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Rare Tumors of Kidney

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ABSTRACT

INTRODUCTION: Collecting duct carcinoma (CDC) is an extremely rare type of renal epithelial tumor that arises from the distal convoluted tubule of kidney. Prognosis of CDC is poor due to its rapid growth and widespread metastasis. It is often difficult to distinguish CDC from pelvic urothelial carcinoma and high grade papillary Renal Cell Carcinoma. Mucinous tubular and spindle cell carcinoma (MTSCC) of kidney is also a rare but low grade epithelial neoplasm. It is often misdiagnosed as sarcomatoid renal cell carcinoma, papillary renal cell carcinoma or other aggressive renal neoplasms.

MATERIALS AND METHODS: This is a study of two extremely rare cases of kidney tumors conducted in a tertiary care hospital. We studied clinical presentation, location within the kidney, radiological findings, histopathological findings, immunohistochemical findings and treatment provided.

CASE REPORT: A case of CDC involving the renal medulla extending up to the cortex of upper pole of right kidney and another case of MTSCC involving the middle and upper part of left kidney have been studied. Patient with CDC was a 71 year old male presented with gross hematuria, backpain, fatigue and weight loss. MTSCC was incidentally detected in a 62 year old female. Histopathological examination of the formalin fixed paraffin embedded tissue sections were done for the diagnosis of these two rare entities. Immunohistochemistry was done for exclusion of other diagnostic entities.

CONCLUSION: CDC is a rare aggressive renal neoplasm which is commonly associated with nodal and distant metastasis at the time of presentation. Early diagnosis of this tumor is essential to improve the survival of the patients. Immunohistochemical analysis helps to differentiate CDC from other renal cell carcinoma (RCC) subtypes. MTSCC is a low grade hypovascular renal tumor. The diagnosis of MTSCC fundamentally depends on histopathological examination (HPE) findings. Surgical resection of the tumor is essential for long term survival.

KEYWORDS: Collecting duct carcinoma, hematuria, histopathological examination, immunohistochemistry, laparoscopic nephrectomy, renal cell carcinoma.

INTRODUCTION

Collecting duct carcinoma (CDC), previously known as carcinoma of the collecting ducts of Belini, was first acknowledged by Fleming and Lewi in 1986 as a distinct subtype of Renal Cell Carcinoma RCC [1]. CDC is a malignant epithelial neoplasm arising from principal cells of collecting ducts of Bellini of kidney. It accounts for less than 1% of renal tumor [2]. The incidence of (RCC is increasing throughout the world. CDC is extremely rare and often misdiagnosed [3,4,5,6]. CDC is an aggressive RCC with bad prognosis and continuously evolving diagnostic features, with typical but not entirely distinct histologic and immunohistochemical characteristics [7,8]. Despite histological and immunohistochemical characteristics of CDC, it is often difficult to distinguish CDC from pelvic urothelial carcinoma, high grade papillary RCC due to diverse features of CDC [9]. CDC is usually found in or proximal to the renal pelvis and appears grey or white without substantial necrosis or hemorrhage [10]. Men are more commonly affected than females with wide age range between 13 and 87 years [2].

Mucinous tubular and spindle cell carcinoma (MTSCC) is a rare type of RCC, which is included in the 2022 WHO classification as one of the 20 subtypes of RCC [11]. It is a low grade tumor composed of a mixture of tubules and spindle cell components with a variable amounts of extracellular mucin in the stroma [12,13]. Less than 100 cases of MTSCC have been reported till date [12]. Its origin is still doubted, either originates from distal nephron or proximal tubule as a variant of papillary RCC [12]. MTSCC is more common in females of wide age range [2,12]. Majority of them are detected incidentally. This low grade neoplasm is often misdiagnosed as sarcomatoid RCC, papillary RCC. or other highly aggressive tumors of kidney [2].

MATERIALS AND METHODS

This is a prospective study conducted in a tertiary care hospital comprising of two extremely rare cases, involving the kidney - collecting duct carcinoma and mucinous tubular and spindle cell carcinoma.

CDC was found in a 71 year old male involving the medulla and adjacent cortex of upper pole of right kidney.

MTSCC was found in a 62 year old female involving the middle and upper part of left kidney. Histological and immunohistochemical analysis was performed on formalin fixed paraffin embedded tissue samples. Various IHC were done to come to a diagnosis.

CASE 1: Collecting Duct Carcinoma (CDC) of Right Kidney

A 71-year-old man was admitted to the surgery department for the evaluation of back pain, gross hematuria for 1 month. He had a significant weight loss associated with fatigue.

Contrast enhanced computed tomography (CECT) of abdomen and pelvis revealed a solid, hypoattenuating poorly demarcated tumor measuring 3x5 cm with minimal contrast enhancement occupying medulla and cortex of upper pole of right kidney along with a subcapsular well demarcated satellite lesion measuring 1x2.5 cm.

Based on the clinical and radiological findings, a laparoscopic radical resection of the right kidney was performed along with large hilar lymphadenectomy. Both the specimens were sent to the department of Pathology. Macroscopic examination revealed one specimen of right kidney with a 5x4x3.5 cm poorly demarcated greyish-white friable soft tissue mass with areas of necrosis, occupying a portion of upper pole (Fig.1). The tumor was found to invade the renal pelvis and a well demarcated whitish subcapsular satellite lesion was found measuring 2.2x2x1 cm. One large hilar lymph

node was also received measuring 3x2x1 cm. Histopathological examination (HPE) of the tumor displayed a variety of architectural patterns which included solid sheets, cords, nests, tubule-papillary, intra-cystic papillary patterns. Individual tumor cells tended to be cuboidal with eosinophilic cytoplasm and highly pleomorphic nuclei with prominent nucleoli (Fig.2-9). A columnar and hobnail configuration was also seen. Stroma showed desmoplastic changes. Mitosis including atypical mitotic figures were also found. There was presence of hemorrhage, necrosis along with areas showing sarcomatoid differentiation. The hilar lymph node showed reactive changes.

Immunohistochemistry (IHC) revealed that the tumor cells were positive for EMA (Fig.10), CK7(Fig.11), with focal positivity for Vimentin (Fig.12). However, they were negative for CK20, CD10, p63.

Based on the histopathological and immunohistochemical findings a diagnosis of CDC was established.

The patient was discharged 10 days after surgery and a repeat CT scan was performed 6 months after surgery which revealed no metastasis or recurrence.

CASE 2: Mucinous Tubular and Spindle Cell Carcinoma (MTSCC) of Left Kidney

A 62-year-old female patient attended the surgery OPD with chief complaints of dyspepsia, indigestion and intermittent pain in the epigastrium since 6 months. She was advised ultrasonography (USG) whole abdomen which revealed chronic cholecystitis and also detected a large (9x7) cm well demarcated heterogenous hypoechoic solid mass occupying the middle and upper part of the left kidney. Hence, the kidney tumor was incidentally detected, in absence of any symptoms related to the kidney. Following detection of left kidney tumor on USG whole abdomen, the patient was advised Contrast Enhanced MRI (CEMRI) of abdomen and pelvis which showed (97x74x70) mm left renal mass occupying the middle and upper part of left kidney with mixed iso-density and slightly longer T_1 and T_2 signal with moderate enhancement and clear boundary.

The kidney tumor was managed first and she underwent laparoscopic radical nephrectomy. Macroscopic examination revealed one (10x9x7) cm greyish white well demarcated mass occupying almost the entire left kidney. HPE revealed tightly packed elongated and anastomosing tubules lined by low grade cuboidal cells merging with bland spindle cells in a myxoid stroma (Fig.13-15) containing basophilic extracellular mucin which was highlighted by alcian blue (Fig.16). Individual tumor cells had eosinophilic cytoplasm, prominent nucleoli and rare mitosis. IHC showed CK7 positivity, CK8/18 positivity, EMA positivity and vimentin positivity. Integrating the histological and immunostaining results, a diagnosis of MTSCC was made.

The patient recovered well post surgery and did not receive any adjuvant therapy and was scheduled for laparoscopic cholecystectomy 1 month after laparoscopic radical nephrectomy. The patient is doing well for last one year.

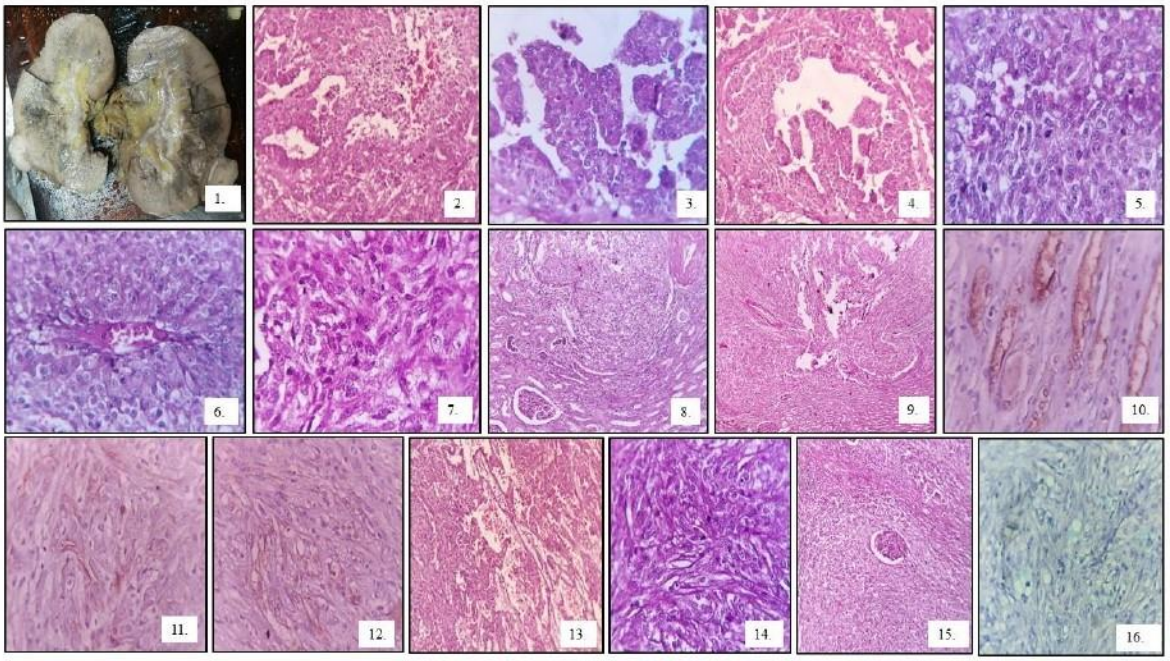

Figure 1: Gross picture of CDC of right kidney showing a poorly demarcated friable greyish white mass with areas of necrosis occupying upper pole with invasion of renal pelvis and a well demarcated whitish subcapsular satellite lesion.
Figure 2: H&E(100X) CDC tumor cells arranged in the form of tubules.
Figure 3: H&E (400X) CDC showing tumor cells arranged in intra cystic papillary pattern.
Figure 4: H&E (100X) CDC showing tumor cells arranged in intra cystic papillary pattern.
Figure 5: H&E (400X) CDC showing tumor cells arranged in sheets.
Figure 6: H&E (400X) CDC showing tumor cells arranged in papillary pattern.
Figure 7: H&E (400X) CDC showing sarcomatoid differentiation.
Figure 8: H&E (100X) CDC showing tumor cells infiltrating in between the normal appearing renal tubules.
Figure 9: H&E (100X) CDC showing tumor cells involving the renal medulla.
Figure 10: IHC (400X) Positive expression of EMA in the tumor cells in CDC of right kidney.
Figure 11: IHC (400X) Positive expression of CK 7 in the tumor cells in CDC of right kidney.
Figure 12: IHC (400X) Focal vimentin positivity in the tumor cells in CDC of right kidney.
Figure 13: H&E (100X) MTSCC showing tightly packed elongated and anastomosing tubules lined by low grade cuboidal cells merging with bland spindle cells in a myxoid stroma containing basophilic extracellular mucin.
Figure 14: H&E (400X) MTSCC showing bland looking spindle cells.
Figure 15: H&E (100X) MTSCC showing tumor cells infiltrating around a glomerulus.
Figure 16: H&E (400X) MTSCC showing myxoid stroma containing basophilic extracellular mucin highlighted by Alcian blue.

DISCUSSION

CDC is a rare type of RCC, which metastases early and has high mortality rates [14,15]. It accounts for less than 1% of all renal tumors. CDC is more common in adult males aged between 41 years and 71 years with male to female ratio of 2:1[2,16]. In our case, the patient was male aged 71 year. He had complaints of hematuria for one month only and diagnosed with a tumor of 5 cm.

Diameter. Majority of the patients with CDC presents with symptoms at the time of diagnosis. Most common symptom is gross hematuria followed by backpain and fatigue. Other symptoms include weight loss, palpable flank mass, constitutional symptoms [3,6], our patient presented with gross hematuria, backpain, fatigue and weight loss. Grossly, CDCs are predominantly located in the renal medulla, however larger tumors secondarily involve cortex [8,17], similar involvement seen in our case. CDC is commonly occurring in Right kidneys, which was seen in our case [17]. Cut section of these tumors are greyish-white or tan-white with irregular borders and variable amounts of hemorrhage and necrosis. Satellite lesions can be identified, usually in the subcapsular area, which was also found in our case. Microscopically, CDCs show a variety of architectural patterns which includes solid sheets, cords, nests, tubules, tubule-papillary patterns and intra-cystic papillary patterns. Desmoplastic stroma is present in all cases of CDC and is one of the diagnostic criteria [17,18], Our case also had similar findings. Individual tumor cells tend to be cuboidal with pale eosinophilic to clear cytoplasm, highly pleomorphic nuclei with prominent nucleoli, some cells exhibit columnar to hobnail configuration, mitosis including atypical mitotic figures are readily seen. Hemorrhage and necrosis are also seen [2,17,18]. Almost similar histopathological features were found in our case with CDC. Once CDC is diagnosed by HPE, ancillary techniques including IHC is done to exclude other diagnostic entities. CDC is positive for PAX8, High molecular weight cytokeratins and CK7, in our case CDC tumor cells were found to be positive for EMA, CK7 and vimentin. CDC is a very aggressive form of RCC with median survival ranging from 10 to 13 months [4,6,17,19]. Confined CDCs are less common, however isolated cases have been reported that have shown long term survival [20,21]. In our case, the patient did well post surgery and there was no history of recurrence or metastasis during the 6 months follow up.

MTSCC is a rare subtype of RCC which according to WHO [2] accounts for less than 1% of RCCs. Studies reveal that MTSCC is more common in females of broad age range [2,22,23], as seen in our case. MTSCC patients are usually asymptomatic or presents with incidental symptoms such as hematuria or flank pain [24,25], however in our case it was incidentally detected while doing investigations for chronic cholecystitis. Abdominal and/or pelvic CT or MRI with or without contrast is strongly preferred. CEMRI was done in our case. HPE reveals MTSCC mainly comprises of tubules lined by cuboidal epithelium and spindle cells within varying amounts of mucinous stroma [12,13]. Cellular origin of MTSCC is still debated, majority of the scholars believe that MTSCC arises from distal nephron, collecting ducts while some scholars believe it to originate from proximal nephron [12]. IHC reveals that this tumor is positive for the distal nephron markers (CK7, EMA) while negative for the proximal nephron markers (CD10). The case we presented here showed CK7, CK8/18, EMA and vimentin positivity in the tumor cells.

Surgical resection is considered as the mainstay of treatment, our patient underwent laparoscopic radical nephrectomy. Prognosis after surgery is generally good without any adjuvant treatment [26]. Our patient is also doing well after one year of surgical treatment.

CONCLUSION

CDC is a rare aggressive renal neoplasm which is commonly associated with nodal and distant metastasis at presentation. As CDC has poor prognosis, early diagnosis of this tumor is essential to improve survival of the patients. Management of CDC needs multidisciplinary team approach by surgeons, pathologists, medical oncologists and radiation oncologists. Immunohistochemical

analysis may provide adequate and reliable data to differentiate CDC from other RCC subtypes. MTSCC is a low grade hypovascular renal tumor which is also an extremely rare entity. The diagnosis of MTSCC depends on HPE. Surgical resection of the tumor is essential for long term survival.

Declaration by Authors

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