

CASE REPORT

Mucinous cystadenocarcinoma arising in an ectopic kidney simulating a retroperitoneal dermoid cyst: a rare tumour presenting as a diagnostic dilemma

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Abstract

Primary mucinous cystic neoplasms are rare tumours of the kidney, with a very few case reports in the literature. They arise from metaplasia of renal pelvic urothelium. We describe here a 45-year-old male who presented with pain in the abdomen and a lump in the left iliac fossa for two months. Ultrasound and CT scan showed a large, complex, heterogenous mass in the central abdomen and left iliac fossa, suggesting the possibility of dermoid cyst. Excision of the mass showed an enlarged multicystic kidney filled with mucin, destruction of renal parenchyma and a small viable area of grey white tumour. Histopathology revealed a peripherally located mucinous cystadenocarcinoma arising in the background of chronic pyelonephritis and mucinous metaplasia. We report this case for the rarity of the lesion and the associated clinical and radiological diagnostic dilemma.

Keywords: mucinous adenocarcinoma, renal pelvis, mucinous cystic neoplasm, cystadenocarcinoma

INTRODUCTION

Although renal tumours mostly originate from the renal parenchyma, the urothelium rarely gives rise to epithelial tumours.¹ Mucinous adenocarcinoma of the renal pelvis is an extremely uncommon tumour.² Only a few such cases have been published as isolated case reports. The experience and knowledge about renal mucinous cystic neoplasms are extremely limited and they have not been recognized formally in the WHO classification of tumours of the pelvicalyceal system.³ We describe a peripherally located renal mucinous adenocarcinoma arising in a background of chronic pyelonephritis and mucinous metaplasia in a non-functioning ectopic left kidney, clinically and radiologically simulating a retroperitoneal dermoid cyst.

CASE REPORT

A 45-year-old male presented with a history of abdominal pain and abdominal mass for 2 months. Ultrasonography and CT abdomen showed a large, heterogenous mass in the central abdomen and left iliac fossa having both solid

and cystic components and measuring 19x18x12 cm, suggestive of a dermoid cyst. The left kidney could not be visualized (Figure 1a). Intravenous pyelogram gave an impression of absence of the left kidney and normal functioning right kidney (Figure 1b). Tc99m DTPA renogram revealed normal glomerular and cortical function of the solitary right kidney. The left kidney could not be localized. A repeat ultrasonography suggested an ectopic left kidney with a calculus and marked hydro/pyonephrosis. A biopsy was attempted and showed foamy histiocytes and necrosis. With the two clinical and radiological possibilities of hydronephrotic ectopic left kidney and huge dermoid cyst in left iliac fossa with absent left kidney, the patient was referred to our institute and taken up for surgery. Intraoperatively, a large, well marginated retroperitoneal mass was identified arising from left side, crossing the midline, sitting on left iliac vessels and was possibly an ectopic enlarged non-functioning left kidney. An excision of the mass was performed. The postoperative course was uneventful. The patient is well one and a half years after the operation.

Pathology

Grossly, the excised mass measured 17x13x9 cm. The external surface was bosselated (Figure 1c). The cut surface showed large cystic spaces filled with friable gelatinous material, marked thinning out of normal renal parenchyma and peripherally located solid grey white tumour (Figure 1d). Microscopical examination revealed glands, cysts and papillae lined by stratified columnar epithelium with stratification, hyperchromatic pleomorphic nuclei and vacuolated cytoplasm along with signet ring cells lying in mucin pools focally (Figure 2). The adjacent renal parenchyma showed sclerosis of glomeruli, interstitial fibrosis and inflammation, aggregates of foamy and hemosiderin-laden histiocytes, cholesterol clefts and giant cells along with focal calcification and mucin pools (Figure 3 a-e). In addition, mucinous metaplasia of the urothelium was identified (Figure 3f). The tumour did not breach the capsule and the perinephric fat was free from the tumour.

Considering the histomorphological features, a diagnosis of mucinous adenocarcinoma arising in a background of chronic pyelonephritis and mucinous metaplasia was made.

DISCUSSION

Tumours of the renal pelvis are uncommon, with about 90% being transitional cell carcinomas and 10% squamous cell carcinomas.² Mucinous cystadenocarcinoma of the renal pelvis is a very rare tumour that accounts for less than 0.3% of renal pelvic tumours.⁴ Hasebe *et al* first described mucinous cystadenocarcinoma of the renal pelvis in 1960.³ The renal pelvis is the general location for mucinous cystadenocarcinoma but they may arise from the urothelium of the bladder, ureter and renal calyces. Only a few cases with atypical localization have been reported in the literature.¹ The tumour in our case originated from the periphery of the kidney.

The frequent association of this malignancy with chronic irritation, inflammation, infection, hydronephrosis and urinary calculi forms the basis of pathogenesis. Glandular metaplasia of the urothelium developing in response to injury may progress to dysplasia and adenocarcinoma.⁵⁻⁷ Some of the authors have also postulated a teratomatous or coelomic epithelial origin. Another theory of origin is that these tumours may arise from the sequestered

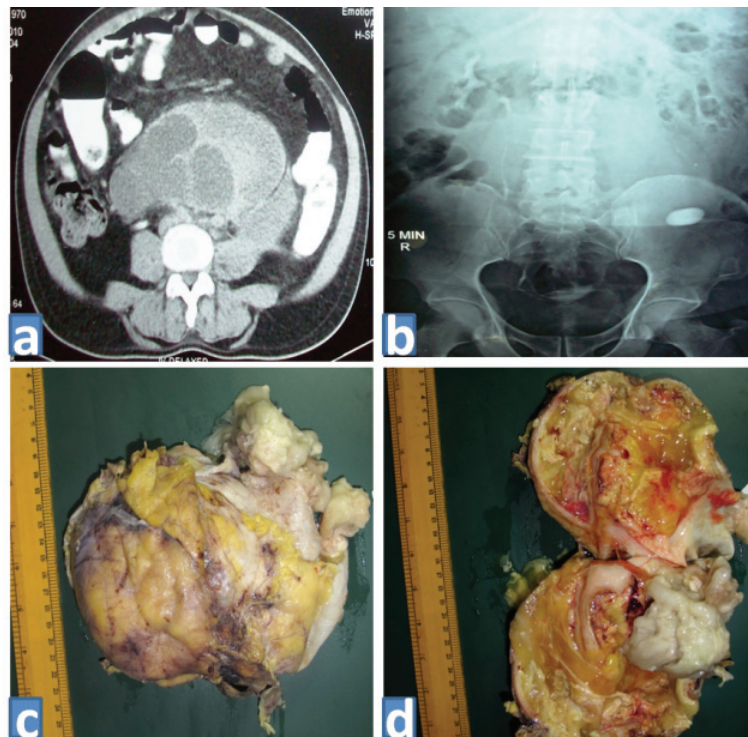


FIG. 1: (a) CT revealing large heterogeneously enhancing abdominal mass; (b) IVP showing normal functioning right kidney and absent left kidney function; (c) Resected left kidney with a bosselated external surface; (d) Cut surface showing enlarged cystic kidney filled with gelatinous material and greyish white tumor

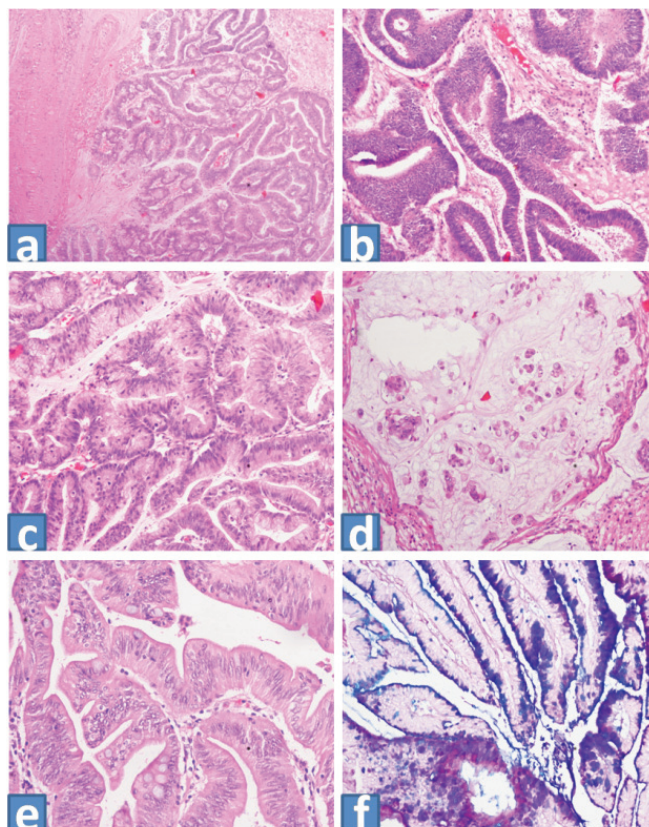


FIG. 2: (a) Complex glandular architecture (X40); (b) Stratification of columnar epithelium (X100); (c) Mucin vacuoles in tumour cells (X100); (d) Signet ring cells floating in mucin pools (X100); (e) Cellular and nuclear atypia (X200); (f) Mucin in tumour cells demonstrated by alcian blue – periodic acid Schiff staining (X100)

renal pelvic epithelium within the parenchyma as a consequence of maldevelopment. The above mentioned hypothesis is supported by the increased incidence of the lesions found in anomalous kidneys.² In our case the ectopic and non-functioning nature of the kidney was the possible predisposing factor.

A few cases have been seen in association with carcinoma-in-situ of the bladder and ureter.^{6,8} Renal mucinous neoplasms can present in wide clinical settings, ranging from pseudomyxoma peritonei to silent cystic lesions without any complication. Although, most patients are asymptomatic, hematuria, mucosuria, flank pain, and palpable abdominal mass are the other clinical presentations.^{1,3,9} Radiological studies may not be diagnostic as in our case.⁵

Aufderheide and Streitz proposed the criteria for malignancy after a review of 28 cases. Their criteria were (1) architectural or cellular atypia, (2) microscopical evidence of invasion of the renal pelvic wall, renal parenchyma, or nodal

or distant metastases, and (3) overt invasion, recurrence, or metastases.¹⁰ The first 2 criteria were observed in the present case. Spires *et al* classified adenocarcinomas into various subtypes: tubulovillous, mucinous and papillary non-intestinal.² As mucinous cystadenocarcinoma is an extremely uncommon entity in the kidney, the patient must be carefully evaluated for primary tumours of pancreas, ovary and appendix which are more common.³ These were not seen in our case.

The prognosis is generally poor, with about 1/2 of the patients dying within 2 years of surgery. Local recurrence possibly results from the spillage of tumour cells during surgery and downward seeding in the distal ureter. Radical nephrectomy and complete removal of the ureter is the preferred surgical treatment. Adequate precautions against spillage should be taken intraoperatively for the suspected cases.¹⁰

To conclude, these tumours represent a separate entity that should be included in the

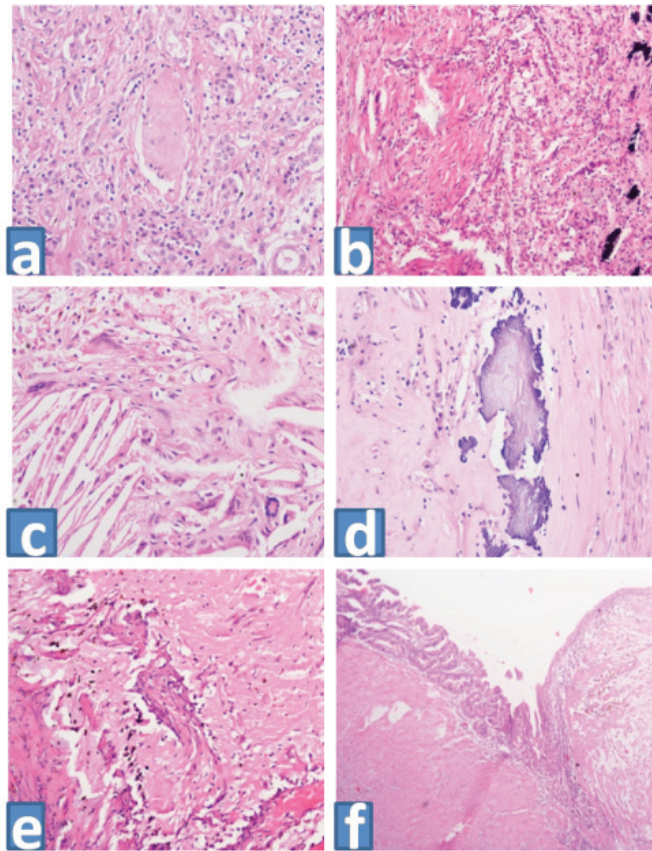


FIG. 3: (a) Chronic pyelonephritis in the adjacent renal parenchyma (X100); (b) Groups of foamy and hemosiderin-laden histiocytes (X100); (c) Cholesterol clefts and giant cells (X200); (d) Focal calcification (X100); (e) Pools of mucin (X100); (f) Mucinous metaplasia of urothelium (X40)

WHO classification of renal and urothelial tumours so that the diagnosis can be suspected preoperatively for adequate treatment of the patient. This possibility should also be kept in mind in patients with a non-functioning enlarged cystic ectopic kidney.

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