

## CASE SERIES

# Collecting Duct Carcinoma of the Kidney: An Overview of 5 Cases

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### ABSTRACT

**Background:** Collecting duct carcinoma of the kidney is one of the rare malignancies of the kidney but highly aggressive one arising from distal part of the nephron in medulla. Early diagnosis may help in prolonged survival of the patients. **Objective:** To know the clinical, radiological and histopathological features of collecting duct carcinoma helping in early diagnosis and treatment in further patients. **Methods:** An institution based retrospective study of five patients including follow up period of minimum 6 months was performed. **Result:** A total of 5 cases (4 males and 1 female) were analysed with average age of 56.4 years with range from 48 to 65 years. All patients were having flank pain with 2 of them also having history of haematuria. None of the patients were having hypercalcemia. CECT abdomen and pelvis was done in 4 patients while MRI abdomen and pelvis along with Doppler of renal vein with IVC was done in one. Of all the tumours three were solid and rest two were complex solid and cystic with average size of 4.7 cm. In 4 patients tumours were arising from central portion of renal parenchyma while in 1 arising from pelvic-ureteric junction radiologically. Regional lymph nodes were enlarged in 4 patients. Radical nephrectomy was performed in 4 patients while one patient underwent nephroureterectomy with bladder cuff excision. On HPE all were confirmed CDC with regional LNs and renal sinus involvement in 4 cases. 2 cases were showing involvement of renal pelvis along with breach of renal capsule while one was having ureteric extension. On IHC all 5 cases were CK19 +ve, 4 cases were CK7+ve while one was negative. **Conclusion:** CDC of kidney exhibits diverse but some special clinicopathological features which may help in early diagnosis/suspect and early treatment. As it is one of the aggressive malignancies with poor outcome early treatment may prolong the survival..

**Key words:** CDC, Clinical, Radiological, Histopathology, Diagnosis, Hypercalcemia.

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### INTRODUCTION

Renal cell cancer almost accounts for 2 to 3 percent of adult malignancies. The most common renal cell cancer is of clear cell variety and it arises from proximal tubular epithelium. The collecting duct is embryologically derived from the ureteric bud, while the remaining of the kidney develops from metanephric blastema. The ureteric bud also differentiates into different structures like calyces, renal pelvis and ureter. Collecting duct carcinoma is one of the rare malignancies of the kidney but it is highly aggressive one arising from distal part of the nephron in medulla. CDC also called carcinoma of Bellini duct accounts for less than 2 percent of all renal malignancies<sup>1, 2</sup>. Some of other renal tumours arising from distal part of the nephron are chromophobe renal cell carcinoma and oncocytoma. Collecting duct carcinoma is also known as medullary renal carcinoma (generally found in sickle cell

patients) and distal renal tubular carcinoma. CDC clinically appears as a renal mass often accompanied by flank pain and haematuria and is frequently mistaken for RCC or transitional cell carcinoma of the renal pelvis<sup>1, 3, and 4</sup>. It can be identified by histological and immunohistochemical features. Macroscopically CDCs are generally located at the confluence of the medulla and renal pelvis, and show a characteristic gray-white-tan colour with absence of foci of necrosis and haemorrhage<sup>5</sup>. Histologically, CDC presents tubulopapillary morphology, often accompanied by desmoplasia, atypia in collecting ducts, and intratubular spread<sup>5</sup>. These features are rarely seen in RCC. On immunohistochemical analysis CDCs stain positive for high-molecular-weight keratin and lectin, proteins typically expressed in the epithelium of the distal tubules<sup>6, 7</sup> while other renal cell carcinomas express antigens widely

expressed in the cells of the proximal tubules, such as low molecular weight cytokeratin and vimentin<sup>7</sup>.

Being an aggressive tumour with poor prognosis early diagnosis of CDC may help in better treatment decision and prolonged survival. Being one of the rare malignancies of kidney its diagnostic protocol has not been led yet. For better understanding of this malignancy, we studied the clinical, radiological and histopathological including immunohistochemical features of 5 patients in AIIMS Patna in retrospective manner spread over 2016 to 2018.

#### Objective:

To know the clinical, radiological and histopathological features of collecting duct carcinoma helping in early diagnosis and treatment in further patients

#### MATERIALS AND METHODS

An institution based retrospective study of five patients including follow up period of minimum 6 months post surgery was performed. All five patients underwent surgical procedure in the department of General Surgery AIIMS Patna and post procedure histopathology and immunohistochemistry was done in the department of Pathology in the same institute. TNM and clinical staging were conducted based upon the clinicopathological information of the cases and the criteria developed by recent American Joint Committee on Cancer (AJCC). Radical nephrectomy was done in four patients while one underwent radical nephroureterectomy with bladder cuff excision. Post

procedure samples were sent in 10 percent formalin to the department of Pathology for histopathology where samples were stained with haematoxylin and eosin. Post procedure histopathology and immunohistochemistry reports were collected and analysed.

#### RESULT

A total of 5 cases (4 males and 1 female) were analysed with average age of 56.4 years with range from 48 to 65 years. All patients were having flank pain with 2 of them also having history of haematuria. None of the patients were having hypercalcemia. CECT abdomen and pelvis was done in 4 patients while MRI abdomen and pelvis along with Doppler of renal vein with IVC was done in one. Of all the tumours three were solid and rest two were complex solid and cystic with average size of 4.7 cm. In 4 patients tumours were arising from central portion of renal parenchyma while in 1 arising from pelvic –ureteric junction radiologically. Regional lymph nodes were enlarged in 4 patients. Radical nephrectomy was performed in 4 patients while one patient underwent nephroureterectomy with bladder cuff excision. On HPE all were confirmed CDC with regional LNs and renal sinus involvement in 4 cases. 2 cases were showing involvement of renal pelvis along with breach of renal capsule while one was having ureteric extension. On IHC all 5 cases were CK19 +ve, 4 cases were CK7+ve while one was negative.

**Table 1. Different observations of all 5 patients**

Patient no.	sex	Age (years)	Largest diameter (cm)	CECT Enhancement	CECT/MRI Location	IHC CK19/CK7
1	male	60	5.4	weak	central	+/+
2	male	56	4.1	weak	central	+/+
3	male	65	6.2	-	calyceal	+/-
4	female	48	4.3	weak	medullary	+/+
5	male	53	3.5	weak	medullary	+/+

#### DISCUSSION

In present study 80% patients were male while average age of patients was 60.4 years with range from 48-65 years. Tokuda et al<sup>8</sup> reported that the average age of onset was 58 years and male patients accounted for 71.6 percent of the cases. According to Xiangyang Wang et al<sup>9</sup> study average age of the patients was 54 years. Clinical manifestations of CDC reported by Yuxiao Hu et al<sup>10</sup> were consistent with those of more common RCCs, including flank pain, haematuria and palpable mass. In present study all cases were having flank pain while 2 of them were having haematuria too.

Radiological imagings are the main methods for CDC diagnosis. These tumours are generally hypo-vascular with ill-defined border, and having invasions to the renal cortex and renal sinus<sup>11, 12</sup>. Pickhardt et al<sup>13</sup>

described the radiological observations of 17 patients with histopathologically confirmed CDC and found medullary involvement (94%) and an infiltrative appearance (65%). They also found that cystic component (35%) and calcification were frequent within the tumours. An additional study by Yoon et al<sup>14</sup> has reported the largest radiological series of 18 cases. They found that medullary location (94%), weak (69%) and heterogeneous (85%) enhancement, involvement of the renal sinus (94%), infiltrative growth (67%), preserved renal contour (61%) and a cystic component (50%) were CT observations frequently identified in patients with CDC. In same series, regional lymphadenopathy, perinephric stranding, vascular invasion and distant metastases were observed in 56, 56, 28 and 33% of the patients. In present study of all the tumours three were solid

and rest two were complex solid and cystic. Also 4 patients' tumours were arising from central portion of renal parenchyma while in 1 arising from pelvic – ureteric junction radiologically. Regional lymph nodes were enlarged in 4 patients (80%).

The pathological examination is the gold standard for diagnosis of CDC. Immunohistochemical examination of epithelial mesenchymal transition (EMT) biomarkers is important for the determination of the origin and the diagnosis of CDC. According to Li M et al study Cancer cell have positive expressions of CK (AE1/AE3), CK7, CK19, EMA, vimentin, CK34BE12, PNA and ulex europeus agglutinin (UEA), and negative expression of CD10 and CK20<sup>15</sup>. In present study all 5 cases were CK19 +ve, 4 cases were CK7+ve while one was negative on IHC.

## CONCLUSION

CDC of kidney exhibits diverse but some special clinical, radiological and histopathological features which may help in early diagnosis/suspect and early treatment. As it is one of the aggressive malignancies with poor outcome early treatment may prolong the survival.

**Conflict of interest:** None to declare

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