## Case Report-3

# Bellini Duct carcinoma- A Case Report

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**ABSTRACT:**

**Bellini duct carcinoma or collecting duct carcinoma is a highly malignant neoplasm arising from the collecting duct epithelium of the kidney. It is a rare tumour with extremely poor prognosis. We report a 53 year old lady who presented with fever, haematuria and weight loss and was detected to have a renal mass with bone metastasis. She underwent nephrectomy and the histopathology revealed a Bellini duct carcinoma. She succumbed to her illness 4 months later.**

## INTRODUCTION

Bellini duct carcinoma (BDC) or collecting duct carcinoma of the kidney is a distinctive subtype of renal cell carcinoma arising from the collecting duct epithelium of the kidney. It accounts for 0.4% to 2.6% of all renal neoplasms.[1] Generally these tumours follow a very aggressive behavior with extremely poor prognosis. At the time of presentation, BDC is metastatic to regional lymph nodes in approximately 80% of cases, to the lung or adrenal gland in 25%, and to the liver in 20%.[2] Median survival after nephrectomy has been reported to be 22 months.[3] Various treatments have been proposed including radiation, immunotherapy, and chemotherapy;[3,4] with generally disappointing results. Here we report a 53 year old lady with Bellini duct carcinoma, because of its rarity.

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CASE: A 53 year old lady presented with history of irregular fever, decreased weight loss and one episode of haematuria. She was investigated elsewhere and was found to have a right sided renal mass. With a presumptive diagnosis of renal tuberculosis she was started on anti-tubercular drugs. There was no improvement in her symptoms even after one month of anti-tubercular drugs and she started complaining of back pain. As there was no improvement in her symptoms even after 1 month treatment she was referred to us. MRI of the abdomen showed a mass lesion with infiltrative margins measuring 5.5x4.0cm in size involving the mid and lower pole of right kidney. The lesion was iso-intense to renal parenchyma on T1 weighted and relatively hypo intense on T2 weighted images. After I.V. injection, lesion showed poor patchy enhancement. The uninvolved upper half of right kidney showed normal parenchymal signal intensity. There was mild dilatation of pelvic calyceal system due to obstruction of renal pelvis level. A MRI of the spine was done which showed features of bone metastasis. Bone scan showed areas of increased uptake in the multiple dorso lumbar vertebrae. CAT scan chest showed enlarged mediastinal nodes. CAT guided FNAC from the renal mass was suggestive of renal cell carcinoma. She underwent right radical nephrectomy and biopsy of interaortocaval lymph nodes. Gross specimen showed a poorly circumscribed grey white firm tumour measuring 4.5x 4 x 3 cm with irregular infiltrative border and early central necrosis located in the medulla extending to renal cortex, renal pelvis and perinephric fat (Fig. 1). Microscopically the viable area of the main infiltrative tumour consisted predominantly of highly irregular duct and tubular structure lined by single layer of cuboidal to low-columnar cells. They had high

grade pleomorphic nucleolus, thick nuclear membrane and small to moderate amount of eosinophilic cytoplasm (Fig. 2). The stroma showed marked desmoplasia with interdigitating invasion of tumour cells. A small foci of in situ dysplasia was seen adjacent to the tumour (Fig. 3) Tumour involvement was present in the capsule, renal pelvis, perinephric fat, and all three interaortocaval lymph nodes.

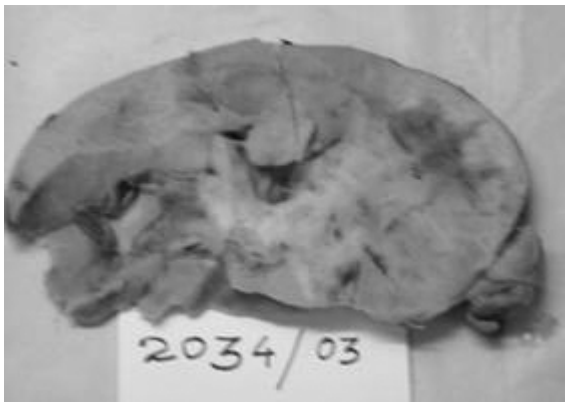


Figure1 Gross specimen showing poorly circumscribed infiltrative grey white tumour with irregular border located in the medulla extending to renal cortex

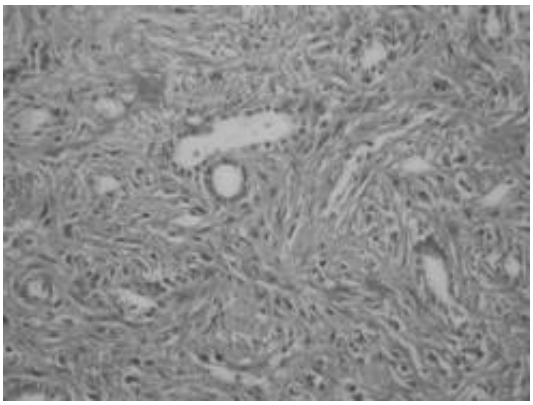

Figure.2 Highly irregular duct and tubular structure lined by single layer of cuboidal to low-columnar cells. The cells show high grade pleomorphic nucleous, thick nuclear membrane and small to moderate amount of eosinophilic cytoplasm.

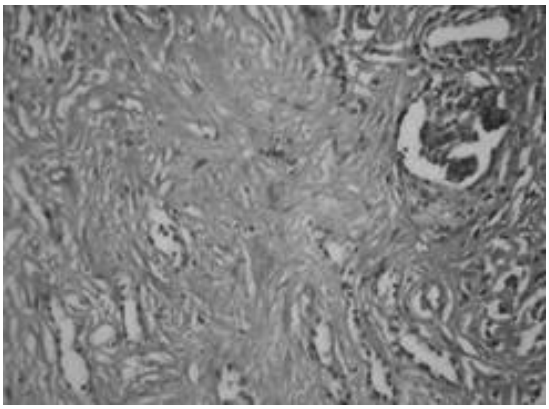

Figure.3 Stroma showing marked desmoplasia with inerdigitating invasion of tumour cells.

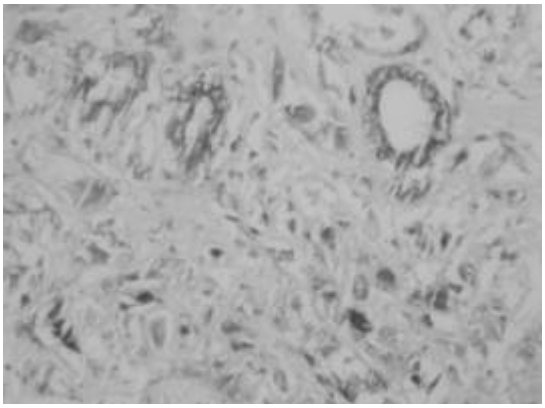

Figure.4 Immunohistochemistry showing reactivity of malignant cells for CK-19 (High molecular weight cytokeratin)

Immunohistochemical studies revealed that the tumour cells were positive for pancytokeratin, cytokeratin19 (CK19), and vimentin (Fig. 4). Based on these findings she was diagnosed to have collecting duct carcinoma kidney. The case was staged as pT3aN2M1. In view of the metastatic disease she was given combination chemotherapy with paclitaxel and carboplatin. However, her tolerance to chemotherapy was poor and hence chemotherapy was discontinued after one course. She succumbed to progressive disease 4 months later.

## DISCUSSION

Bellini duct carcinoma (BDC) predominantly affects men, and a majority of patients are white. The average age of reported patients is approximately 53 years.[1,3] These tumours are usually located in the medulla or central part of

**Table.1**

| Author (Year) (Ref) | No of cases | Drugs used | Response |
|---|---|---|---|
| Dimopoulos (1993)[2] | 6 | IFN/IL-2 | PR-1 |
| | | | SD-2 |
| Peyromaure (2003)[12] | 1 | IFN | PR |
| Dimopoulos (1993)[2] | 1 | 5-FU, MMC, IFN | SD |
| Gollob (2001)[13] | 1 | Paclitaxel/carboplatin | PR |
| Milowsky (2002)[14] | 1 | Gemcitabine/adr | PR |
| Peyromaure (2003)[12] | 2 | Gemcitabine/cisplatin | CR |

IFN- Interferon, IL-2 interleukin-2, 5FU- 5-fluouracil, MMC- Mitomycin-C, Adr-Adriamycin, CR- complete response, PR- partial response, SD- stable disease

the kidney. They are white and gray and histologically show a tubular and papillary pattern. BDC generally pursues a more aggressive course than conventional renal cell carcinoma.[1,4] The prognosis is poor, as more than 50% of the reported patients die within two years of presentation.[2,4,5] Metastases to regional lymph nodes, bone, adrenal glands, lung, skin and meninges have been reported.[3,5]

The usefulness of FNAC or voided urine cytology is limited and controversial.[6] Fleming and Lewi[7] defined the diagnostic criteria and established collecting duct carcinoma of the kidney as a separate histological entity arising in the renal medulla. BDC is divided into two histological types.[8] The papillary type is grayish-white in colour with a yellow tinge, and microscopically the tumour cells demonstrates an eosinophilic cytoplasm, nuclear polymorphism, distinct nucleoli with a papillary or tubulopapillary structure and a resemblance to the distal collecting duct epithelium[5,9]. Therefore, if the tumour is small, it will be located in the renal medulla only as a nodular mass. On the other hand, the mixed type is grayish-white in color macroscopically, with microscopic features of tubulopapillary adenocarcinoma and areas of transitional cell differentiation.[5,9] The tumour cells shows a ring-shaped arrangement mimicking the mesonephric duct. Staining for high molecular weight cytokeratins and UEA-1 gives strong support to a collecting duct origin in the distal nephron.[9] Cytogenetic abnormalities like hypodiploidy, numerical and structural aberrations of chromosomes 1 and X or Y and high frequency of loss of hetero have been reported in BDC[10]. Radiological features of BDC are similar to those of other types of renal tumours.

There is no optimal treatment for BDC. For metastatic collecting duct carcinoma radical nephrectomy alone does not seem to be useful except in a palliative setting.[11] Radiation therapy has been found to be of no use. In one report, radiotherapy was used in patients with local recurrence after nephrectomy for non-metastatic tumour, but this treatment showed no response.[2]

The efficacy of immunotherapy is also debated. Dimopoulos et al.[2] reported one partial response and 2 stable disease out of six patients treated with a combination of IL-2 and interferon-a. Peyromaure *et al.,*[12] reported one patient who had a partial response to interferon lasting for 6 months.

Several combination chemotherapy regimens have also been investigated in single

case reports with varying responses (Table.1). In the series of Dimopoulos *et al.*,[2] one patient achieved stable disease lasting for 16 months. Gollob *et al.*,[13] described a patient with metastatic collecting duct carcinoma who achieved an 80% reduction in her tumour burden, including complete regression of lymph node metastases and significant shrinkage of a renal mass. Later she underwent nephrectomy and was disease-free for 20 months. Immunohistologic and molecular analysis indicate that BDC more closely resembles transitional cell carcinoma than renal cell carcinoma. As the combination of gemcitabine and cisplatin has been demonstrated to be effective in transitional cell carcinoma (TCC) it might work in BDC as well. Recently, Milowsky *et al.*,[14] reported a patient treated with doxorubicin and gemcitabine for BDC with regional and left supraclavicular nodal involvement. These authors noted a partial response after 3 cycles; however, the patient experienced disease progression after[6] cycles. Peyromaure *et al.*,[12] reported complete response in two patients with metastatic BDC using a combination of gemcitabine and cisplatin. One patient remained free of disease 27 months after nephrectomy, and the other patient achieved complete response after 3 cycles lasting 9 months.

In conclusion, BDC is a rare tumour with a clinically rapid and progressively malignant course. This makes it important to determine the origin of all renal tumours and to distinguish it from common types of renal cell carcinoma because it follows an aggressive course and respond poorly to immunotherapy.

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