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Case Report

Horseshoe Kidney and Concomitant Urinary Collector System Tumor: A Case Report

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Abstract: Urothelial carcinomas are the second most common genitourinary tumors after prostate cancer. Urothelial tumors may be located in the lower (e.g. bladder, urethra) or upper (e.g. renal pelvis, ureter) urinary tract. Bladder tumors are responsible for 90% of urothelial carcinomas. However, incidence of upper urinary tract tumors increased recently, as a result of strict implementation of bladder cancer follow-up protocols and improved endoscopic imaging techniques. Ureteral tumors are less common than tumors of the renal pelvis but their prognosis is relatively poor. In the light of the current literature, we present here a 62-year-old male patient who admitted to our clinic with occasional recurrence of hematuria for 2 years and diagnosed with the hydronephrosis in the right horseshoe kidney and concomitant bladder cancer in his examination.
Keywords: Horseshoe kidney, ureter tumor, transitional epithelial cell cancer, hydronephrosis

INTRODUCTION

Upper urinary tract urothelial carcinomas are rather uncommon. Transitional epithelial cell cancers of the renal pelvis are responsible for 5% of all urothelial cancers, while ureteral tumors constitute only one-third of the renal pelvis tumors [1]. Ureteral tumors are less common than tumors of the renal pelvis but their prognosis is relatively poor. Patients usually present with hematuria, right flank pain, palpable mass in the abdomen and frequent urinary tract infections. Otherwise, they may be detected incidentally during their application for any other reason.

In this present study, we intend to present a patient with horseshoe kidney and ureteral tumor which can be seen very rarely in the literature.

CASE REPORT

A 62-year-old male patient admitted to our clinic with the complaint of occasional recurrence of hematuria for the past two years. He had a nonsignificant history, except smoking, with normal physical findings. No pathology was detected in his examination two years ago, when he first applied for hematuria. He had no additional disease and his laboratory analysis revealed that creatinine was 2 mg/dl while the other parameters were within normal limits. Ultrasonography revealed the right hydronephrosis, horseshoe kidney and a 3x4 cm mass in the bladder.

The patient was prepared for operation based on these findings. Papillary tumor formation was observed during bladder cystoscopy where the right side wall of the bladder joined the bottom side. Left orifice was in normal place and appearance but the right orifice could not be analysed. The right orifice became visible after complete resection of the tumor so that papillary tumor formation was observed in the ureteroscopic examination performed from this point on. Pathology result was reported as a high-grade papillary urothelial carcinoma. Then computed tomography (CT) was planned. CT revealed right hydronephrosis and horseshoe kidney (Figures 1,2,3). Using a midline incision in the patient, radical nephroureterectomy with isthmusectomy and bladder-cuff removal was performed.

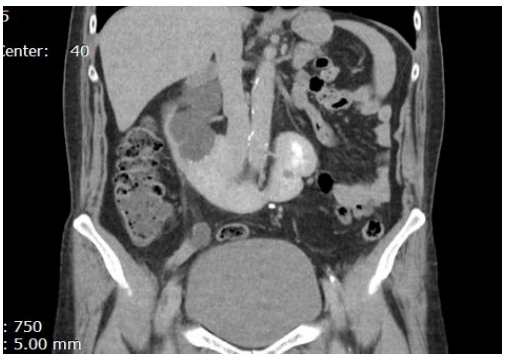

Fig-1: Sagittal section of total abdominal CT: right hydronephrosis and horseshoe kidney

Fig-2: Transverse section of total abdominal CT: dilated lower end of the right ureter

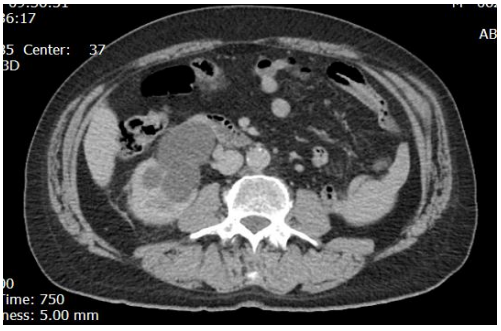

Fig-3: Transverse section of total abdominal CT: right hydroureteronephrosis

DISCUSSION

Upper urinary tract tumors constitute 1-2% of all urogenital tumors. On the other hand, only 5% of the urothelial tumors occur in the upper urinary tract [2]. Transitional epithelial cell cancers comprise 90% of the upper urinary tract tumors. The remaining 10% is composed of squamous cell carcinoma and adenocarcinoma. These cancers are 4 times more common in renal pelvis than in ureter. The incidence of having also contralateral tumors of the upper urinary tract is between 1-5.8% [3]. Despite similarities with bladder cancer, upper urinary tract tumors are detected in less than 4% of the cases with bladder cancer [4]. In contrast, bladder cell carcinoma was detected in 30-75% of the cases with transitional epithelial tumor of the upper urinary tract during follow-up. At the time of diagnosis, 60% of upper urinary tract tumors display invasive nature [3, 4]. They usually reach highest level of incidence during the ages of 70s and 80s and they are 3 times more common in men than in women [4].

Smoking, Balkan endemic nephropathy, papillary necrosis, excessive analgesic use, excessive coffee consumption, exposure to chemicals, urinary tract stones, chronic infections are the main risk factors involved in the etiology [1]. Among these etiological factors, smoking was present in our patient. The most common complaint of the patients when they applied to a hospital is macroscopic or microscopic hematuria, observed at the rate of 75% [5]. Obtuse pain in the right side is observed in 20-40% of patients and palpable mass in the lumbar region in 10-20% [4]. Patients may also present with anemia, weight loss, nausea, vomiting, loss of appetite and bone pain due to progression of the disease [6]. It can be detected incidentally during investigations and monitoring for any other reason at any healthcare center, and also can be seen during percutaneous stone surgery [7]. Intravenous pyelography (IVP) can be taken in suspected patients. IVP reveals a tumor related filling defect in the 50-70% of patients, whereas 10-30% display hydronephrosis due to invasive disease and blockage of the contrast medium passage into the ureter, in a similar manner to ureteropelvic junction obstruction. CT urography (CTU) has replaced IVP and become the gold standard method for imaging the upper urinary tract [8]. Detection rate of the upper urinary tract urothelial cell carcinoma by using this imaging method is satisfactory. CTU has a sensitivity of 96% and/while specificity of 99% in detecting small size polypoid lesions of 5-10 mm [9]. MR urography is the imaging modality of choice where CTU fails to diagnose [10].

Tumor stage and tumor grade are the best indicators of surveillance in these tumors while other factors include presence of concomitant carcinoma in situ, number of positive lymph nodes, presence of DNA ploidy, tumor localization (ureter or renal pelvis), tumor multifocality, presence of concomitant bladder tumor, surgical type, patient age, performance status of the patient and female gender. Ureteral tumors are more common in the lower part of the ureter, depending on the direction of urine flow, namely about 70% in the lower ureter, 25% in the mid ureter and only 5% in the upper ureter [2]. In our patient, however, the tumor was located at the lower 1/3 part of the ureter and in a segment about 5-6 cm. Besides, tumoral formation was also found around the right orifice of the bladder.

Upper urinary tract tumors are characterized by a poor prognosis. Metastasis is detected in approximately one fifth of patients at admission. In their study involving 32 patients in 2010, sharing 15 years of clinical experience, Demirtas et al determined tumor stage as the factor affecting progression of the tumor [11]. In these cancers, 5-year cancer-specific survival rates were reported as 100% for Ta/Cis tumors, 92% for T1 tumors, 73% for T2 tumors, 41% for T3 and 6 months of average survival for T4 tumors [2].

Horseshoe kidney and tumors of the renal collecting system have been rarely reported as case reports in the literature. Renal tumor incidence has been reported to be 3-4 times more for those with horseshoe kidney than normal population, which can probably be due to chronic infection, renal stones and obstruction [12]. Renal cell carcinoma is the most common type. Tumor usually manifests itself by involving isthmus, even though it may be localized in any region of the kidney [13]. In addition, increased tendency to renal pelvic tumors and Wilms' tumors has been reported for horseshoe kidneys [14]. National Wilms' tumor study

group detected horseshoe kidney in 41 of 8617 patients between 1969-1998 and reported the incidence as 0.48% [15].

Radical nephroureterectomy involving surrounding bladder mucosa at the lower end of ureter still remains as the gold standard, despite organ-sparing surgery procedures such as retrograde resection, antegrade resection, fulguration and partial resection [16]. This surgical approach can be implemented as open surgery, laparoscopic or combination of both methods as well as robot-assisted surgery, with the development of technology in recent years. Furthemore, detection of the tumor in the bladder during cystoscopic imaging prior to the resection would even worsen the prognosis and increases the risk of developing transitional epithelial cancer in the contralateral upper urinary tract [2]. In such cases, very strict follow-up is required via cystoscopy every 3 months and computed tomography every 6 months. So we performed radical nephroureterectomy with excision of bladder "cuff" in our patient. In addition, we followed the patient with cystoscopy every 3 months and computed tomography every 6 months, since we anticipated recurrence.

In coclusion, open surgery technique for the management of large-volume invasive papillary UC can be safely and effectively applied to patients with horseshoe kidneys, thereby limiting morbidity while adhering to the principles of oncologic surgery.

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