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Carcinoma of Bellini's Duct: A Case Treated at the Mohammed VI University Hospital Centre, Marrakesh

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Abstract **Case Report**

Bellini duct carcinoma is a very rare and aggressive histological type of renal cell carcinoma. It accounts for less than 1% of renal cancers. It originates in the epithelial layer of the Bellini duct, which is the distal part of the nephron, with a median survival time of less than one year in the metastatic stage. We report the case of a 69-year-old patient admitted for a painless swelling of the right flank, which had developed gradually. Computed tomography revealed a large mass in the right middle kidney. The patient underwent an extended right nephrectomy. Histopathological examination coupled with immunohistochemical analysis revealed carcinoma of the renal collecting ducts. Positron emission tomography (FDG PET scan) did not reveal any hypermetabolic foci associated with lymph node or metastatic involvement. The patient did not receive adjuvant chemotherapy and was placed under surveillance. The outcome is exceptionally favourable 9 months after surgery: no recurrence, locoregional metastases or distant metastases.

Keywords: Renal cell carcinoma, Bellini collecting ducts, collecting duct carcinoma, poor prognosis.

INTRODUCTION

Bellini duct carcinoma is a very rare and aggressive histological type of renal cell carcinoma. It accounts for less than 1% of renal cancers [1]. Unlike other clear cell renal carcinomas, which represent the majority of renal cancers and develop in the renal cortex, Bellini duct carcinoma develops in the medulla. It originates in the epithelial layer of the Bellini duct, which is the distal part of the nephron, and appears histologically closer to urothelial carcinomas than to renal carcinomas [2].

Collecting duct carcinoma is a very aggressive disease, often associated with a poor prognosis and diagnosed at an advanced stage (>T2), with a median survival time at the metastatic stage of less than one year [3]. Approximately 60% of patients will present with lymph node invasion or distant metastases, preferentially in the bone, lung and liver [4].

Collecting duct carcinoma is not very responsive to conventional treatments used for clear cell renal cell carcinoma. Currently, there are very few studies evaluating their optimal management. Treatment of localised disease is based on total or partial nephrectomy surgery when technically feasible. Despite optimal surgical management, recurrences are common. It should be noted, however, that prolonged remissions have been observed after surgical treatment alone in patients with localised disease. In these patients, overall survival is identical to that of patients with clear cell renal carcinoma, which is 122.5 months (121.0–123.9) [2]. There is currently no argument for neoadjuvant or adjuvant chemotherapy. In the metastatic stage, platinum-based polychemotherapy remains the most commonly used strategy. Early diagnosis and optimal surgical management could improve the prognosis of this disease [2].

OBSERVATION

Our 69-year-old patient, who had been diabetic for five years and was on ADO, consulted in December 2024 for a painless swelling on his right side, which had developed gradually. The onset of symptoms dated back six years, with the gradual appearance of right lumbar pain without fever or haematuria, all occurring in a context of stable general health. Clinical examination

revealed a lumbar lump on the right side, which was not painful on palpation.

Hydatid serology was negative, and renal function was impaired with creatinine levels at 14.64 mg/L and creatinine clearance at 50.6 ml/min.

The CT scan revealed a right mid-renal mass measuring 9x8x7cm associated with a right renal hilum lymph node measuring 7mm in the short axis. Two small simple left renal cysts classified as Bosniak stage I were also noted (Figure 1).

The patient underwent an extended total nephrectomy and had an uneventful postoperative course.

The pathological examination revealed a renal parenchyma largely dissociated by an infiltrating carcinomatous proliferation measuring 6 cm. This was arranged in papillae of varying sizes, diffuse layers and diffuse nests. The carcinoma cells were medium to large in size, with enlarged nuclei, slightly irregular contours, hyperchromic in places with vesicular chromatin, and nucleoli visible at high magnification. The cytoplasm was abundant, clarified, eosinophilic and granular elsewhere, with more or less well-defined cytoplasmic borders. The stromal reaction is fibrous, sparse, furrowed by blood vessels with clean walls, sometimes hyalinised and with regular endothelium, with the presence of vascular emboli. Presence of foci of tumour necrosis estimated at 10%. This tumour proliferation infiltrates the pyelocaliceal cavities and invades the renal capsule in places without exceeding it.

No infiltration of perirenal fat, sarcomatoid or rhabdoid components, or hilar lymph nodes. The ureteral resection margin is within healthy tissue, as are the vascular resection margins (renal artery and vein). In immunohistochemical studies, the tumour cells did not express the following antigens: anti-CD10 antibodies, anti-CD117 antibodies, P504S (Racemase) antibodies. They expressed anti-CK7 antibodies. In conclusion, the pathological examination revealed a 6 cm carcinoma of the renal collecting ducts, infiltrating the pyelocaliceal cavities and focally invading the renal capsule without extending beyond it, and the presence of intratumoral vascular emboli. There was no infiltration of the perirenal fat, no sarcomatoid or rhabdoid component, and no hilar lymph node involvement (pT1bN0Mx).

A staging assessment using thoracoabdominal-pelvic computed tomography revealed bilateral pulmonary nodules and micronodules, the largest of which measured 10 mm in diameter and was located in the dorsal segment of the right upper lobe, appearing to be secondary. There was also fatty infiltration in the nephrectomy site with millimetric retrocaval, lombo-aortic and bilateral iliac lymph nodes.

This prompted us to further investigate the extent of the disease using positron emission tomography, which did not reveal any hypermetabolic foci associated with lymph node or metastatic involvement (Figure 2).

Following a multidisciplinary consultation, the patient did not receive adjuvant chemotherapy and was placed under surveillance given the absence of secondary lesions on the PET scan. The outcome is exceptionally favourable nine months after surgery: no recurrence, locoregional metastases or distant metastases.

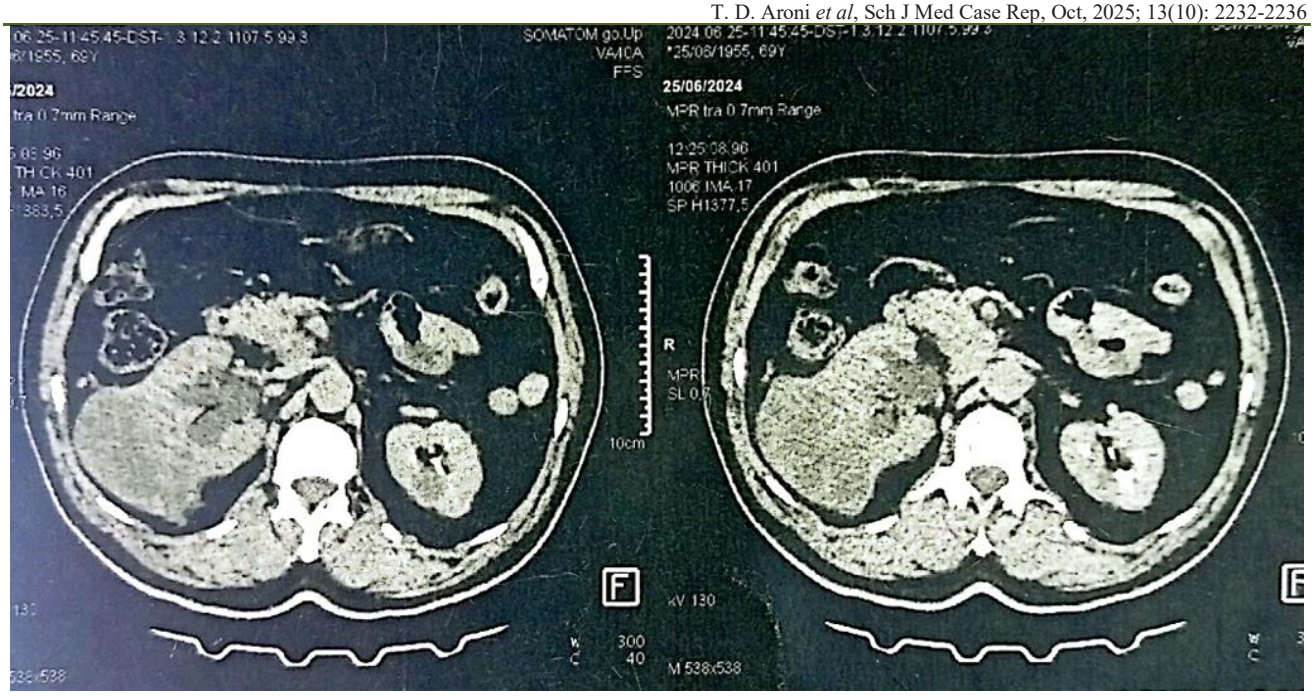

Figure 1: Uroscanner: Right mid-renal mass measuring 9x8x7cm

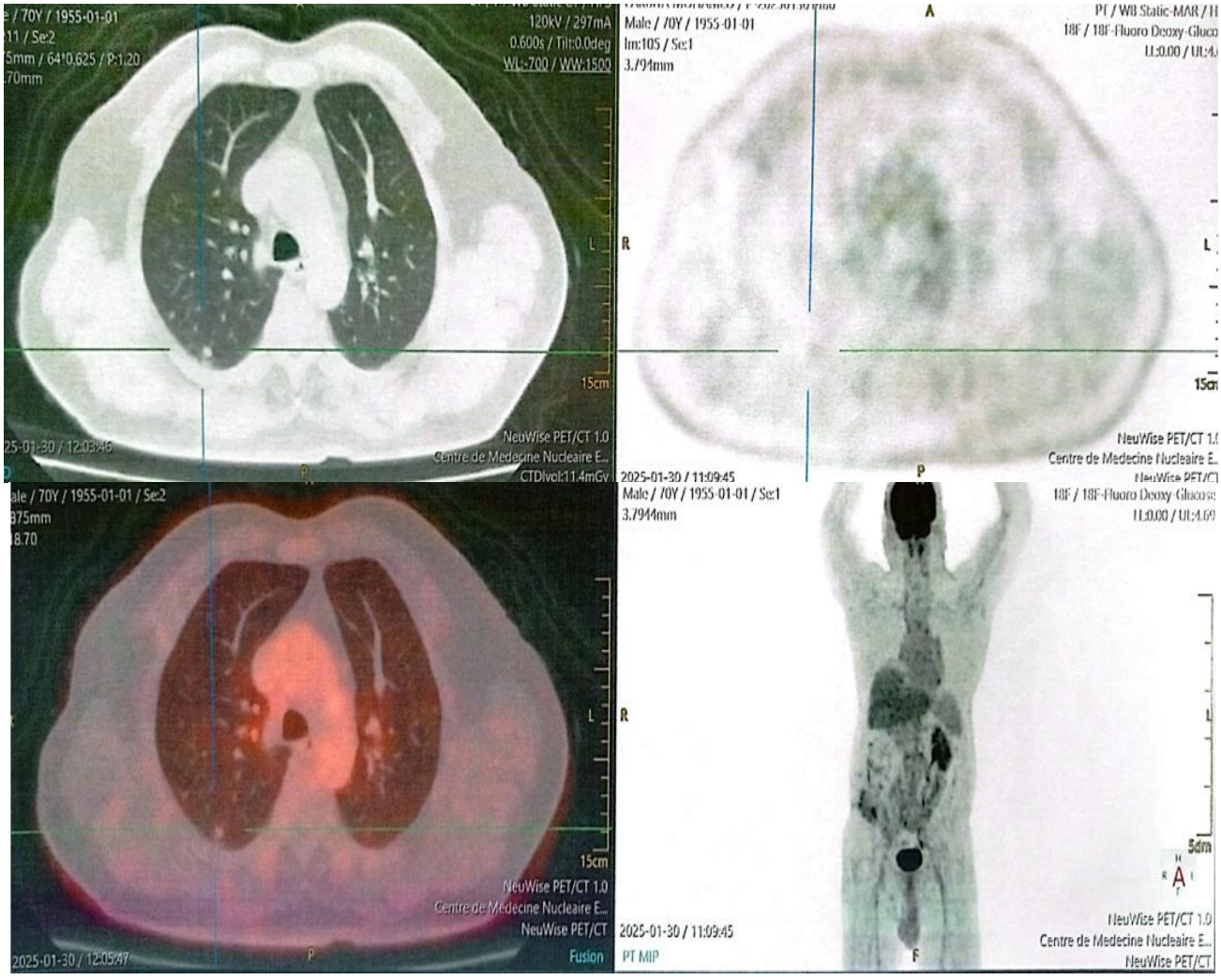

Figure 2: PET scan: No hypermetabolic foci associated with lymph node or metastatic involvement

DISCUSSION

Bellini duct carcinoma is a rare renal tumour, accounting for approximately 1% of epithelial cancers. This entity was first reported in 1976 [5] by Mancilla Jimenez and was adopted by the WHO in 1981. The majority of patients affected are between the 6th and 7th decades of life, as reported in our case (69-year-old patient).

There is a male predominance with a sex ratio of 2:1 [6]. There is no side predominance [6] and no synchronous or metachronous bilateral involvement has been reported. Half of the patients reported by Chao [7] had a family history of cancer. In our patient, no family history of cancer was reported.

Chronic renal failure, currently recognised as a risk factor for renal carcinoma, appears to be less clear-cut for Bellini carcinoma, with only one case having been reported in a haemodialysis patient [8]. Most clear cell renal carcinomas are discovered incidentally. In contrast, Bellini carcinoma is most often symptomatic [3]. The most common symptoms are macroscopic haematuria (50-66.7%), low back pain (40%) and sometimes a palpable mass in the lumbar region or flank [1]. In our patient, the presenting symptom was a painless swelling in the left flank. Sometimes the diagnosis is made at an advanced stage of the disease when there is a deterioration in the patient's general condition with weight loss and lymph node or visceral metastases from the outset (26%). The presence of a vena cava thrombus has been reported in two cases [9]. In 8% of cases, the discovery is incidental during radiological examination [10]. Radiologically, Bellini carcinoma is not specific.

The diagnosis of Bellini carcinoma is anatomopathological. Macroscopically, Bellini carcinoma is most often located in the central region of the kidney, in the parenchyma itself, and is a large tumour. It is firm, yellow or greyish in colour when cut, generally poorly defined [6,9] and often has satellite nodules and haemorrhagic changes. Microscopic analysis reveals a glandular tumour consisting of irregularly shaped tubes and medullary topography. These are most often large cells with eosinophilic cytoplasm and large, highly nucleolar nuclei with a high nuclear grade [9]. For FONDIMARE, the macroscopic appearance associated with a tubular, microcystic and papillary architecture, as well as the cytological appearance, are highly suggestive of the diagnosis [9]. There may also be focal sarcomatoid features [6]. The presence of dysplastic lesions in the collecting ducts near the tumour is also a diagnostic feature [11]. Cytogenetics should provide a better understanding of the molecular events involved in Bellini carcinoma. Unlike renal adenocarcinoma, where chromosomal aberrations mainly affect chromosomes 3, 7 and Y, SCHOENBERG found a loss of heterozygosity in chromosomes 8p and 13q in 50% of a series of 6 Bellini carcinomas [12]. Recent work has focused on evaluating prognostic factors that are more specific to this histological type in order to improve the management of these patients [13].

Total nephrectomy has a place in the treatment strategy for patients, even those with metastases. Adjuvant chemotherapy with gemcitabine and platinum salts, also used in urothelial carcinomas, may be offered while awaiting further evidence on the contribution of targeted therapies [14].

CONCLUSION

Bellini carcinoma is a rare malignant renal tumour with an extremely poor prognosis, which depends mainly on the stage of the disease at the time of diagnosis. Diagnosis is mainly histological, based on immunohistochemical analysis.

The standard treatment is extended total nephrectomy, which can lead to prolonged remission if the disease is localised, and cytoreduction in the metastatic stage is associated with improved overall survival.

The contribution of targeted therapy is currently being evaluated. However, given the rarity of our histological entity, the standard treatment remains total extended nephrectomy, combined with adjuvant chemotherapy using a combination of gemcitabine and platinum salts for metastatic stages.

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