the patient was subjected to right nephrectomy. Peroperatively, the right kidney appeared like a cystic mass, from which 4 l of mucoid material was drained.

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Staghorn calculi measuring $7 \times 4 \times 3$ cm was found in the pelvis. Multiple calculi were also removed along with the mucoid substance.

Grossly, the kidney was markedly enlarged and measured $16 \times 12 \times 10$ cm. The capsule could easily be peeled out. The entire kidney was replaced by a cystic mass consisting of multiple cysts filled with mucoid material and gelatinous solid areas. Papillary excrescences were seen in some of the cysts. The cortex was markedly thinned out [Figure 1]. The ureter measured 1.5 cm in length. The lumen appeared dilated and filled with mucus. However, no growth or papillary excrescences were seen in the entire length of the ureter.

Histological examination of the tumor showed 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 [Figure 2]. The ureter was dilated and the mucosa was lined by pseudostratified columnar epithelium with apical mucin and hyperchromatic nucleus and intramucosal atypical gland formation, as described in the *in situ* adenocarcinoma of the colon or a borderline mucinous tumor of the ovary [Figure 3]. It was seen extending till the resected margin of the ureter. However, there was no infiltration into the wall of the ureter.

Carcinoembryonic antigen (CEA) level done postoperatively was within a normal range. Patient was given radiotherapy postoperatively. After a period of one year, patient presented with back pain radiating to the legs for duration of one month. Magnetic resonance imaging of the spine revealed multiple cervical and lumbar vertebral lesions, with recurrence of growth in the right renal fossa and enlarged retroperitoneal lymph nodes. There was cord compression at the level of 10th thoracic vertebra.

DISCUSSION

Mucinous adenocarcinoma of the renal pelvis are rare tumors and are mainly reported from Asian countries.^[4-9] There are few documented reports according to which the mucinous adenocarcinoma of the renal pelvis usually occurs following glandular metaplasia of the transitional epithelium induced by long-standing chronic inflammation, sometimes secondary to renal stones.^[2-4] Though it has been presumed that an adenoma-carcinoma sequence like that of a colonic tumor exists, no such study has been done to establish such association. Our case was associated with borderline type of mucinous tumor in the ureter and had renal stones suggesting a possible intestinal metaplasia of the transitional epithelium leading to mucinous adenocarcinoma. Ross and D'Amato reported a case of papillary mucinous cystadenoma



Figure 1: Photograph of the renal mass shows multiple cysts filled with mucoid material

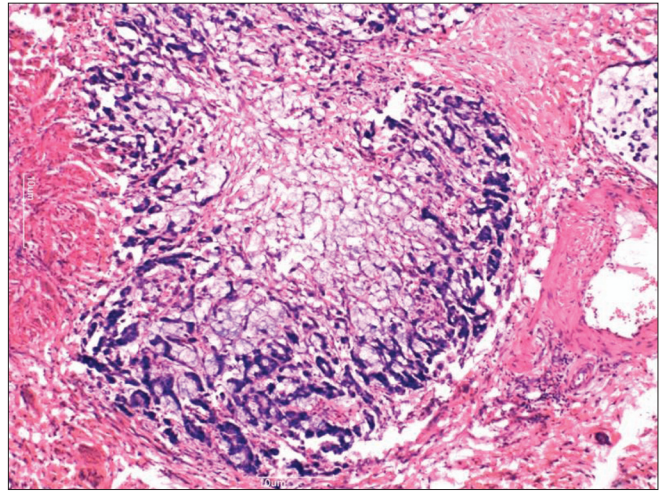


Figure 2: Photomicrograph shows malignant glands and signet ring cells in large pools of extracellular mucin (Hematoxylin and Eosin, $\times 10$)

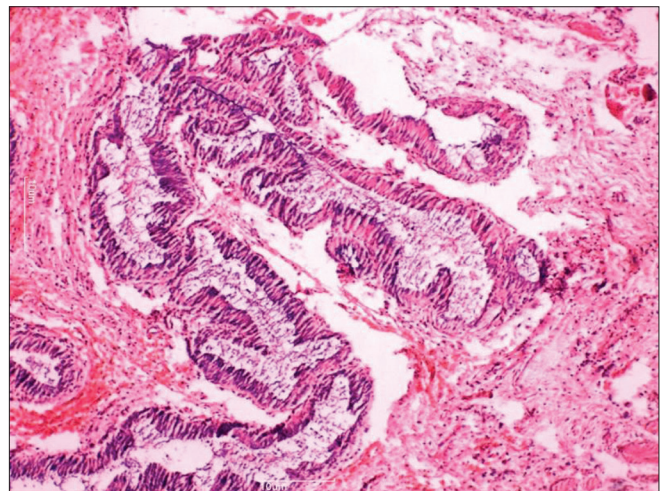


Figure 3: Photomicrograph of ureter shows mucosa lined by pseudostratified columnar epithelium with hyperchromatic nuclei and papillary tufting (Hematoxylin and Eosin, $\times 10$)

of probable renal pelvic origin in a horseshoe kidney. They suggested that the same criteria used for evaluating mucinous ovarian tumors might also be appropriate for mucinous tumors of the renal pelvis.^[10] Cases of mucinous cystadenoma arising from the renal pelvis has also been reported from India.^[7] However, there has been no study to check evolutions of these tumors. There are no reports of adenocarcinoma *in situ* as occurred in this case.

Grossly, abundant mucin is seen in most cases, as was evident in our case.^[8,9] There has been a report of pseudomyxoma peritonei associated with mucinous adenocarcinoma.^[8] Some tumors have shown high level of CEA like in colonic mucin-secreting adenocarcinomas and it has been suggested to be used as a marker for prognosis and recurrence.^[11] However, serum CEA level was normal postoperatively in our case and did not show any raise during the follow-up period.

The treatment of these tumors is radical nephrectomy and total ureterectomy, including the intravesical part.^[12] However, as there was no preoperative diagnosis, total ureterectomy was not done in our case. A review of the radiological images did not reveal any significant findings in favor of the diagnosis in addition to the presence of hydronephrosis and calculi. In view of the presence of cysts filled with large pools of mucin and gelatinous solid areas in most of the documented cases of mucin-secreting adenocarcinoma, a strong clinical suspicion is needed to establish the diagnosis. Peroperative frozen section study or cytology may help to confirm the diagnosis. Our case presented with recurrence within a period of one year; however, there are reports of good prognosis without recurrence even after three years after surgery.^[5]

CONCLUSION

Adenocarcinoma of the renal pelvis is closely associated with intestinal metaplasia of transitional epithelium induced by long-standing chronic inflammation, renal calculi, and persistent hydronephrosis. They are high-grade tumors. However, the

presence of *in situ* type of adenocarcinoma in the ureter associated with mucinous tumor has not been documented. In the absence of significant diagnostic radiological features in favor of this diagnosis, peroperative diagnosis with frozen section or cytology may be helpful to confirm the diagnosis and planning of an appropriate surgery. There is a need to study the etiopathogenesis and evolution of these tumors in the Indian subcontinent, wherein the highest number of cases have been recorded.

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