Primary Squamous Cell Carcinoma of the Renal Parenchyma: A Rare Case with Review of Literature

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Abstract

Primary renal squamous cell carcinoma (SCC) is an extremely rare primary malignancy of the kidney with only few cases that have been reported in the literature. The indolent course of the disease and the lack of specific clinical and radiological features may be responsible of the delay in the diagnosis leading to an advanced stage of disease at diagnosis with poor outcomes. We present the case of a 65-year-old man with long standing renal stone disease and recurrent pyelonephritis in his history, admitted for management of locally advanced renal tumor with distant metastases where histology of biopsy revealed SCC. We report this case to underline the rarity of the renal location as primary site of squamous cell carcinoma, the clinic-radiological and histopathologic features, in addition to treatment management and prognosis.

Keywords: Calculi; Chemotherapy; Kidney; Prognosis; Squamous Cell Carcinoma

Introduction

Kidney is an extremely rare primary site of Squamous Cell Carcinoma (SCC). It accounts less than 1% of all urinary tract malignancies [1] and it is usually associated with urolithiasis and hydronephrosis. The diagnosis is often delayed because of the absence of specific signs leading to the diagnosis at advanced stages with poor therapeutic results.

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Case Presentation

We report the case of a 65-year-old man. His medical history is marked by active smoking and long standing renal stone disease with recurrent pyelonephritis. He presented 4 months before his admission in our department of medical oncology at Hassan II University Hospital, pain over left flank region associated with important loss of weight (12kg) then complicated by gross hematuria. Physical examination of the abdomen found a painful percussion on the left kidney area. Hematological and biochemical parameters especially renal function tests were normal. Plain radiograph of the abdomen showed a voluminous left renal calculi with hydronephrosis (Figure 1).

Renal CT scan showed a big left kidney with staghorn calculi and a voluminous renal tumor locally advanced associated with a mass of the left lumbar fossa measuring evoking a metastatic lesion, and retro-peritoneal and para-aortic lymph nodes (Figures 2 and 3). The thoraco-abdomino-pelvic scan revealed distant metastases in lung, pleura (Figure 4) and bones. It showed also a thrombosis of the left renal vein. Therefore, the renal malignancy was diagnosed as Stage IV. Biopsy report revealed a Squamous Cell Carcinoma (SCC) well differentiated, infiltrating extensively into the surrounding renal parenchyma. Immunohistochemical analysis showed a positive staining for cytokeratin 7 (CK7) and p63. Curative dose of heparin was prescribed for the thrombosis of renal vein and patient has received the first cycle of chemotherapy based on carboplatine (AUC5) and gemcitabine (1000mg/m²) and the evolution was marked by a rapid deterioration of his performances status.

Discussion

Primary renal squamous cell carcinoma is a rare kidney tumor accounting for 0.5-0.8% of all kidney malignancies [1]. It affects mainly the age group of 50-70 years with a slight female predominance. It is characterized by aggressive local behaviour and rapid distant spread.
Some factors were found to be etiological of renal SCC such as recurrent urinary tract infections, chronic inflammation and renal calculi which are the leading factors [2,3]. But the causal association between renal calculi and development of SCC is still unclear [5]. Other factors were also identified including smoking, chemicals, analgesics containing phenacetin, schistosomiasis, vitamin A deficiency, and hormonal imbalance [4,5]. Hydroureter was also reported in upper urinary tract carciners and was found to be associated with advanced stages of the disease [6]. Radiologically, there are no specific-imaging features. The CT scan may show a solid mass, with obstruction and hydroureter, but it may also reveal a renal infiltrative process without distinct mass [7,8]. The main differential diagnosis for a renal SCC associated with calculi is the Xanthogranulomatous Pyelonephritis (XGP), because it is also associated with renal calculi causing an obstruction with hydroureter and development of an inflammatory mass destroying the renal parenchyma simulating a primary renal tumor [7,9]. Based on these findings about the strong association between renal SCC with urolithiasis, the long indolent course and the misleading apparence, screening should be considered in patients with known urolithiasis. In our case, the patient presented as apparent risk factors: smoking, long history of renal calculi and recurrent pyelonephritis. Imaging findings showed a large infiltrative tumor in the renal parenchyma of the left kidney, associated with big calculi leading to an important hydroureter, and responsible of distant metastases. Given the lack of specific clinical and radiological characteristics in renal SCC histopathological confirmation is required to avoid misleading diagnosis [7]. Histologically, renal SCC is commonly moderately or poorly differentiated and more infiltrative than the transitional cell carcinomas [10]. At Immunohistochemistry, it often stains positive for AE1/AE3, cytokeratin 7 (CK7), and negative for CK20, CEA, S100 and neuroendocrine markers [11]. For patients with early stage of renal SCC, radical surgery is the standard of care [9], while for metastatic stages, there is no standard in treatment and conventional chemotherapy using cisplatin based regimens is still the mainstay of treatment [12] despite the poor results in term of response and the poor survival results with a 5-year survival rate less than 10% and a median survival of 5 months [13]. Therefore, novel targeted therapeutic approaches are highly needed.

**Conclusion**

Primary renal SCC is a rare entity, considered to have a strong association with a renal calculi and urinary tract infections. In the absence of imaging-specific features the diagnosis of SCC should be taken into consideration in patients with risk factors especially long standing renal stone disease.

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