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Bellini Collecting Tube Carcinoma with Hepatic Invasion: Diagnostic and Therapeutic Difficulty: Case Report and Literature Review

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Abstract

We report the case of a 60-year-old patient hospitalized in our training for the management of a renal mass, An ultrasound and an abdominal scanner showed the presence of a process of bulky heterogeneous renal tissue which infiltrates the right lobe of the liver measured at 88X87mm la, we performed a total enlarged right nephrectomy associated with a right hepatic lobectomy. The appearance of cells in "kitten ring" and the tubular appearance in "furniture nail" first evoke a carcinoma of the bellini collecting tubes, an immunohistochemical complement to confirm the diagnosis. Although rare, carcinoma of the collecting tubes poses a problem of differential diagnosis all the more since in our observation there was a massive infiltration of the liver making the diagnosis extremely difficult.

Keywords: Carcinoma; Diagnosis; Nephrectomy

Introduction

Carcinoma of the bellini collecting tubes extremely rare, representing less than 1% of renal tumors. It was first described in 1956 by Masson [1] and it was not until 1981 that this tumor was identified in the classification of renal tumors by the World Health Organization (WHO) as one of the variants of carcinomas with renal cells called "Bellini collecting duct carcinoma" (CCCB). This tumor is of high nuclear grade, generally aggressive and of advanced stage at the time of diagnosis [1,2]. The median survival after enlarged nephrectomy is less than two years. Its low frequency limits the reporting of new clinical or therapeutic data [2]. Only the early diagnosis of Bellini seems to offer a real improvement in survival [3]. Concerning the literature, it is mainly presented in the form of clinical cases [5-7]. This entity poses a problem of differential diagnosis for the pathologist and therapeutic management for the urologist. The diagnosis was more difficult was because there was a hepatic invasion. The goal of this work is to illustrate through an observation the diagnostic and therapeutic difficulty of CCB, Our observation of the CCCB is particular because it is a tumor which posed a problem of preparatory diagnosis between the hepatic origin and renal mass.

Case Report

Our 60-year-old patient consulted for right low back pain associated with episodes of gross hematuria. The physical examination found an altered general state with an alteration in the state of performance and on palpation of the right lumbar fossa a mass giving lumbar contact. Biological evaluation showed an acceleration of the sedimentation rate. hyperleukocytosis, normal hepatic check-up, ultrasound and abdominal scanner have shown the presence of a large heterogeneous renal tissue process with areas of necrosis this process massively infiltrates the right lobe of the liver measured at 88×87 mm, The tumor mass slightly absorbs the contrast agent after the injection, without renal venous thrombus or cellar (Figure 1).

The evaluation of the extension includes a thoracic pelvic abdominal scanner aiming at the presence of a suspect nodule in the dorsal segment of the upper lobe of the right lung whose malignancy is eliminated by a pulmonary biopsy under scanner, the surgical intervention consisted of an enlarged total right nephrectomy with adrenalectomy associated with a right hepatic lobectomy by subcostal route. Macroscopically it is a mass of 875 g including a kidney, an adrenal, a right hepatic lobe and

a gall bladder, it is a white tumor, without cysts, developed from the medulla towards the renal cortex (Figures 2 and 3), the tumor presents under the microscope a mainly tubular or tubulopapillary architecture, with an inflammatory stroma and the aspect of the cells in "kitten ring" and the tubular aspect in "upholstery nail" first arousing a carcinoma of Bellini collecting tubes, an immunphistochemical supplement to confirm the diagnosis, the immunohistochemical study showed a very marked labeling of tumor cells with cytokeratins (Dako) (Figure 4).



Figure 1: Heterogeneous abdominal mass scanner, containing a mid-renal polar tumor right upper kidney with massive liver invasion.

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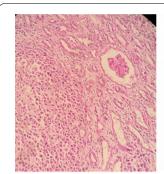




Figure 2: Piece of nephrectomy+right hepatectomy macroscopic postoperative aspect.



Figure 3: Macroscopic appearance of the mass after opening.



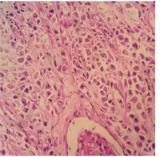


Figure 4: Renal carcinoma of the collecting tubes infiltrating the renal tissue with an abundant fibro-inflammatory stroma reaction.

The tumor cells showed a slightly less intense labeling with the following markers Ulex Européus Agglutinin, Vimentine, EMA and BNH9 when the cytokeratin 20 was negative, and especially a p63-/PAX8+profile. The diagnosis of Bellin Fuhrman grade 4 collecting duct carcinoma associated with massive hepatic invasion was retained. Postoperative follow-ups were simple after the intensive care stay, on the other hand, the thoracic CT at one objective month of metastatic pulmonary lesions leading to a rapid deterioration of the general state as well as the death of the patient within the framework of a multifailure organic.

Discussion

The CCCB is rare, represents approximately 1% of renal tumors [1]. It is defined according to the classification of the WHO as a malignant epithelial tumor which would come from the cells of the collection tubes of Bellini. The clinical signs of CCCB are not specific; it occurs

at an average age of 59 years. A male predominance is noted with a sex ratio of 2/1 [2,4]. There is no side prevalence [5] and no synchronous or metachronic bilateral involvement has been reported. Half of the patients reported by Chao [6] had a family history of cancer. In our patient, no notion of family history of cancer was reported. Bear reports a case of CB associated with a clear cell carcinoma on the contralateral kidney [7], What concerns the mode of discovery of Bellini's carcinoma is most often symptomatic [3,8]. The majority of patients present with an alteration in performance status at the time of diagnosis, with lumbar or abdominal pain or, at the start, an alteration in general condition, characterized first by weight loss [3,8]. The presence of gross hematuria is inconsistent and sometimes never found [3,8], This may be related to intra-parenchymal development, the point of departure of the tumor or its hypovascular character [9]. Whatever the radiological examination, the presence of a single lesion and renal infiltration must in any case refer the diagnosis to the urologist [3]. On ultrasound, there is no particular tumor characteristic. In CT scans (Figures 1 and 2), Bellini's carcinoma is distinguished from other infiltrating renal tumors: lymphoma, metastases, sarcomatoid carcinoma, urothelial tumor with renal extension [3]. Another differential diagnosis is xanthogranulomatous pyelonephritis.

The tumor mass is often slightly increased with the injection of contrast medium, reflecting a relative hypoperfusion of Bellini, with a frequent association of a renal thrombus or a vena cava [10,11]. our observation presents another diagnostic difficulty due to a massive hepatic invasion hence a doubt rather on an aggressive hepatic tumor with right renal invasion, the situation is rarely observed given the rapid and fatal development of the CCCB or the invasion of organ and rarely observed, to our knowledge, this is the first observation of CECC with massive hepatic invasion. No pathognomonic signs have been described. Some authors have performed angiographies of these tumors, highlighting strictly avascular or hypoperfused masses [3,9]. The CCCB is generally located in the medulla then extends to the cortex, to the hilum of the kidney, to the capsule and to the perirenal fat see the neighboring organs, the tumor cells of the CCCB are generally eosinophilic with the characteristic aspect "upholsterer nail" They secrete mucin, the characteristic cellular appearance and the immunohistochemical profile made it possible to retain the diagnosis of CCCB, highlighted with PAS (periodic acid-Schiff), BA (alcian blue) or mucicarmin. There are significant cytonuclear atypies as well as increased mitotic activity. The architecture is very variable tubular and tubulopapillary, pseudo papillary, microcystic or cribriform or even solid. These structures are irregular, angular and embedded in a desmoplastic stroma rich in neutrophils, this poses a problem of pathological diagnosis with other tumors, in particular urothelial carcinoma with glandular flexion hence the interest of the histochemical immune study [12,13]. The CCCB is radio-resistant; the treatment of choice is surgery, an enlarged total nephrectomy. Medical treatments for inoperable metastatic CCEFs use immunotherapy and various chemotherapies, but the results are generally disappointing [14,15]. Our patient treated by enlarged nephrectomy with right hepatic lobectomy died in the postoperative months due to multivariate failure secondary to metastases. The prognosis for carcinomas of the Bellini collecting ducts is generally poor; survival does not exceed 20% at 2

Conclusion

Bellini's carcinoma is a rare renal malignancy, with a poor prognosis because of the frequency of its discovery at the metastatic stage. The histological diagnosis is mainly based on immunphistochemical

analysis. The carcinoma of the collecting tubes poses a problem of differential diagnosis with the transitional carcinoma with glandular inflections whose evolution is more favourable. Histological study and immunohistochemistry allow the diagnosis to be corrected. Extended nephrectomy does not improve the prognosis of this metastatic carcinoma, which is why new multicentre protocols are being evaluated.

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