

Collecting Duct of Bellini Renal
CarcinomaLiu J^{1,2,3*}, Li Y^{1,2,4*}, Su Z^{1,2,3}, Duqun Chen^{1,2,4}, Ni L^{1,2}, Mao X^{1,2}, Yang S^{1,2} and Lai Y^{1,2}¹Department of Urology, Peking University Shenzhen Hospital, China²The Guangdong and Shenzhen Key Laboratory of Male Reproductive Medicine and Genetics, Peking University Shenzhen Hospital, China³Department of Urology, Shantou University Medical College, China⁴Department of Urology, Anhui Medical University, China

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CC-BY 4.0Keywords Renal cell carcinoma;
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Abstract

Collecting Ducts Carcinoma (CDC), also known as Bellini Duct Carcinoma (BDC), is a rare variant of Renal Cell Carcinoma (RCC) arising from the epithelium of the distal collecting ducts. It has an aggressive course and poor treatment response. Up to date, there are only few reports of CDC, which may be due to unrecognized or unfamiliarity of its features and characteristics. Therefore, we present a retrospective study was done in 5 cases of CDC and makes a systematic review of the literature.

Introduction

Collecting duct carcinoma or Bellini duct carcinoma is a very rare variant of kidney carcinoma, characterized by an aggressive course and an extremely poor prognosis [1-4,5]. Unlike the more common variants of renal cell carcinoma that arise from the convoluted tubules of the renal cortex, this aggressive malignancy is derived from the renal medulla, possibly from the distal collecting ducts of Bellini. Another distinctive feature of collecting duct carcinoma is its propensity for showing infiltrative growth, which differs from the typical expansile pattern of growth exhibited by most renal malignancies at pathologic examination [2]. Collecting duct carcinomas demonstrate an aggressive clinical course. Less than one-third of patients survive more than 2 years beyond diagnosis, and up to 40% have metastatic disease at presentation [3].

Material & Methods

Between 2006 and 2012, there were 400 nephrectomies and nephron sparing surgery was performed due to renal tumors in Peking University First Hospital. Among them, five cases of Bellini duct carcinoma were reported (0.8% of patients). This study is a retrospective analysis of cases with Bellini duct tumor of the kidney.

These patients were between 34 and 68 years of age, two females and three males. The two female and one male patient presented with a history of osphalgia along with hematuria and discovered a neoplasm through the CT scan. Other two men mainly complained about backache, renal tumors were discovered incidentally on ultrasound examination of the abdomen. In addition, one of the men revealed lumbar vertebra metastasis. Routine preoperative examinations were performed (laboratory and imaging). In two case there was microscopic haematuria, in three patients increase of creatinine was noted. CT of abdomen confirmed the diagnosis of renal tumours. The medium size was 8cm, 5cm, 5cm, 5cm and 3.5cm accordingly. In all cases there was no evidence of enlarged lymph nodes and chest x-ray were unremarkable in each case. All patients were treated with radical nephrectomy. Operations and post operative period were uneventful in all Bellini patients.

Histopathology examination combined with immunophenotype revealed in all patients Bellini duct carcinoma. All tumors extended beyond the fibrous renal capsule and infiltrated the perirenal adipose tissue. An immunohistochemical profile was established and was positive for CD7 in 2 cases, vimentin in 4 cases, Epithelial Membrane Antigen (EMA) in 2 cases and high-molecular weight cytokeratins in 1 case. Two female patients are still alive after surgery, three males passed away within one year, all due to disseminated cancer (Table 1).

Discussion

CDC is a rare aggressive type of renal epithelial malignancy, comprising about 1% to 3% of all renal neoplasm [4]. Presently only 270 cases have been reported worldwide [4]. It is more common in males and in the younger population (58-61 years). Like our patient, most cases present with gross hematuria, a renal mass or flank pain [5]. In 1998, the International Classification of Tumors by the World Health Organization divided this carcinoma from renal cell carcinoma and designated it 'CDC' in malignant epithelial tumors of renal parenchyma.

Table 1: The pathologies and prognoses of CDC patients.

No.	Sex	Age	Location and size of neoplasm (cm)	Routine preoperative examination	TNM classification	Prognosis
1	F	59	right kidney,4.7cm*4.0cm*3.2cm	microscopic hematuria	T1bN0M0	14 months survival with tumor free
2	F	34	right kidney,4.8cm*4.0cm*4.0cm	increase of creatinine	T1bN0M0	17 months survival with tumor free
3	M	46	left kidney,8.3cm*7.1cm*6.7cm	increase of creatinine	T3bN0M1	died of CDC with Lumbar vertebra metastasis
4	M	68	right kidney,5.0cm*4.5cm*4.0cm	increase of creatinine	T3aN0M0	died of CDC with Lung metastasis
5	M	47	left kidney,3.1cm*3.0cm*2.2cm	microscopic hematuria	T3aN0M0	died of tumor recurrence

F: Female; M: Male

In most countries, CDC appears to be unreported since its differentiation requires extensive histopathologic and immunohistochemical examinations [4]. CDC is a high-grade renal adenocarcinoma arising in the medulla of the kidney with a prominent infiltrating tubulopapillary pattern exhibiting high-grade nuclear atypia and extensive stromal dysplasia [4,5]. Immunohistochemical markers include lectin *Ulex europaeus*, Epithelial Membrane Antigen (EMA), and High-Molecular Weight Cytokeratins (HMW). Vimentin reactivity is variable. In our case, histologic characteristics with such stains were used to diagnose CDC.

Of all renal neoplasms, CDC has the worst prognosis with most patients eventually developing metastatic disease [4,5]. A majority of our cases initially presented as localized disease, which were completely excised by radical nephrectomy. Unfortunately, a man was revealed lumbar vertebra metastasis when he presented to hospital. The mechanism can occur either by direct invasion or by hematogenous spread, with the former assumed to be the proposed means of metastasis in our patient.

There are no existing guidelines for the treatment of CDC and only a few retrospective studies have investigated the significance of potential treatments [4,5]. The benefit of radical nephrectomy is achieving good oncologic outcome, two cases of us have a long-term survival. Some reports show good response with partial nephrectomy, with patients surviving 24 months without evidence of recurrence or metastasis [6]. Three patients developed metastasis and died in 12 months even after a well-performed nephrectomy. Meticulous surveillance should be made 3 years after the postoperative period. Due to its limited therapeutic options in CDC and urothelial carcinoma, radiotherapy has been used for control of local recurrence but does not prevent tumour progression [7]. Sunitinib, an orally available inhibitor of multiple receptor tyrosine kinases, including vascular endothelial growth factor receptor, platelet-derived growth factor receptor, and others, has shown to have observable oncologic benefit in non-clear cell renal carcinoma, specifically CDC [8]. Hideaki Miyake et al reported a case that the patient was treated with sunitinib rather than systemic chemotherapy following by radical nephrectomy, after 4 courses of sunitinib therapy, metastases to the lungs and left rib decreased by more than 30% compared with findings before sunitinib treatment [9]. Even though it later showed treatment resistance, it still provided us a useful option of treatment. Recent studies have suggested that the combination of gemcitabine and cisplatin provides an objectively good response for patients with CDC [4,5]. Therefore, we wonder that is it effective to use sunitinib combined with gemcitabine and cisplatin to prevent development of resistance in the treatment of CDC? This needs us a further study.

In conclusion, CDC is a rare tumor with a clinically rapid and progressively malignant course. In every case, immunohistochemical profile which was necessary to define this subtype of RCC. Surgery is the first choice of treatment to CDC. It is also important to consider a good oncologic outcome by administering adjuvant therapy, such as chemotherapy, and targeted therapy for a better prognostic outcome.

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