A huge collecting duct carcinoma of the kidney in an elderly woman: Diagnosis and management challenge (uncommon condition)

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ABSTRACT

Collecting duct carcinoma, also known as Bellini duct carcinoma (BDC) is a rare type of renal tumor, arising from the distal collecting ducts. The prognosis of this disease is extremely poor due to its rapid progression with widespread metastasis. The present study reported a case of CDC involving the left renal region of a 68-year-old female patient. CT scan showed a huge mass occupying the lower portion of the left kidney. The patient underwent enlarged nephrectomy. Anatomopathological examination showed collecting duct carcinoma of the kidney. Patient’s evolution was exceptionally favorable: no recurrence, no locoregional metastasis and no distant metastasis.

Introduction

Collecting duct carcinoma (CDC) is a rare subtype of renal epithelial neoplasm, accounting for <2% of all the renal cell carcinoma (RCC) cases.1 This aggressive malignancy is considered to be derived from the collecting duct of the kidney and has a poor prognosis in the majority of patients with a median survival time of only 30 months following nephrectomy2

Various treatments have been reported in the literature, including immunotherapy, radiation therapy, chemotherapy and radical excision. In order to improve the understanding on the biological behavior of CDC, the present study reported the case of a 68-year-old female patient with typical pathological features of CDC.3 In addition, the clinical, pathological and immunohistochemical aspects of the disease were reviewed.

Case report

Ms. F.T., 68 years old patient otherwise healthy, presented progressive painless of the left flank (since 3 years), without fever or haematuria, worse performances status (PS = 2) and with quality of life involvement. Clinical examination showed left flank mass. The rest of general examination was otherwise normal.

The ultrasonography showed a left retro peritoneal mass, pushing back the spleen. The CT scan was consistent with 12 cm mass at the lower part of the left kidney, with significant and heterogeneous enhancement and lympho-node involvement in the renal pedicle (Fig. 1).

Intra-operative exploration before enlarged Nephrectomy had showed lympho-node involvement we have performed a limited lympho-node dissection in the perihilar region along left renal vein. (Fig. 2), without any clavien dindo complications (see Fig. 3).

On Histological Macroscopic examination a lower and median polar tumor measuring 12 × 9 × 11 cm, with capsule and kidney fat invasion. 4 lymph nodes were found. They measure from 3 mm to 10 mm. Only one of them is infiltrated without extranodal capsule involvement.

On microscopic examination, round to polygonal neoplastic cells with prominent nucleoli and abundant eosinophilic cytoplasm arranged in sheets and tubulopapillary pattern with marked desmoplastic reaction was seen along with areas of necrosis and hemorrhage.

Immuno-histochemical analysis was positive for high molecular weight cytokeratin (IHC)-CK5/6, Pancytokeratin and Vimentin but negative for CD10, CD117 and this established the diagnosis.

The final pathologic diagnosis was left renal Collecting Duct Carcinoma (BELLINI TUMOR), pT3N1. Because of the histological similarities with urothelial tumors, the patient benefited from 3 chemotherapy cures based on cisplatin and gemcitabine with very good tolerance, the CT the control by CT scanner performed every 6 months for 4 years does not show a local or metastatic recurrence. The Recurrence Free survival is exceptionally favorable.

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Discussion

CDC mostly affects younger patients and often presents with non-specific features such as haematuria, flank pain, a palpable abdominal mass or distant metastasis. Our patient was an elderly female with alteration of performance status, no haematuria, and regional lymphadenopathy.

Microscopically, the common features include a tubulopapillary architecture, atypical hyperplastic changes, clear cytoplasm, evident stromal reaction with fiber hyperplasia and detached single cells with a hobnail surface.

In the present case, the results of the microscopic examination revealed certain characteristic features of CDC, including a tubulopapillary growth pattern, significant pleomorphism of the neoplastic cells. Positive immunohistochemical staining for high molecular weight cytokeratin (IHC)-CK5/6, Pancytokeratin and Vimentin.

Radical nephrectomy and regional lymphadenectomy is commonly adopted in the management of CDC. These tumors exhibit highly aggressive behavior and tendency for early distant metastasis with a median survival of only 12 months even after surgical extirpation.

At present, GC (gemcitabin cisplatin) regimen is considered as a first-line systemic treatment in metastatic CDC as no other specific chemotherapy agent has demonstrated a beneficial effect. GC regimen is advocated as CDC has a mesonephric origin.
In the present study, left nephrectomy was performed without chemotherapy or immunotherapy. No recurrence or metastasis was observed 7 months after surgery; however, further follow-up is required.

Conclusion

CDC is a rare, aggressive renal tumor that is frequently associated with nodal and visceral metastases at presentation. Since patients with CDC often present a poor prognosis, the early detection and diagnosis of this disease are vital. Newer therapeutic modalities need to be explored and developed for treatment and improving the prognostic outcome of the patients with this rare tumor.

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Fig. 3. Histological features of the Collecting duct carcinoma.