Bellini duct carcinoma revealed by cutaneous metastasis: A case report

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ABSTRACT
INTRODUCTION: Cutaneous metastasis of renal cell carcinoma is rare and the majority of these metastases are asynchronous. The scalp and face are the major sites of metastases, followed by the chest and abdomen. However, the entire body surface can be affected. When diagnosed, patients are multistatic in 50–80% of cases during follow-up post nephrectomy.

CASE PRESENTATION: We report here a patient who consulted a dermatologist for multiple skin nodules that appeared 3 months prior. A skin biopsy of a nodule was performed and the pathological examination and immunohistochemistry profile confirmed a metastasis of Bellini Carcinoma, which is a renal cell carcinoma of the collecting duct. A thoraco-abdomino-pelvic scan showed a left renal tumor locally advanced with lung and liver metastases. Chemotherapy was indicated and the patient died four months after diagnosis.

DISCUSSION: Bellini carcinoma is a very rare type of carcinoma of renal cell origin with a very poor prognosis as it is diagnosed already at a metastatic stage in the vast majority of cases. After analysis of the data from the literature, our case is the second reported case of a Bellini carcinoma revealed by cutaneous metastases. The peculiarity of our observation is metastases occurred on all four limbs and at the trunk level, and the asymptomatic characteristic of Bellini’s carcinoma, which is a rare situation.

CONCLUSION: The originality of this observation is based on the mode of presentation of a rare renal tumor by an even rarer metastasis of the skin.

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1. Introduction

Bellini Carcinoma, also known as collecting duct carcinoma is a rare tumor characterized by its aggressiveness behavior and has a poor prognosis [1,2]. Most cases are metastatic at the time of diagnosis [1,2]. However, the cutaneous location of metastases is rare and the majority of these metastases are asynchronous. The entire body surface can be affected but the scalp and face are the major sites of metastases, followed by the chest and abdomen [1,2].

We report here the second documented case of Bellini carcinoma revealed by cutaneous metastases. This work has been reported in line with the SCARE criteria [3].

2. Observation

We report a case of a 52-year-old patient with no previous medical history who consulted a dermatologist for multiple skin lesions that have appeared 3 months prior. The patient had not reported any urinary symptom (lower back pain, hematuria…), but he noticed a weight loss with asthenia.

On examination, several cutaneous lesions of varied size and aspect were located on the limbs and trunk (12 lesions in total). At first, the lesions appeared to be hard, infiltrated, and erythematous (Fig. 1), and rapidly became necrotic and infected ulcers (Fig. 2). No abdominal mass was found and the rest of the examination was normal.

A skin biopsy was carried out. Histological examination showed an undifferentiated carcinoma. The diagnosis of multiple cutaneous metastases was suggested and was confirmed by a thoraco-abdomino-pelvic CT scan looking for primary cancer, finding a 52 × 64 mm left renal tumor enhancing at arterial phase contrast. The tumor was described as infiltrating the peri-renal fat with loss of the fatty border within the pancreas, spleen, and the left colic

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3. Discussion

Renal cell carcinoma of the Bellini collecting ducts is included in the 2004 WHO classification of renal epithelial tumors and accounts for less than 1% of renal carcinomas worldwide [2,4].

The mean age of diagnosis is 66 years old with a standard deviation of range of ages from 20 to 80 years [2]. Unlike most renal cell carcinomas that are discovered fortuitously, Bellini’s carcinoma is often symptomatic and most patients have lower back pain and general deterioration [2,4,5].

On the other hand, hematuria may be absent (intra-medullary development of the tumor has a hypovascular characteristic) [6,7]. Our patient had no low back pain nor hematuria, but there was a marked alteration of the general state of health.

This tumor is often aggressive, of high grade and advanced stage at the time of diagnosis and 80% of Bellini carcinoma cases are T3 N+ at the time of diagnosis with an average size of 80 mm [6].

At the time of diagnosis, most patients are metastatic at the regional lymph nodes. This is seen in 80% of the cases, but some cases also have a distant spread with 40% of pulmonary metastases, 30% of liver metastases, and 20% of bone metastases [1,2].

The peculiarity of our observation is twofold. On one hand, our patient had multiple cutaneous metastases on all four limbs and at the trunk level (12 lesions in total) associated with a diffuse ganglionic, hepatic and pulmonary metastases at the time of diagnosis. A single case of Bellini’s carcinoma with cutaneous metastases as the initial clinical manifestation was reported by Grande et al. [1]. The patient was a 38-year-old man presenting a 6 cm left mediastinal tumor with unique cutaneous metastasis on his scalp. On the other hand, the second feature is the asymptomatic characteristic of Bellini’s carcinoma, which is a rare situation.

Histologically, it is a minimally differentiated adenocarcinoma, predominantly of tubular architecture, often tubulo-papillary. However, its histological presentation may vary [4]. The carcinoma of the collecting tubules is a high-grade carcinoma with marked atypia. Therefore, it’s not necessary to specify the Fuhrman grade according to the latest recommendations of the International Society of Urological Pathology [8].

The immunohistochemical study differentiate Bellini duct carcinoma from a high-grade papillary carcinoma, adenocarcinoma

flexure (Fig. 3a). It also showed multiple mediastinal, lateroaoctic, and left renal pedicle lymph nodes metastasis (Fig. 3b) associated with multiple liver and pulmonary metastases.

A second cutaneous biopsy was performed, the histological examination and the immunohistochemistry profile confirmed a metastasis of a renal Bellini carcinoma (Fig. 4).

The patient was in poor general condition, had no hematuria or low back pain. A left nephrectomy was deemed unnecessary to perform and he was instead considered for palliative chemotherapy with gemcitabine-cisplatin.

The patient died after 4 months of diagnosis.
metastasis, with a possible aid of the use of PAX8 and p63 to distinguish it from an intra-renal infiltrating urothelial carcinoma [4].

The morphology of the renal carcinoma of the collecting tubules and the immunohistochemistry phenotype can be very variable, making its diagnosis sometimes difficult, especially on biopsy specimens [3]. It explains the difficulty in diagnosing Bellini carcinoma on the first biopsy in our case.

Bellini carcinoma has what can be considered a very poor prognosis [1,2,6,9]. The majority of patients are metastatic at the time of diagnosis and half of the patients die within the first year [2,6,7]. The overall survival rate, generally reported, is approximately 60% for the first 6 months, 50% for 1 year and 20% for 2 years after diagnosis [6,7]. In metastatic patients the median survival rate is 6 months [2,6].

Nevertheless, there’s an entity that should be differentiated from Bellini’s carcinomas, referred as low-grade tubulocystic carcinoma, since it is has a good prognosis. It presents as a spongy and cystic medullary tumor, containing cysts lined with eosinophilic cells [9].

The treatment of Bellini’s carcinomas must take into account the fact that it is a very aggressive tumor, with a clinical progression similar to urothelial tumors.

For localized Bellini carcinomas, minimal data is available in published literature for its management and there are only isolated cases or a series of cases [11]. Surgery is indicated allowing durable remission for some cases [6,7].

However, most patients are metastatic at the time of diagnosis. Cytoreduction nephrectomy, which has shown its benefits for clear cell carcinomas, does not seem to improve the prognosis [1,11,12]. It should be reserved for patients in good general health condition and very symptomatic (back pain or hematuria) [1,4]. Some authors believe that it is completely useless and can even be dangerous with immediate death due to the alteration of the general state of health often observed in these patients [6,11]. They propose a tumor biopsy to avoid surgery [2,6].

In our case, nephrectomy was not performed because the patient was in poor general health condition upon entry. He had no hematuria nor low back pain. Grande et al. performed a nephrectomy with excision of the cutaneous metastasis but the patient died after 7 months [1].

Adjuvant treatments, immunotherapy and radiotherapy have not shown efficacy in the treatment of Bellini carcinoma [2].

Chemotherapy is relatively inactive, out of 23 patients treated with gemcitabine and cisplatin, the response rate was 26%, with progression-free survival and overall survival of 7.1 months and 10.5 months respectively [10]. There are no published studies on the efficacy of tyrosine kinase inhibitors [9].
Thus, in a metastatic situation, chemotherapy with gemcitabine-platinum salts can be proposed while waiting for evidence of targeted therapies [11].

4. Conclusion

Bellini carcinoma is a very rare type of carcinoma of renal cell origin with a very poor prognosis as it is diagnosed at a metastatic state in the vast majority of cases. After analysis of the data from the literature, our case is the second reported case of a Bellini carcinoma revealed by cutaneous metastases. Extended nephrectomy is found to not improve the prognosis of this metastatic carcinoma and chemotherapy with gemcitabine and cisplatin may be suggested.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Since this was just a case report and patient’s confidentiality was assured, there was no need for ethical approval based on our institution regulations.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

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