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Case Report

Renal Pelvis and Renal Calices Sarcomatoid Carcinoma: A Case Report

Qiliang Yin¹, Shijie Lan¹, Fengming Jiang² and Di Wu^{1*}

¹Department of Oncology, The First Hospital of Jilin University, Changchun, Jilin, China

²Department of Urinary Surgery, The First Hospital of Jilin University, Changchun, Jilin, China

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ABSTRACT

Introduction: Sarcomatoid carcinoma is one of the malignant neoplasms characterized by the ability to differentiate into both epithelial tissue and mesenchymal tissue. And it is rare in urinary system with a high degree of malignancy and prognosis is poor.

Patient Concerns: The present article reports a 63-year-old man presented with recurrent left flank pain for two years. Radiological tests suggested a space-occupying lesion in left kidney with enlarged paranephric/retroperitoneal lymph nodes.

Diagnosis: The pathological diagnosis following left nephrectomy indicated renal pelvis and renal calices sarcomatoid carcinoma; pTNM: T4N0M0.

Interventions: No radiotherapy or chemotherapy were performed after the surgery and underwent rapid growth.

Outcomes: The patients died six months later.

Conclusion: Renal pelvis and renal calices sarcomatoid carcinomas is a highly malignant tumor and difficult to diagnose at its early stage. Definite diagnosis relies on the combination of immunohistochemistry (IHC) and morphological analyses and there currently is no effective treatment.

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Introduction

Renal pelvis and renal calices sarcomatoid carcinoma is very rare in the clinical practice. The incidence of this disease is unclear. According to our literature search, less than 30 cases of sarcomatoid carcinoma was reported until now [1-3]. Thus, it is impossible to summarize the character of this disease systematically. One renal pelvis and renal calices sarcomatoid carcinoma patient was presented in our center in 2015. The objective of this article was to describe the clinical features of this case and review the pathology, immunohistochemistry (IHC), treatment and prognosis in accordance with the available literature.

Case Report

A 63-year-old man presented with recurrent left flank pain for two years and aggravated for one month was referred to our medical center on 9th

Dec 2015. There were no symptoms with fever, frequent micturition, the urgency of urination, the urine pain and gross hematuria. PE: there was tenderness in left lumbar region. No abdominal mass was accessed. Radiological tests suggested space-occupying lesion in left kidney with enlarged paranephric/retroperitoneal lymph nodes (Figures 1a & 1b). Renal needle biopsy suggested high grade renal carcinoma. IHC panel: Vimentin (+), CK-pan (+), CK7 (+), CD 10 (+), PAX-2 (±), SMA (-), CD34 (-), HMB45 (-) and P63 (-). Left nephrectomy was performed on 18th Dec 2015. The tumor size was about 6*4*4 cm and gray in colour. Mucous membrane of renal pelvis was smooth. The pathological diagnosis following left nephrectomy indicated renal pelvis and renal calices sarcomatoid carcinoma; pTNM: T4N0M0. IHC panel: CK-pan (+), CK7 (partially +), Vimentin (partially +), CD10 (sporadic +) and P63 (-). After the consultation with the pathologist from Peking Union Medical College Hospital, renal cell carcinoma was excluded, and we decided that the diagnosis was undifferentiated sarcomatoid carcinoma.

*Correspondence to: Di Wu, Department of Oncology, The First Hospital of Jilin University, 1 Xin-Min Street, Changchun, Jilin, 130021, China; E-mail: wudi888991@163.com

No radiotherapy or chemotherapy was performed after the surgery. Radiological tests on 18th Mar 2016 suggested the recurrence of tumor

(Figure 1c). According to the desire of the patient's family, no additional treatment was performed. And the patients died six months later.

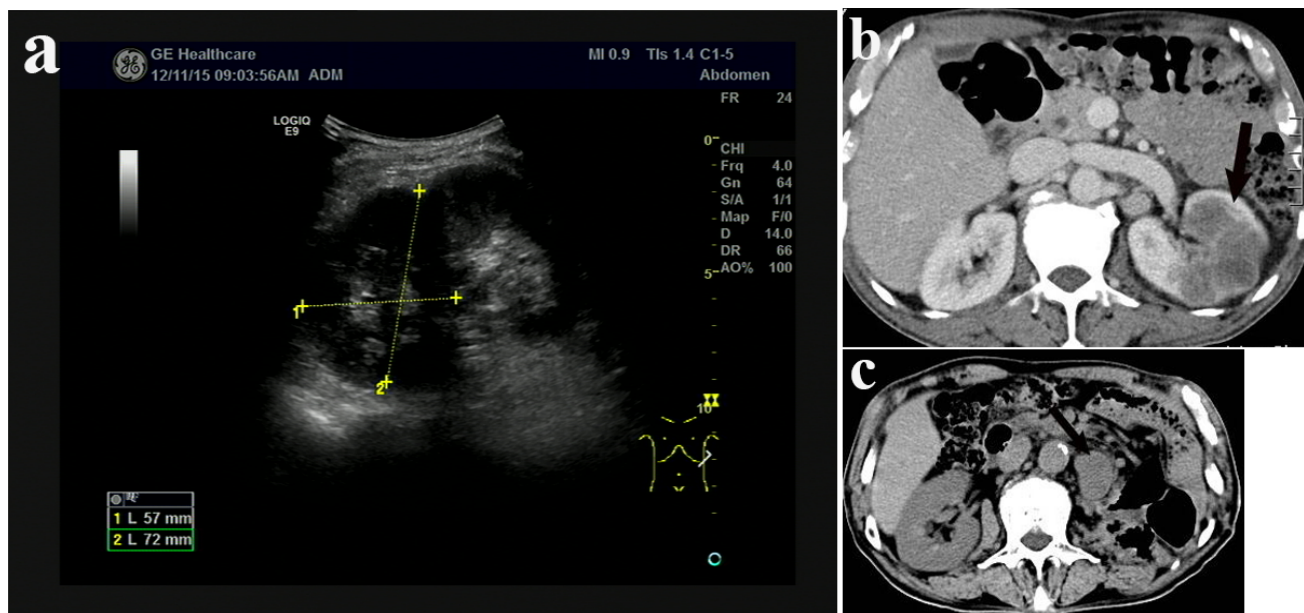


Figure 1: a) Intravenous urogram indicate that left kidney is non-significantly enhanced. Kidney size is 27*30 mm. The enhancement of the lesion decreased slightly in the venous phase. Space-occupying lesion could be identified, and renal cancer could not be excluded. b) Renal CT plain scan and three phases enhancement scan show untypical hypodense shadow in the middle-upper end of the left kidney. The largest section is about 5.6*4.6 cm. CT-value is about 30HU. The density of lesion is low in either of the three phases. The boundary is unclear. These all indicate space-occupying lesion while renal cancer could not be excluded. Enlarged paranephric/retroperitoneal lymph nodes could also be found. c) CT scan shows that after left nephrectomy and lienectomy, the local connective tissue was unclear, and multiple nodules could be found. Tumor recurrence or metastasis could not be excluded.

Discussion

The incidence of renal pelvis and renal calices sarcomatoid carcinoma is low. The reported cases indicated that the ratio of male patient/female patient is 2:1~3:1, and the older patients, right side lesion are more commonly seen [1-3]. There is no specific clinical manifestation indicating the onset of renal pelvis and renal calices sarcomatoid carcinoma. Some non-specific manifestations include gross hematuria and flank pain/ swelling and uronephrosis [1-6]. The early diagnosis is quite difficult for this disease. The sensitivity of radiological tests limits the chance of discovering the tumor in early stage. Radiological

manifestation usually indicates the inferior TNM staging. Definite diagnosis relies on pathological test and immunohistochemistry following nephrectomy and percutaneous renal biopsy. There is no treatment guideline available for this disease. Nephrectomy is performed in most of the patients, while chemotherapy and radiotherapy after nephrectomy are less commonly used [2, 4, 6, 7]. The available evidence suggests that chemotherapy and radiotherapy have little effect on long term survival rate [2, 6]. Prognosis of this disease is poor. Most patients die in the first two years after the diagnosis. TNM stage of the neoplasm is associated with the prognosis [7, 8].

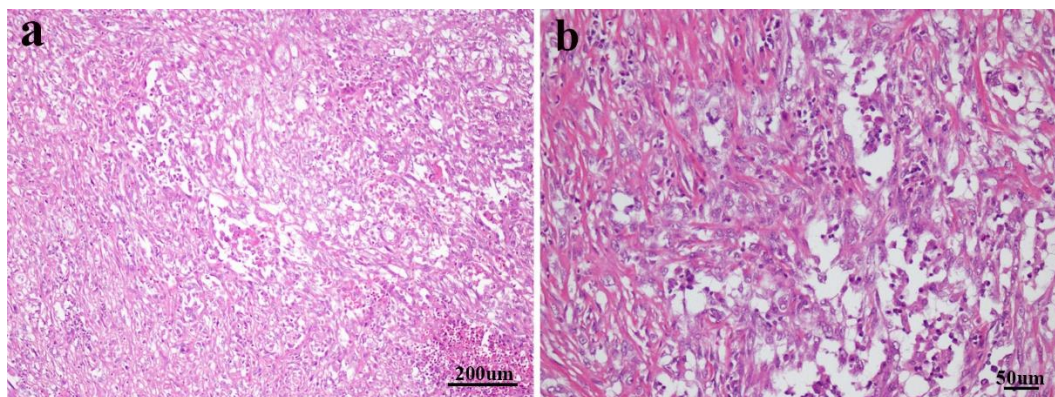
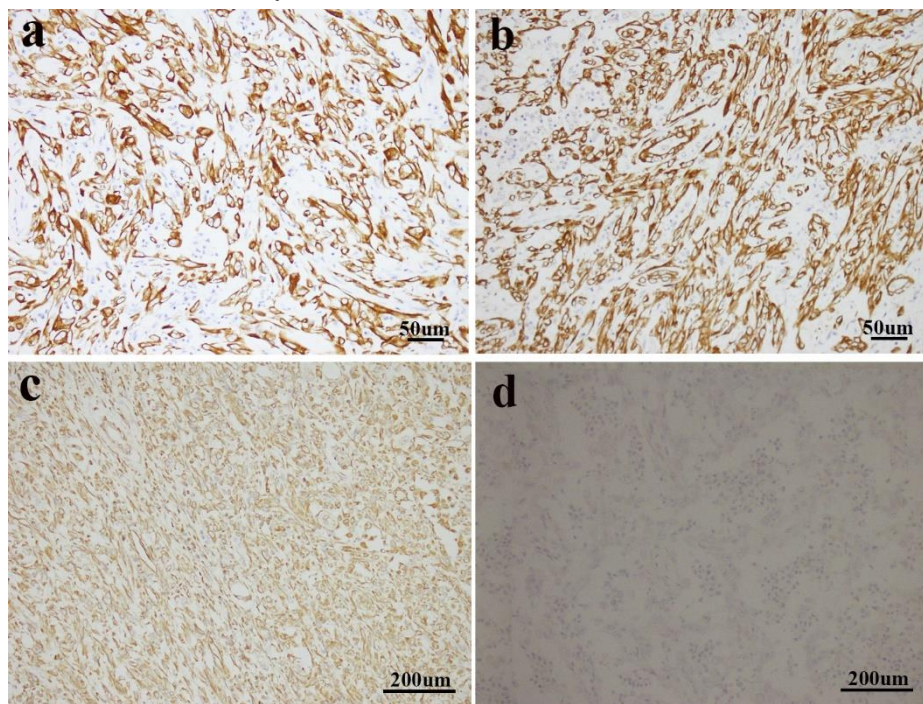


Figure 2: a) The cells of sarcomatous area are spindled and patchy in distribution. Necrosis could be seen in lower right side (stain, hematoxylin and eosin; magnification, x100). b) Some of sarcomatoid cells were epithelioid, rich in cytoplasm, staining weakly, vacuole nucleus and nucleoli could be seen (stain, hematoxylin and eosin; magnification, x200).

Grossly, the mass of renal pelvis and renal calices sarcomatoid carcinoma is usually large (diameter ≥ 4 cm), and it may be described as solid and papillary and the transverse section was white and gray [2, 4-6]. The structure characteristics in this case are identical with this general description. While the mass in this case also infiltrated to sinus renalis, renal parenchyma and perinephric fat. The tumor consists of high grade sarcomatoid tumor cells. Histological examination can also find tumor cells from epithelium origin and other abnormal cells, which indicating the nature of mixed tumor of this disease [1, 2, 4, 6, 9]. The malignant epithelial cells include transitional cell carcinoma cells, squamous carcinoma cell, adenocarcinoma cell. etc. The real sarcoma cells (osteosarcoma, chondrosarcoma, rhabdomyosarcoma or

leiomyosarcoma) were absent in this disease. There is a transitional area between the tissue from epithelium origin and sarcomatoid tissue. Cytoplasm of the tumor cells is eosinophil.

The tumor cells have increased size of the nucleus; increased chromatin content; abnormal chromatism distribution. The tumor usually possesses large number of mitoses, especially producing atypical ones [1, 4, 6]. The tumor cells in this case were spindled and patchy in distribution. Necrosis could be seen. Some tumor cells were epithelioid, rich in cytoplasm, staining weakly, vacuole nucleus and nucleoli could be seen (Figures 2a & 2b).



Figures 3: a & b) Sarcomatoid cells demonstrated positive expression of CK7 and CK-pan (immunostaining; magnification, x200). **c)** Vimentin was strongly expressed in the sarcomatoid area (immunostaining; magnification, x100). **d)** Sarcomatoid cells showed negative expression of P63 (immunostaining; magnification, x100).

Immunohistochemistry (IHC) test is crucial for the diagnosis of renal pelvis and renal calices sarcomatoid carcinoma. Typically, both CK and vimentin stainings are positive. The tumor cells origin from epithelium express CK and EMA, while tumor cells from mesenchymal tissues express vimentin. Both CK and vimentin positivity may indicate the process of metaplasia from epithelium to mesenchymal tissues in the tumor. Desmin, muscle specific actin and S-100 protein are usually not expressed [2, 3, 6, 10]. The IHC results for this patient agreed with the general features above mentioned (Figures 3a-3c). Combined with IHC with morphology results, the definite diagnosis could be confirmed. P63 is homologous with p53. The positive P63 may be associated with the higher grading/higher staging and the sarcomatoid nature of the tumor [3, 11]. We performed P63 staining for this case and it is negatively stained (Figure 3d).

Differential diagnosis also relies on the combination of IHC and morphology analyses. In clinical practice, renal pelvis and renal calices sarcomatoid carcinoma should be distinguished from the following

tumors: 1) Sarcoma, epithelium IHC biomarkers are negative for sarcoma. In addition, cytodema and intermediate filament are absent in sarcoma. 2) Renal sarcomatoid carcinoma, this is a high-grade malignant tumor in renal parenchyma. The differences between this disease and renal pelvis and renal calices sarcomatoid carcinoma are the gross structure and tumor cell origins. Renal pelvis and renal calices sarcomatoid carcinoma are usually papillary, and transitional carcinoma cells could be found in area of sarcomatoid carcinoma [4, 12]. 3) Carcinosarcoma, this contains both carcinoma and sarcoma in its tumor mass. Pathologists can find osteosarcoma, chondrosarcoma and rhabdomyosarcoma, while IHC evidence in epithelium differentiation lacks in carcinosarcoma. Sarcomatoid carcinoma mainly consist of epithelium tumor cells. Both CK and Vimentin are expressed in sarcomatoid carcinoma [2, 3, 5]. The differential diagnosis between two diseases have no impact on the intervention to the patients. They both indicate quick progression and poor prognosis and the clinical manifestations are similar [4].

There are no standard treatment guidelines for this disease. Most reported cases underwent nephroureterectomy, while almost no chemotherapy or radiotherapy were performed on these patients [2, 7]. Previous study proposed that surgery should be avoided for the tumor staging higher than T4, because these patients may not benefit from surgery [13]. EGFR expression in urethra sarcomatoid carcinoma is upregulated, which indicates that targeting therapy could be a potential treatment option for renal pelvis and renal calices sarcomatoid carcinoma [7]. Furthermore, the prognosis of this disease is poor. Most died in the first two years after diagnosis and long-term survival is scarce [2, 3, 7, 14]. Renal pelvis and renal calices sarcomatoid carcinoma respond poorly to chemotherapy or radiotherapy, and this assistant treatment has little effect on overall survival benefit [2, 6, 7, 13, 14]. Previous studies demonstrated that pathological stage has been associated with prognosis [13].

Conclusion

Renal pelvis and renal calices sarcomatoid carcinoma is rare in clinical practice. It is very difficult to make an early diagnosis due to the lack of specific clinical manifestations. Definite diagnosis relies on the combination of IHC and morphological analyses. There are no standard treatment guidelines for this disease. Most reported cases underwent nephroureterectomy. The prognosis is associated with pathological staging. More data are needed to make evidence-based treatment decisions.

Abbreviations

IHC: Immunohistochemistry

CK: Cytokeratin

EMA: Epithelial Membrane Antigen

SMA: α -Smooth Muscle Actin

EGFR: Epidermal Growth Factor Receptor

Consent

Patient has provided informed consent for publication of the case.

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