Case Report

Primary mucinous adenocarcinoma of the renal pelvis with signet ring cell formation

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ABSTRACT

Primary mucinous adenocarcinoma is exceedingly rare in renal pelvis neoplasms, and a signet ring cell formation in this kind of tumor is even more uncommon. Our report presents a unique case of this tumor with a concise review of the literature. From this case, we can conclude that primary mucinous adenocarcinoma with a signet ring cell formation may be a sign of poor prognosis.

KEY WORDS: metastasis, mucinous adenocarcinoma, pathological diagnosis, renal pelvis, signet ring cell

INTRODUCTION

Primary mucinous adenocarcinoma of the renal pelvis (MARP) is an exceptionally rare tumor with a poor prognosis, while MARP with a signet ring cell formation is even more uncommon[1,2]. Our report presents a case of MARP with signet ring cell changes diagnosed by pathological examination. We conclude that MARP with signet ring cell differentiation is a predictor of poor prognosis.

CASE REPORT

A 30-year-old woman was admitted to our hospital because of abdominal pain and discomfort. She denied having hematuria or a fever. Physical examination revealed an abdominal mass. After an abdominal ultrasound evaluation, a large cystic tumor was found in her left retroperitoneal area. Tests of routine blood, blood coagulation function, and liver and kidney function were unremarkable. Elevated serum levels of CA199 and CEA were 56.7 U/ml (normal value: less than 27U/ml) and 236.3 ng/ml (normal value: less than 5ng/ml) respectively. Computed tomography and magnetic resonance imaging were performed and showed a 11.0 cm solid-cystic calcified tumor located in the left kidney (Figure 1). Local enlarged lymph nodes and multiple lesions in the liver and lung were discovered. A clinical diagnosis of primary renal cancer and distant metastasis was considered. The patient consented to a laparoscopic radical nephrectomy via the intraperitoneal route without lymph node dissection. Gross examination of the specimen showed a cyst-solid mixed tumor containing a significant amount of thick, jelly-like mucus (Figure 1). Hematoxylin-eosin (Figure 2) staining showed an abundant mucin pool. Neoplastic cells with hyperchromatic pleomorphic nuclei infiltrated the renal pelvis and some floated in the mucin pool. However, the tumor did not invade the adjacent adrenal gland and renal parenchyma. Some neoplastic cells had signet ring cell features. The mucous membrane of the renal pelvis was completely eroded by tumor cells. The patient followed up for three months and did not have surgical complications or disease progression. She continued therapy on Chinese medicine only.

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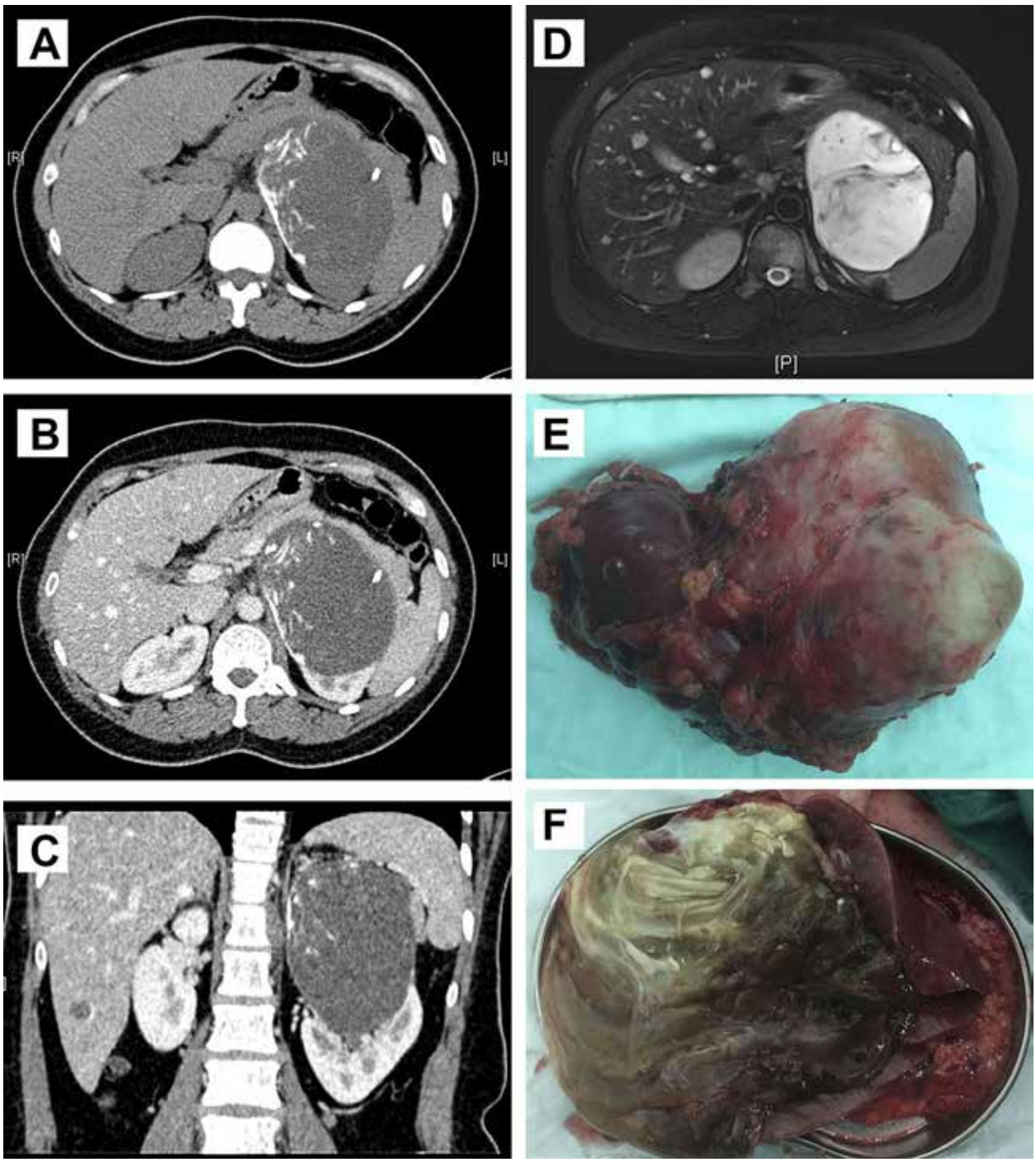

Fig 1: Axial computed tomography (CT) (A, non-enhanced; B, contrast-enhanced) and coronal CT (C) images show a giant cyst-solid mixed tumor located in the left kidney. Multiple lesions in the liver were also discovered (B, C, and D, magnetic resonance images). Gross investigation showed a cyst-solid mixed tumor containing an abundant thick, jelly-like and taupe mucus (E and F).

DISCUSSION

The rarity of primary MARP makes a preoperative diagnosis difficult to achieve, as patients with MARP usually present with nonspecific symptoms or may even be asymptomatic. Some patients complain of hematuria. Our case presented with abdominal pain only, which is considered a sign of late-stage MARP[1].

The pathogenesis of MARP is still unknown. According to previous clinical reports, it may be associated with chronic inflammation and long-term irritation by stones[2]. Our case also had stones or calcified plaques in the tumor; however, it is uncertain

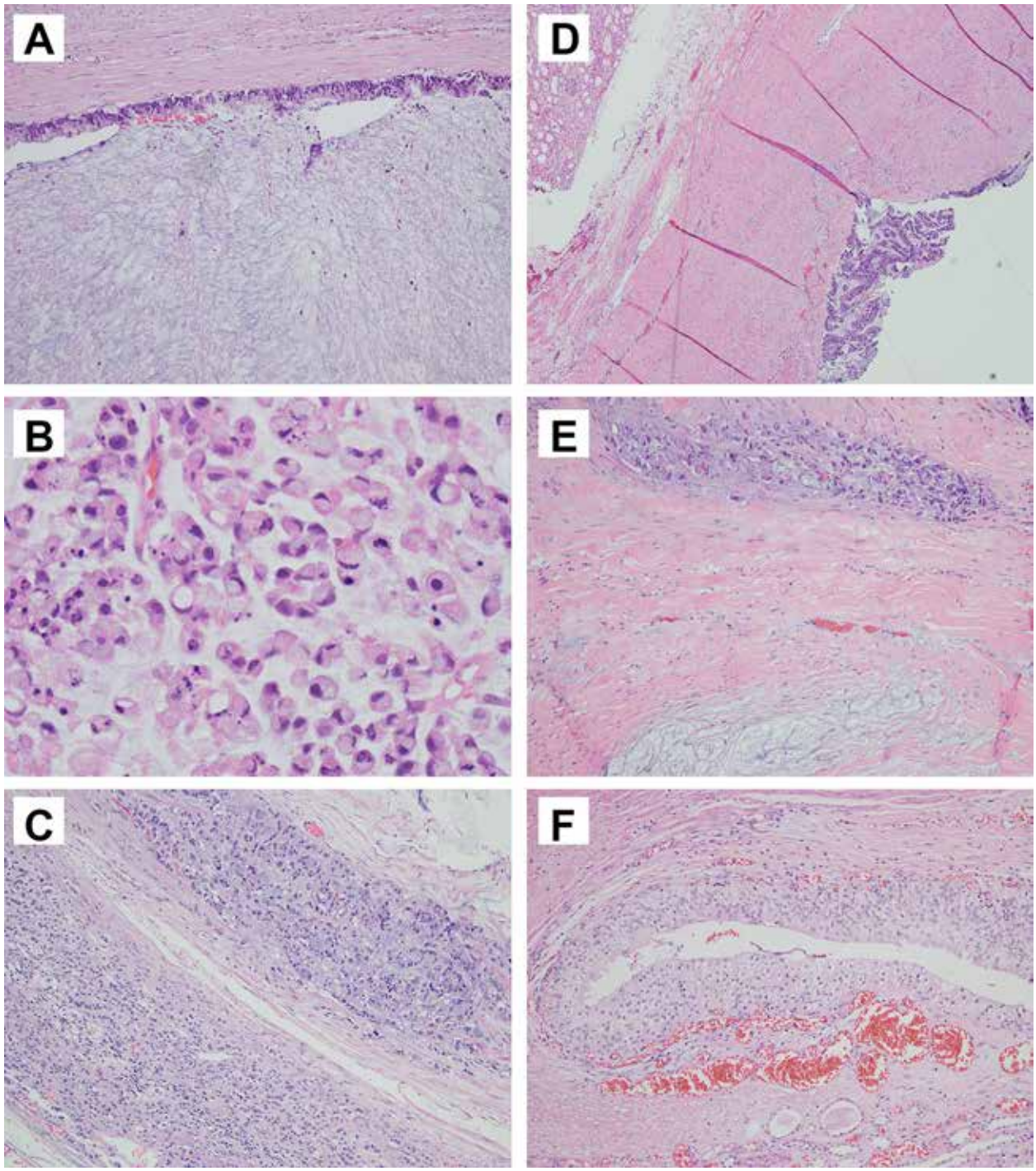

Fig 2: Hematoxylin-eosin (HE) staining presented an abundant mucin pool and neoplastic cells with hyperchromatic pleomorphic nuclei infiltrating the renal pelvis and floating in the mucin pool (A, 100×). Some of the neoplastic cells had signet ring cell features (B, 200×). The adjacent adrenal gland (C, 100×) and renal parenchyma (D, 100×) were not invaded by the tumor. The neoplastic cells infiltrated into the renal pelvis resulting in mucosal and mesenchymal destruction (E, 100×), while no neoplastic cells were observed in other parts of the renal pelvis (F, 100×).

whether they preceded the tumor or were secondary to it. We assume the latter, as multiple calcified lesions were observed in the pool of mucus. Elevated CEA and CA19-9 were considered predictors of MARP prognosis[3,4]. The same results were also discovered in the presented case. Previous data indicated that MARP has a poor prognosis as most patients die within 2 to 5 years[4]. MARP with a signet ring cell formation is rarely uncommon, and we consider it a sign of poorer prognosis as mucinous adenocarcinoma in the breast

or colon, with signet ring cell formation typically results in a significantly poorer prognosis[5,6]. As mentioned above, we observed local enlarged lymph nodes together with multiple lesions in the liver and lung but noted no progression of the disease three months postoperatively.

CONCLUSION

In this study, we reported a unique case of primary MARP with a signet ring cell formation, which may indicate a poor prognosis.

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Author's contribution: Wei Yongbao prepared the draft of manuscript. Cheng Hui and Li Tao participated in the case diagnosis and management and follow-up. Li Tao sponsored the study. All authors read and approved the final manuscript.

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