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Case report

Primary mucinous adenocarcinoma of the renal pelvis with carcinoma in situ in the ureter

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KEYWORDS

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Abstract Primary epithelial tumor of the renal pelvis is rare and only 100 cases are reported in the literature [1]. 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. One case was reported with carcinoma in situ of the ureter (2).

Immunohistochemically: The tumor showed positivity for CK7, CK20, CK8/18, GATA-3, MSH-2, MSH-6, MLH-1, Ber-EP4, and S-100-P with focal positivity for CDX-2, weak positivity for PMS-2 and negativity in TTF-1 and Her-2. Molecular pathological analysis revealed microsatellite stability and without mutation in K-ras-gene. Thus, a diagnosis of mucinous adenocarcinoma of the renal pelvis with in situ adenocarcinoma of the ureter was made.

Introduction

Mucinous adenocarcinoma of the renal pelvis was first described in 1960 by Hasebe et al. [3], and it was the least common of the renal pelvis tumors, which include transitional cell carcinoma (85–90%), squamous cell carcinoma (10–15%) and adenocarcinoma (< 1%). Adenocarcinomas are further subdivided into (a) tubulovillous, (b) mucinous, and (c) papillary non-intestinal categories. The first two groups resemble intestinal adenocarcinoma and constitute 93% of the cases [4]. Mucinous adenocarcinomas are presumed to be

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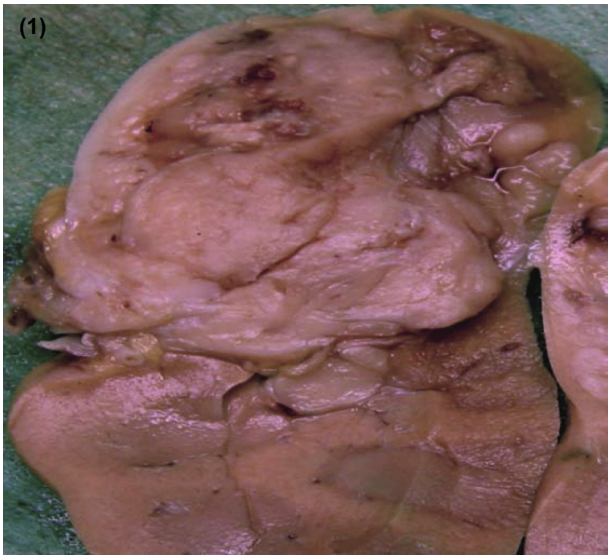

Figure 1 Photograph of the renal mass in renal pelvis with infiltration into the renal tissue.

originating from the intestinal metaplasia of the transitional epithelium [5].

Mucinous adenocarcinoma of the renal pelvis is a rare tumor and is mainly reported from Asian countries [6]. 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 [6]. It has been presumed that an adenoma-carcinoma sequence like that of a colonic tumor already exists. We report a case of mucinous adenocarcinoma of renal pelvis with in situ adenocarcinoma of the ureter.

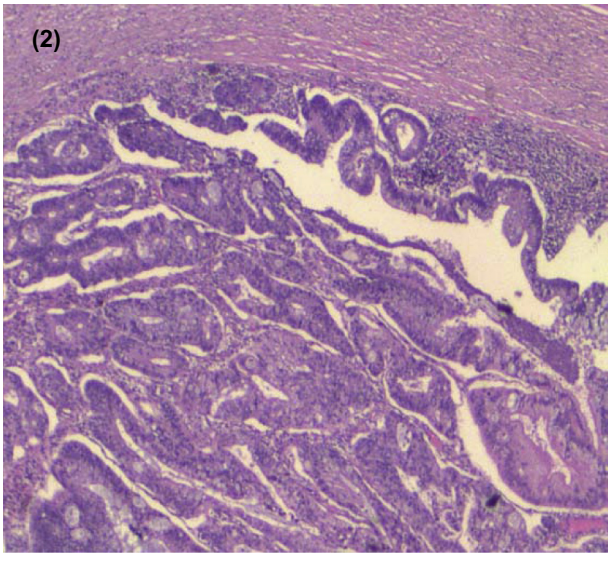

Figure 2 Photomicrograph shows malignant glands and signet ring cells in large pools of extracellular mucin (Hematoxylin and Eosin.x10).

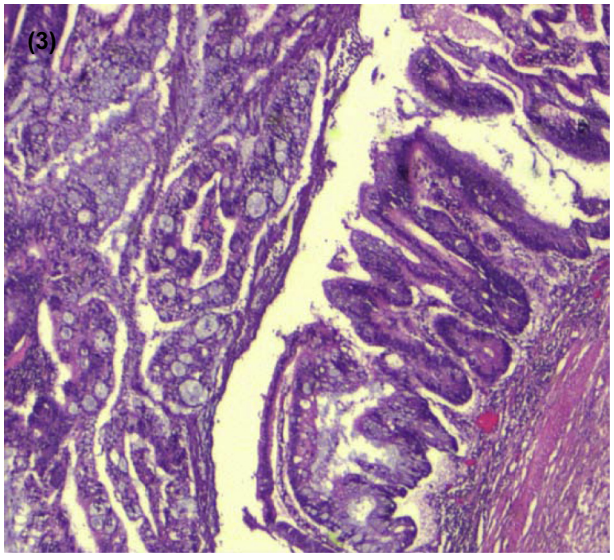

Figure 3 Photomicrograph shows on the right side the intestinal metaplasia of the renal pelvis with dysplastic changes. On the left side of the photomicrograph it shows malignant glands (Hematoxylin and Eosin.x10).

Case report

A 51-year-old female patient presented with swelling in the right side of the abdomen for a period of 1 year which was gradually increasing in size. She 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. Intravenous urography showed a right nonvisualized kidney with multiple stones. Computed tomography scan showed renal mass of the right kidney, two calculi and the ureter was dilated up to the level of iliac vessels. The provisional diagnosis was of non-functioning kidney and renal mass. The patient was subjected to right nephrectomy and partial ureterectomy. Preoperatively the kidney appeared like a cystic mass

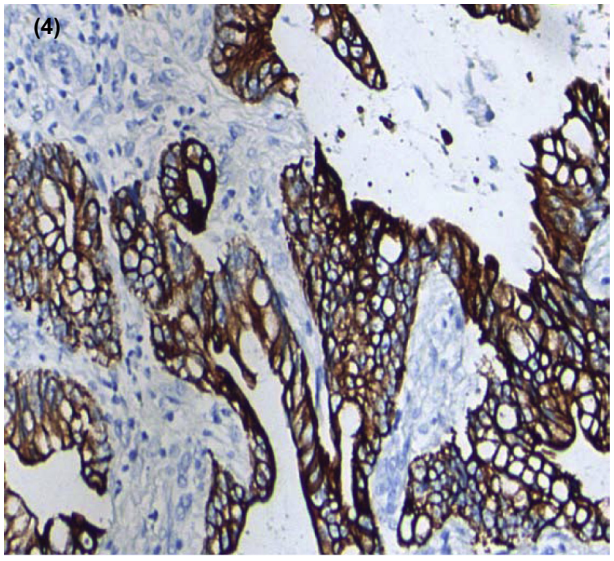

Figure 4 Neoplastic cells with strong positivity for CK8/18.

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with multiple stones, from which multiple calculi were also removed along with the mucoid substance.

Grossly, the kidney was enlarged and measured $16 \times 12 \times 10$ cm. The capsule could easily be peeled out. The entire kidney was replaced by a mass consisting of multiple cysts filled with mucoid material, gelatinous solid areas and small stones. Papillary excrescences were seen in some of the cysts. The cortex was thinned out (Fig. 1). The ureteric stump measured 1.5 cm in length. The lumen appeared dilated and filled with mucus. However, no growth or papillary excrescences were seen in 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 scattered poorly differentiated signet ring tumor cells were seen infiltrating into the renal cortex (Fig. 2). The ureter was dilated and the mucosa was lined by pseudostratified columnar epithelium with apical mucin and hyperchromatic nuclei and intramucosal atypical gland formation, as described in the in situ adenocarcinoma of the colon or a borderline mucinous tumor of the ovary (Fig. 3). It was seen extending till the resected margin of the ureter. However, there was no infiltration into the wall of the ureter.

Immunohistochemically: The tumor showed positivity for CK 8/18 (Fig. 4), CK7 (Fig. 5), CK20, GATA-3, MSH-2, MSH-6, MLH-1, Ber-EP4, and weak positivity for S-100-P (Fig. 6) and PMS-2 (Fig. 7) with negativity for Her-2. Focal positivity was detected for CDX-2 (Fig. 8).

Molecular pathological analysis revealed microsatellite stability which means that carcinoma of probable sporadic and not familial origin and without mutation in *K-ras-gene*. The TNM-classification was pT3, pN0 and after 6 months has developed metastasis in the chest wall (pM1).

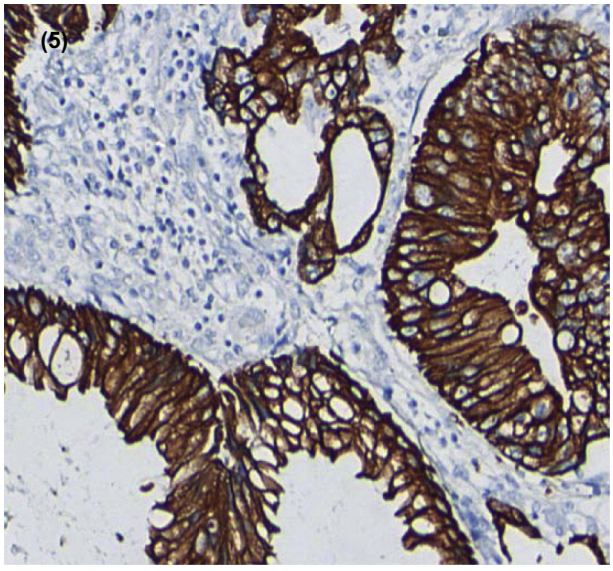

Figure 5 Neoplastic cells with strong positivity for CK7.

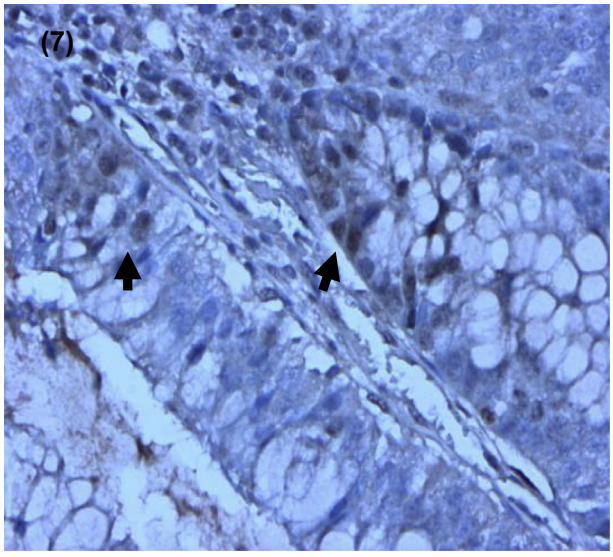

Figure 7 Focal weak positivity for PMS-2.

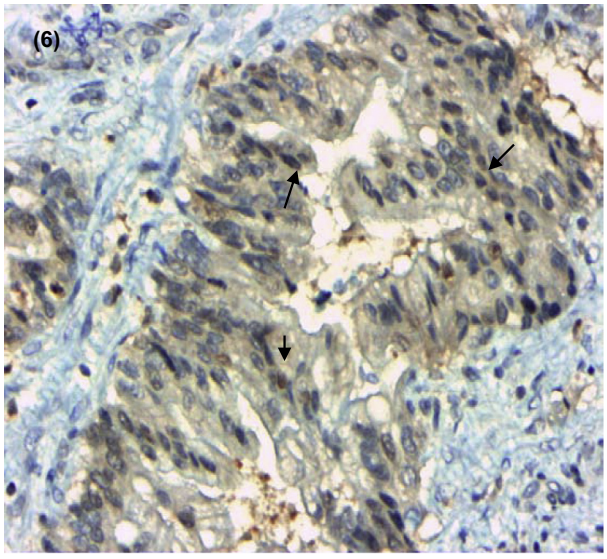

Figure 6 Focal nuclear positivity for S-100-P (arrow).

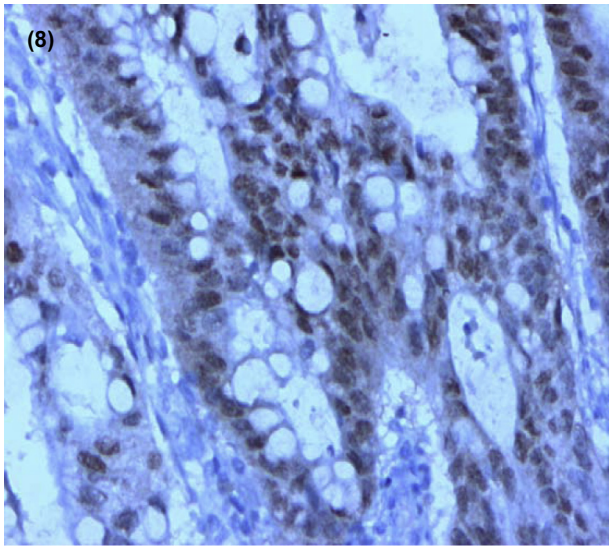

Figure 8 Nuclear positivity for CDX-2.

To exclude metastasis CT-scan was performed to exclude other abdominal or thoracic tumors. Endoscopic investigation was performed to exclude other tumors in the gastroduodenal tract. Complete gynecological analysis was performed to exclude associated tumors in genitourinary tract.

Discussion

Our case was associated with borderline type of mucinous tumor in the ureter and had renal pelvis 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 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 [7]. Cases of mucinous cystadenoma arising from the renal pelvis have also been reported from India [8]. The treatment of these tumors is radical nephrectomy and total ureterectomy, including the intravesical part [9]. However, as there were no preoperative measures to detect such carcinoma in situ in the ureter and as the standard operation for pelvis tumors is nephrectomy with partial ureterectomy, 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 of carcinoma in situ 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. Preoperative cytological diagnosis or intraoperative frozen section study may help to confirm the diagnosis of carcinoma in situ in the ureter and establish the diagnosis of adenocarcinoma of the renal pelvis. There was negativity in the immunohistochemistry for GATA-3 and also weak positivity for S-100-P. There are the only relatively specific markers for urothel. This is why, primary tumors of the gastrointestinal tract by endoscopy and primary tumors of the gynecological origin were excluded. Primary Tumors of the lung by X-ray and CT-examination were also excluded. Our case presented with recurrence in the form of distant metastasis within a period of 1 year; however, there are reports of good prognosis without recurrence even after 3 years after surgery [6].

However, there has been no study to check evolutions of these tumors. There was one case reported with adenocarcinoma in situ as occurred in this case [2].

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 indicated also by microsatellite stability [10]. The carcinoma in situ in the ureter would not be radiologically detected and this is why only partial ureterectomy with nephrectomy was performed. In the absence of significant diagnostic radiological features in favor of this diagnosis, intraoperative diagnosis with frozen section or cytology may be helpful to confirm the diagnosis and planning of an appropriate surgery, nephrectomy with complete ureterectomy which favors good prognosis and survival with little incidence of recurrence.

Conflict of interest

I confirm that there is no Conflict of interest.

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