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Case report

Primary enteric-type mucinous adenocarcinoma of the renal pelvis masquerading as cystic renal cell carcinoma: A case report and review of the literature

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ABSTRACT

Primary mucinous adenocarcinoma of the renal pelvis is easily misdiagnosed and it was rarely reported in the literature. We describe in this study a case of 40-year-old male patient presented with right lumbar pain of one year duration and elevated level of carcinoembryonic antigen (CEA). After a series of imaging examinations, the initial impression was a cystic renal cell carcinoma. Right radical nephrectomy was performed on the patient. The postoperative pathological examination indicated a primary mucinous adenocarcinoma of enteric type of the renal pelvis. After surgical removal of the tumor, an immunotherapy was administrated to prevent recurrence. The patient survives upon this report. A review of pertinent literature is also presented.

1. Introduction

Most tumors of the renal pelvis are urothelial in origin. Primary adenocarcinomas of the urinary system are very rare and those with enteric type, mucinous or signet-ring cell phenotypes have only rarely been reported in renal pelvis in a few dozens of literatures to date. Glandular metaplasia, nephrolithiasis and repeated inflammatory irritation are predisposing factors. In view of the relative rarity of this tumor and its deceptive imaging, we here report a case of incidental primary enteric-type mucinous adenocarcinoma of the renal pelvis with clinical symptoms and imaging characteristics mimicking a cystic renal cell carcinoma. Because the tumor is easily misdiagnosed and currently no standard treatment protocol has been proposed, we also present a summary of related literatures published to date.

2. Case report

A 40-year-old male presented with intermittent dull aching pain in the right lumbar region of one year duration. Before this hospital admission, he has been diagnosed as right renal cyst during routine examinations for three years. There was no history of dysuria, hematuria, or pyuria. On physical examination, no positive sign was appreciated. Routine hematological investigations were within normal limits. Some oncology markers were sampled before surgery, and carcino-embryonic antigen (CEA) showed a moderately elevated level at 31.4 ng/ml. Serum CA19-9 was at close to its normal upper limit (32.5 u/ml). Because the elevation of CEA usually indicated the primary origin site at the gastrointestinal tract, we ran an upper gastrointestinal endoscopy and a colonoscopy to exclude primary lesions from the GI tracts. No gastric or colonic mucosa was found to be abnormal.

Ultrasonography of the abdomen revealed a solid-cystic mass on the right kidney measuring 9.0cm × 6.8 cm. Nodular enhancement was appreciated at the cyst wall, and a malignancy of the mass was suspected (Fig. 1). The dynamic contrast-enhanced computed tomography (CT) scan revealed a grossly cystic mass of the right kidney with very thin wall and clear margin (Fig. 2). The CT attenuation (HU) of the tumor was 18 HU. The attenuation stayed almost the same during enhanced phases (19 HU). Because no enhancement was appreciated, pure kidney cyst with bleeding was the initial impression drew by the radiologists. The subsequent PET-CT showed slower FDG metabolism in the cystic mass of the right kidney. Malignant abnormality with low metabolism could not be excluded. With the provisional diagnosis of huge cystic renal cell carcinoma (CRCC), the patient was subjected to right radical nephrectomy. At operation the right kidney was found to be very large and the cortex appeared to be thin (Fig. 3A). On gross pathological examination, the kidney measured 15 × 10 × 5 cm. On cut

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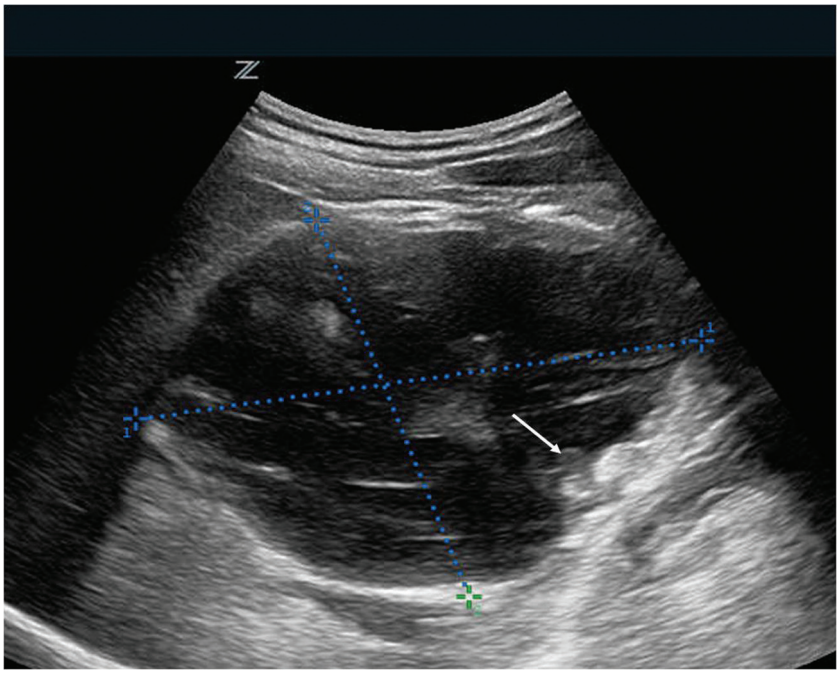

Fig. 1. On the ultrasonograpgy, nodular enhancement was appreciated at the cyst wall, which revealed malignancy of the mass.

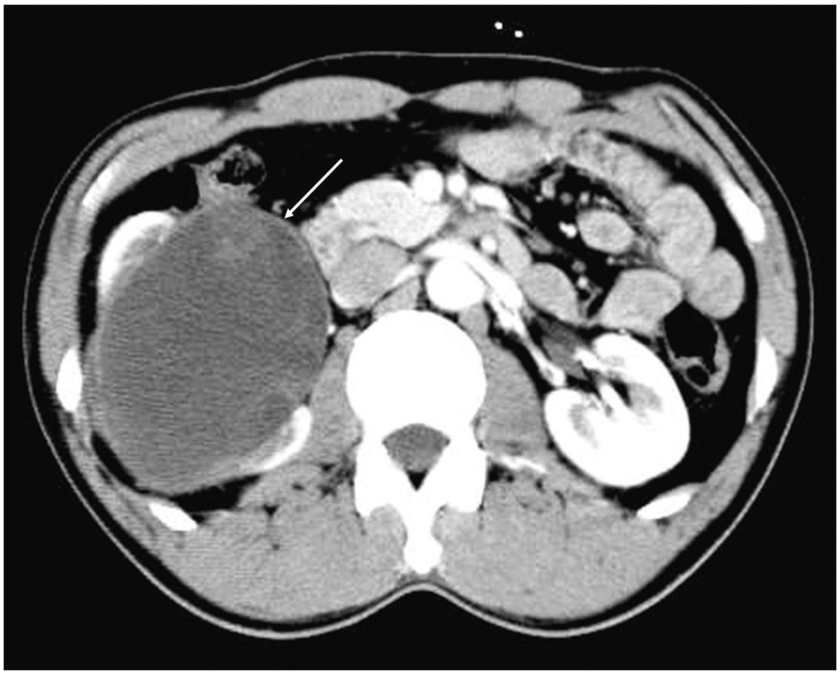

Fig. 2. A gross cystic mass of the right kidney with very thin wall and clear margin was shown on the CT scan and no significant contractions existed.

surface, jelly-like substance filling the whole renal pelvis and calyx was appreciated (Fig. 3B).

The final diagnosis of a primary enteric-type mucinous adenocarcinoma of the renal pelvis was established on pathological examination. Microscopic examinations of the tumor revealed a tubular adenocarcinoma of enteric type with abundant exosmotic mucinous depositions that predominantly localized in the renal pelvis and infiltrating into the renal parenchyma (Fig. 4A and B). Immunohistochemical studies revealed that the tumor cells were positive for cytokeratin 20 (CK20) (Fig. 5), CEA (Fig. 6), CDX2 (Fig. 7) and MUC-2 (Fig. 8). Renal interstitial infiltration of chronic inflammatory cells could be easily identified (Fig. 9). On the basis of the histomorphological and immunohistochemical features of the tumor and the clinical impression of a secondary adenocarcinoma of the renal pelvis from other organs was excluded, a diagnosis of mucinous adenocarcinoma of enteric type originally arising at the renal pelvis was finally made.

After surgery, the patient was given interleukin-2 (IL-2) and ubenimex empirically to prevent recurrence. At one month post-operation, the patient was asymptomatic. His CEA level was within the normal range by 1 month after discharge and remained at low