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Case Report

Anephric Patient due to Bilateral Upper Urinary Tract Urothelial Carcinoma: A Case Report -

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SUMMARY

Bilateral urothelial carcinoma of the upper excretory tract (UUTUC) has been rarely reported in the literature. This article describes the observation of a 70-year-old patient, chronic smoker 20 pack/year, who complained of hematuria and whose biological and radiological examination revealed a bilateral UUTUC without concomitant bladder cancer. The left and the right Nephro-ureterectomy surgery with bladder cuff went off without any complication with an interval of 89 days in which the patient took some rest and received dialysis sessions because he was diagnosed with chronic end-stage renal failure. The aim of this article is to present this unusual case and demonstrate what we believe to be the best surgical management for this type of patient.

INTRODUCTION

Urothelial Carcinoma (UC) is the fourth most common type of cancer in both men and women. In this carcinomas, bladder cancer is by far the most common (90-95 %), Unlike Upper Urinary Tract Urothelial Carcinomas (UUTUC) which represent only 5-10 % [1] Moreover, bilateral upper urinary tract urothelial carcinoma is even rarer. In this article, we report a case of a patient with bilateral UUTUC, with a focus on diagnostic tools, and also the partnership between multiple medical and surgical departments to handle this challenging disease.

OBSERVATION

A.L is 70 years old, the chronic smoker at a rate of 20 packs/year. A few months before his admission, the patient complained of left back pain with macroscopic hematuria. His abdominal-pelvic CT scan revealed a mass in the upper pole of the left kidney, a large cortico-medullary cyst in the right kidney, as well as a significant upper right urinary tracts dilatation upstream of a spontaneous hyperdensity of the terminal portion of the ureter measuring about 15mm. We completed with an MRI (for a better characterization) that had shown: A 3cm lesion of the right retro-metric ureter arriving at the meatus responsible for an ureterohydronephrosis with atrophic appearance of the right kidney, indicating a chronic obstruction (Figure1). On the left kidney, we found a suspicious mass in the upper pole with thick septa (Figure2). There was no lymphadenopathy or bladder abnormality or other secondary cancer. His biological exams revealed anemia (hb: 8.7g/dl), for which he was transfused, as well as an altered renal function: Urea: 1.08g/l; creatinine: 70.28mg/l; the rest of the test was normal. The patient was treated by nephrologists for chronic renal failure with twice-weekly dialysis sessions and underwent a cystoscopy in the urology department showing a normal bladder with a double J stent placement in the left side. His selective urinary cytology of the left urinary tract revealed the presence of an atypical high-grade cell.

The discussion in a multidisciplinary consultation meeting concluded that a left Nephro-Ureterectomy (NUT) first with bladder cuff should be done then a second selective urinary cytology for histological confirmation on the right side then right NUT with right bladder cuff, with a bladder-sparing management and regular cystoscopic follow-up. The post-operative follow-ups for the first surgery (Figure 3) were simple. The second selective urinary cytology revealed the presence of atypical high grade cells, and we performed the second surgery: right NUT+ bladder cuff (Figure 4) after about 89 days from the first one, during which the patient regained strength and continued his dialysis sessions. We should notice that the CT scan after 3 months of the first surgery was normal besides the right ureteral mass already known. The anatomopathological examination of both sides had shown an urothelial carcinoma pT3N0Mx with a

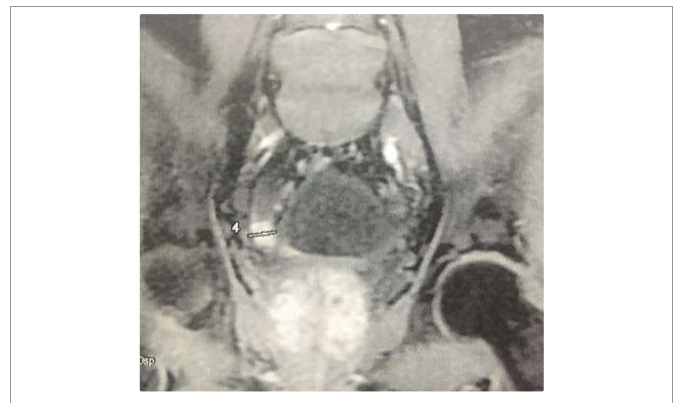


Figure 1: Hyperdensity of the right ureter's terminal part.

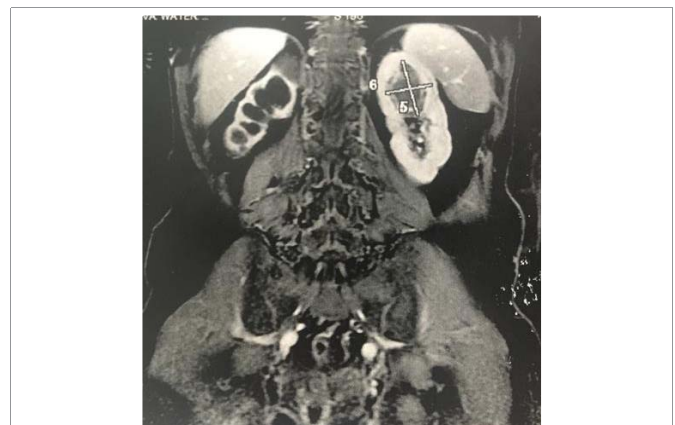


Figure 2: Suspicious mass of the left kidney.

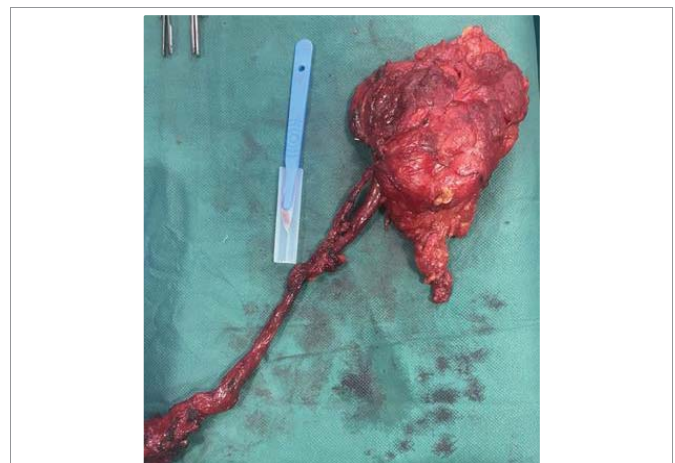


Figure 3: Left nephroureterectomysurgical specimen.



Figure 4: Right nephroureterectomy surgical specimen.

safe bladder cuff adjuvant chemotherapy should be done but with his renal function, we preferred to continue by a simple observation. At the time of writing this article, the patient is at day 20 of his second operation, anephric, in good general condition, with follow-up by nephrologists for his bi-weekly dialysis.

DISCUSSION

Tumors of the upper urinary tracts are a rare entity with an incidence ranging from 1 to 5% [2] and are more common in Balkan areas with endemic kidney diseases. Their age of occurrence is between 65 and 70 [3] with a male predominance [4]. The renal pelvis is four times more affected than the ureter. However, the distal ureter remains also to a preferential localization in 70 to 75 percent of cases [2]. The main risk factors often reported in historical series are: Age, sex, race, smoking, cyclophosphamide, history of infection and urinary lithiasis, hypertension, arteriosclerosis, Chronic Kidney Failure (CKD), and abuse of phenacetin [2-4]. Bilateral UUTUC shares the same Risk factors, as well as exposure to aristolochic acid, which is a nephrotoxic agent known for its mutagen and carcinogen role who induces a specific mutation in the codon 139 of the p53 gene (AAG TAG; Lys Stop) [3]. Two theories have been proposed to explain the pathophysiology of the existence of two synchronous upper excretory tract tumors [5]. Firstly, the theory of “Field Cancer” which states that mutagens in urine are in contact with the entire urothelium. As a result, these agents would be able to induce the development of multiple tumor clones in different levels of the ureter.

Secondly, the theory of “intraluminal seeding of tumor cells,” which supports the clonal development of multifocal cancer. Thus, multiple localizations and carcinological recurrences would be linked to intraluminal migration and tumor cell grafting into the urinary tree wall or to the intraepithelial expansion of cells from the primary tumor. Clinically, bilateral UUTUC may be discovered fortuitously [4] or after microscopic or gross hematuria. Urinary cytology is the main method for diagnosis as well as a CT urogram and cystoscopy, which plays an important role in searching for an associated bladder

tumor. The major issue after detecting bilateral UUTUC is surgical management: should surgery be performed on both sides at the same time or be deferred? Are we going to opt for a radical or conservative technique? In a Swedish series [4] of 936 patients treated over 28 years, 15 patients had bilateral synchronous upper tract tumors. Four of them had bilateral surgery on the same day, and seven others had surgery with a median interval of 55 days (34-240 days). According to the authors, Two-step surgery appears to be an acceptable protocol and there was no evidence of significant tumor growth during the interval. Concerning the technical aspect of surgery, Fernandez I and al [2] recommend that in infiltrating and/or high-grade tumors, radical treatment would be suitable namely nephro-ureterectomy with bladder cuff, and that superficial and well-differentiated, single, low-grade, small tumors undergo conservative treatment with BCG or Mitomycin C in situ to reduce recurrence. We should mention that adequate renal function before surgery is essential to choose the conservative option. Our patient underwent a two-step radical bilateral surgery with no complications during or after the procedure. Based on our experience, we recommend a two-step surgery, which we believe is more manageable for the operator, more bearable for the patient, with the easiest post-operative follow-up.

CONCLUSION

Owing to the rarity of UUTUC, there are insufficient data to provide strong recommendations. In our case, even after bilateral upper excretory tract surgery, it is important to remember that urothelial carcinoma is known for its frequent and early recurrences. If we choose sparing-bladder management, patients must be monitored by cystoscopy for the rest of their lives.

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