A case series of sarcomatoid transitional cell carcinoma of renal pelvis and collecting duct carcinoma of kidney

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ABSTRACT

Sarcomatoid transitional cell carcinoma (TCC) of the renal pelvis and collecting duct carcinoma (CDC) of the kidney are rare types of malignant renal tumors with aggressive behavior. Here, we report a case series of these tumors, in males of age 65 years and 72 years, respectively. In both the cases, the left kidney was affected and they underwent the left-sided radical nephroureterectomy. Histologically, sarcomatoid TCC showed two components with invasive TCC and areas with spindle cells and osteoclastic type of giant cells. The immunohistochemistry showed diffuse positivity for CK7 and negativity for CK20 and p63 in both the components. CDC showed tubule-papillary patterns with marked desmoplasia. The patients were disease free after a follow-up of 12 months and 2½ years, respectively, without any adjuvant therapy.

Keywords: *Carcinoma, Collecting duct cell carcinoma, Kidney, Renal pelvis, Sarcomatoid transitional cell carcinoma*

Sarcomatoid transitional cell carcinoma (TCC) of the renal pelvis is an extremely rare tumor and accounts for 0.3% of all urothelial carcinomas [1] with sarcoma-like components arising from the malignant transitional epithelium and shows aggressive behavior. Due to its rarity, there are only a few studies concerning the treatment efficacy of sarcomatoid TCC. At present, surgical resection remains the first line of treatment.

Collecting duct carcinoma (CDC) is another rare histological variant of renal cell carcinoma (RCC), arising from the epithelium of distal collecting ducts, also called Bellini duct cell carcinoma. It represents approximately 2% of all the RCCs and is highly aggressive [2]. We, herein, report the clinical and pathological features of one case each of sarcomatoid TCC and CDC.

CASE REPORT

Case 1

A 65-year-old male patient presented with intermittent hematuria and the left flank pain for 6 months duration. On physical examination, the vitals were stable and there was no palpable abdominal mass. The computed tomography (CT) scan revealed an ill-defined lesion in the pelvis of the left kidney (Fig. 1a) and a left laparoscopic nephroureterectomy was performed. The gross picture showed that the nephrectomy specimen measured 9 cm×7 cm×5 cm, along with perinephric fat and ureter of length 10 cm. Cut section showed an ill-defined gray-white to tan, firm lesion, with some friable areas, in the pelvicalyceal region, measuring 5 cm in greatest dimension (Fig. 1b). No lymph nodes were identified.

Microscopic examination showed a tumor composed of two components: Invasive TCC (Fig. 2a and b) and spindle cell area with some bizarre cells (Fig. 2c), matrix-like material, and many osteoclastic types of giant cells (Fig. 2d). The adjacent kidney showed chronic glomerulonephritis changes. Immunohistochemistry was diffusely positive for CK7 in both the components and negative for CK20 and p63, indicating epithelial origin. A diagnosis of sarcomatoid TCC of renal pelvis was made. No adjuvant chemotherapy was given. A follow-up was done after 12 months of surgery, and the patient had no complaints and was disease free.

Case 2

A 72-year-old male presented with the left side flank pain and three episodes of hematuria, in 2 weeks period. On physical examination, the vitals were stable and there was mild tenderness of the left flank with no palpable mass. The CT scan revealed an ill-defined, low-attenuation lesion, located in the renal medulla of the left kidney. He underwent the left-sided nephroureterectomy, with a pre-operative diagnosis of T2N0M0.

A gross picture showed the nephrectomy specimen (Fig. 3a) measured 15 cm×8 cm×5 cm, along with perinephric fat and ureter of length 5 cm. Cut sections showed a gray-white to tan lesion at the confluence of medulla and cortex with irregular

outline and are infiltrating into the perinephric fat, at the pelvis. The renal capsule showed adhesions in some areas. Microscopic examination showed a tumor with tubule-papillary morphology accompanied by marked desmoplasia (Fig. 3b and c). The lining epithelium of the adjacent collecting ducts showed cytological atypia (Fig. 3d), suggesting a diagnosis of CDC of the kidney. Adjuvant chemotherapy or radiotherapy was not given. A follow-up done after 2½ years showed that the patient was disease free and without recurrence.

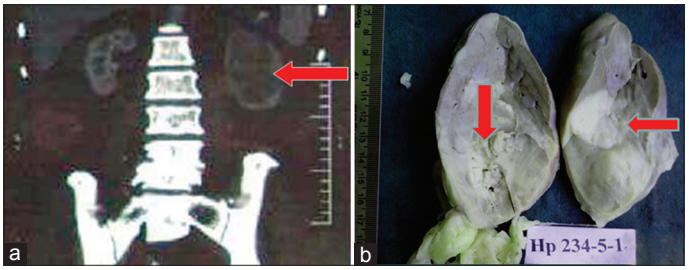

Figure 1: (a) Computed tomography scan picture showing the left kidney lesion; (b) gross picture of the left nephrectomy specimen along with perinephric fat

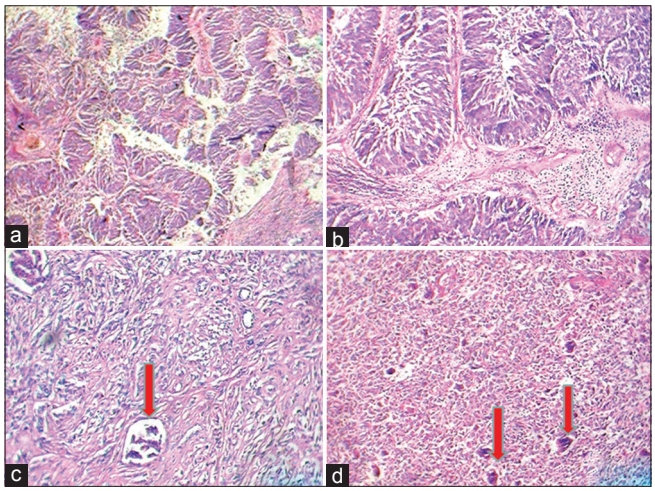

Figure 2: (a) Invasive transitional cell carcinoma (TCC) component, H and E, ×10; (b) TCC, H and E, ×40; (c) tumor with spindle cell areas, infiltrating into adjacent kidney parenchyma, glomerulus, H and E, ×10; (d) tumor with osteoclastic type of giant cells, in spindle cell areas, H and E, ×10

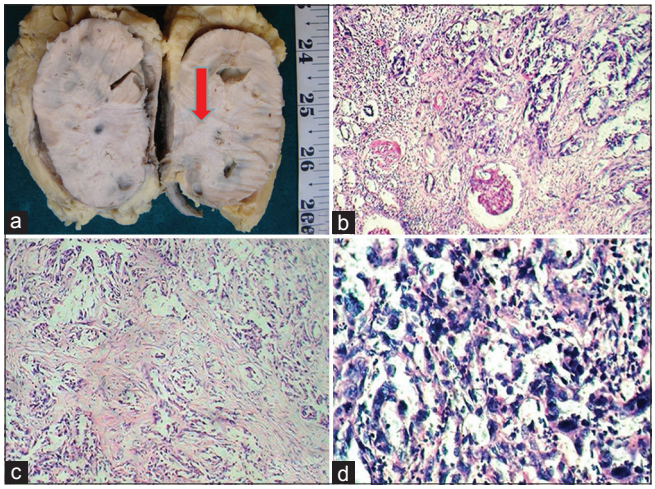

Figure 3: (a) Gross picture showing cut section of the left nephroureterectomy specimen; (b) tumor with tubulopapillary patterns, infiltrating into the adjacent renal parenchyma, shows glomerulus, H and E, ×10; (c) tumor with inflammatory desmoplastic stroma, H and E, ×10; (d) tumor cells with marked pleomorphism, H and E, ×40

DISCUSSION

Sarcomatoid carcinoma is a high-grade epithelial neoplasm characterized by a biphasic appearance with epithelial component and another area with a sarcoma-like appearance. Carcinosarcoma is a term frequently confused with sarcomatoid carcinoma, which consists of a malignant epithelial component and specific features of mesenchymal differentiation, such as chondrosarcoma, osteosarcoma, and liposarcoma. Due to similar morphologies in microscopic appearance with hematoxylin and eosin staining, immunohistochemistry is necessary for a diagnosis.

TCC is the most common type of malignancy occurring in the renal pelvis. However, a sarcomatoid variant of TCC is extremely rare. Perez-Montiel *et al*. [3] studied 108 cases of high-grade urothelial carcinomas and only eight were sarcomatoid TCCs, with the advanced involvement of parenchyma and very poor prognosis with a mean survival time of 11.2 months. Adjuvant chemotherapy offered no response. Froehner *et al*. [4] suggested that it would be appropriate to treat these tumors in the same manner as conventional high-grade TCC, with the gemcitabine-based regime.

In a retrospective study by Chen *et al*., [5] among eight cases diagnosed as renal pelvis sarcomatoid carcinoma, all were males and the majority presented with intermittent hematuria. Radiological findings showed a tumor in the renal pelvis. On gross examination, the mean tumor size was 5.8±4.5 cm. Of eight patients, six succumbed to the disease while two patients were free of disease at 54 and 120 months, respectively. The present case of sarcomatoid variant of TCC is also a male 65 years, presenting with hematuria and tumor of size 5 cm greatest dimension in the renal pelvis. The patient remains disease free, 12 months after laparoscopic nephroureterectomy and without adjuvant chemotherapy.

CDC is a rare and an unusual variant of RCC. Cheville *et al*. [6] studied 2385 patients, whose first surgery between 1970 and 2000 was a radical nephrectomy for sporadic unilateral RCC, which included only 6 cases (0.3%) of CDC. The first report that the ducts of Bellini could give rise to tumors was made by the findings of atypical hyperplastic change in the collecting duct epithelium, distant from a renal papillary tumor [3]. Later, it has been grouped in RCC with a subdesignation "Bellini duct carcinoma" in an international classification of tumors by the WHO [7]. Since then just over 100 cases have been describes [8]. Criteria for diagnosing CDC are central location, irregular tubular architecture, high-nuclear grade, reactive with high-molecular-weight cytokeratin, inflammatory desmoplastic stroma, and extensive renal, extrarenal, and lymphovascular infiltration [7]. The typical CDC has a poor prognosis with many being metastatic at presentation. About two-thirds of patients die of their disease within 2 years of diagnosis [9].

In a study by Wilson Sui *et al*. [10], among 2016, 86 cases of RCC, there were 577 cases of CDC and they presented with

higher grade and stage, node positive, and metastatic disease. Overall, median survival for CDC was 13.2 months compared to 122.5 months for clear cell RCC. They concluded that CDC is an aggressive subtype of RCC and adjuvant chemoradiation after nephrectomy showed survival benefit over single-mode therapy. In the present case, the tumor is at the confluence of medulla and cortex, with tubulo-papillary patterns, of high nuclear grade and extensive desmoplasia. The tumor shows infiltration into perinephric fat and is labelled as CDC of kidney-papillary type. The patient succumbed to disease 26 months after radical nephroureterectomy, without any adjuvant chemotherapy.

CONCLUSION

Sarcomatoid TCC of the renal pelvis and CDC of the kidney are very rare high-grade neoplasms with a very poor prognosis. For early localized disease, a radical, aggressive operation should be considered to improve the outcomes. However, systemic chemotherapy with a gemcitabine-based regimen may be offered for sarcomatoid TCC, due to its urothelial origin.

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