



An Unusual Transitional Cell Carcinoma in Megaureter in the Patient with Duplicated Collecting System

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Abstract

We report a rare case of primary urothelial carcinoma in megaureter in a patient with duplicated collecting system and ureterocele. A 63-year-old male, previously diagnosed with left sided ureterocele and megaureter, presented with gross hematuria. Cystoscopy revealed bleeding from left ureteral orifice. Urine cytology showed poorly differentiated urothelial carcinoma. MSCT urography, with delayed excretory scans, showed duplicated dysplastic kidney with impaired function, with enhancing neoplasm within dilated upper-pole moiety ureter, not outlined on delayed scans. The patient underwent a successful left nephroureterectomy. Pathology revealed high-grade papillary urothelial carcinoma confined to the ureter. There was no evidence of recurrence during two-year follow-up.

Keywords: Duplication; Ureter; Urothelial carcinoma; Dilatation

Introduction

The urinary tract malformations are rare with duplicated collecting system being the most common congenital malformation occurring in approximately 0.8% of the population [1,2]. It is more frequently found in women. Duplication can be complete or incomplete with incomplete duplication being three times more common [3]. Duplication can be associated with other abnormalities such as ureteral ectopia, vesicoureteral reflux, ureterocele and megaureter [4]. Ectopic ureter draining the upper-pole moiety of the kidney frequently ends in ureterocele, and can be dilated and tortuous (megaureter), which is associated with parenchymal atrophy, particularly of upper-pole renal moiety. Furthermore, vesicoureteral reflux into the lower-pole renal moiety often leads to chronic inflammatory parenchymal changes [5]. A quarter of all upper urinary tract urothelial tumors occur in ureters, with 60% to 75% located in the lower third [6]. Urothelial carcinoma is relatively rarely reported in patients with duplicated collecting system [7-17]. Three cases of urothelial carcinoma in megaureter [18-20], and bifid ureter [21,22] are found in the literature, respectively. Multiphasic Multislice Computed Tomographic Urography (MSCTU) is the method of choice in the assessment of upper urinary tract in patients with hematuria, the most common presenting symptom of ureteral tumor. The method is superior especially in patients with concomitant urinary tract anomalies or impaired renal function, and offers the best possibility of tumor staging [22].

Case Presentation

Material and methods

A 63-year-old male was admitted to the Urology emergency department due to intermittent painless gross hematuria. Physical examination and blood tests were unremarkable. The patient had been previously diagnosed with left sided ureterocele and megaureter demonstrated with ultrasound. Ureterocele was treated with endoscopic procedure 5 years ago in other institution. Cystoscopy was performed revealing a normal bladder and bleeding from intravesically dilated left ureteral orifice. Urine cytology suggested poorly differentiated urothelial carcinoma. MSCTU was performed including non-enhanced scans, postcontrast arterial, venous and excretory phase scans with additional delayed scan 4 h after the injection of contrast medium. Irregularly shaped shrunken left kidney with duplicated collecting system and considerable parenchymal atrophy, particularly in the upper moiety, was seen. Distal part of upper moiety ureter was tortuous and largely dilated (megaureter), crossing lower moiety ureter at the level of iliac crest (Figure 1). Two centimeters caudally from that point, within megaureter, a slightly hyperdense 3 cm × 5 cm irregularly shaped tumor, showing early postcontrast enhancement, was suspected. Dilated ureteral lumen was not completely filled with tumor (Figure 1-3). Arterial phase scans also revealed duplicated renal artery on

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Received Date: 12 Dec 2019

Accepted Date: 09 Jan 2020

Published Date: 17 Jan 2020

Citation:

Brnić Z, Schmidt S, Kovačević D, Bušić-Pavlek I, Krušlin B, Sjekavica I. An Unusual Transitional Cell Carcinoma in Megaureter in the Patient with Duplicated Collecting System. *Ann Clin Case Rep.* 2020; 5: 1786.

ISSN: 2474-1655

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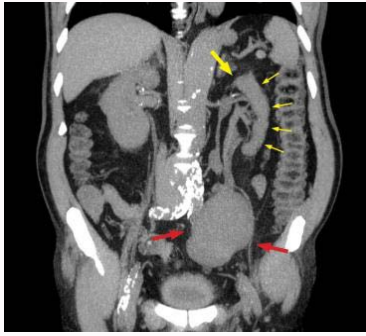


Figure 1: Non-enhanced coronal CT scan shows dysplastic left kidney with duplicated collecting system and reduced thickness of parenchyma (yellow thin arrows) with complete atrophy of upper-pole moiety (yellow thick arrow); dilated distal upper-pole moiety ureter is seen, with slightly hyperdense proliferative mass suspected within the lumen of distal megaureter (red thick arrow).



Figure 2: Excretory phase coronal CT scan shows delayed opacification of lower-pole moiety collecting system with signs of chronic obstructive uropathy (red arrows) and complete lack of excretory function of upper unit (yellow arrows).



Figure 3: Delayed excretory phase coronal CT scan shows signs of compression of contrast opacified lower-pole ureter (red arrow) with megaureter containing the tumor.

the left side, with lower-pole artery emerging from the wall of partially thrombosed aortic aneurysm (Figure 4). Standard excretory phase scans after 10 min showed incomplete opacification of lower moiety pelvicalyceal system, with signs of chronic obstructive uropathy, while atrophic upper moiety unit was not opacified (Figure 2, 3). Complete opacification of lower moiety collecting system, at delayed excretory scans after 4 h, revealed compression of lower-moiety ureter between megaureter and the psoas muscle 2 cm proximal to the tumor. No signs of periureteric fat infiltration were seen (Figure 3,5). The density of megaureter content (20 HU) suggested bloody urine, without any increase even on delayed excretory phase scans,



Figure 4: Arterial phase oblique CT scan shows enhancing tumor in distal part of dilated upper-pole moiety ureter; duplicated left renal arteries (red arrow) with the lower emerging from the wall of partially thrombosed aortic aneurysm.



Figure 5: Delayed excretory phase sagittal CT scan shows compression of lower-pole ureter with megaureter (red arrow) containing the tumor not outlined by contrast urine because of complete lack of function of upper unit.



Figure 6: Non-enhanced axial CT scan shows thickened wall of urinary bladder (yellow arrows) with 3 cm left-sided ureterocele (red arrow).

due to non functional upper moiety unit. Urinary bladder wall was thickened and trabeculated, with intraluminal orthotopic ureterocele at the orifice of the left upper-moiety ureter (Figure 6). No enlarged lymph nodes in the retroperitoneum were detected. The preoperative diagnosis was defined as ureteral tumor staged T1M0N0. Left total nephroureterectomy was performed as the definitive surgical treatment. A dysplastic upper pole ureter with soft tissue mass within its distal portion was shown at surgery. After left Gibson incision the urinary bladder was opened, and both ureteral orifices on the left side were excised. The left kidney and both ureters were sent to pathology for analysis. The gross appearance of the surgical specimen revealed a dilated dysplastic upper-moiety ureter 35 mm in diameter; containing 3 cm × 6 cm friable papillary tumor located 2 cm from the distal end of ureter (Figure 7). The tumor was histologically (WHO 2004) described as high-grade invasive papillary urothelial carcinoma (T1NxMxG3), which invaded lamina propria, but remained confined to the ureteral wall. Adjacent to upper-pole ureter within the fat, the lower-pole ureter of normal structure, slightly dilated, but not invaded by the tumor, was seen. Renal parenchyma showed focal sclerotic changes of glomeruli with normal appearance of tubuli (Figure 8).



Figure 7: Gross specimen of distal portion of upper-pole ureter which is dilated to 35 mm showed intraluminal crumbly dark tumor mass 3 cm x 6 cm in size and adjacent thickened ureteral wall; lower-pole ureter of normal appearance is seen in fat adjacent to the ureter with tumor.

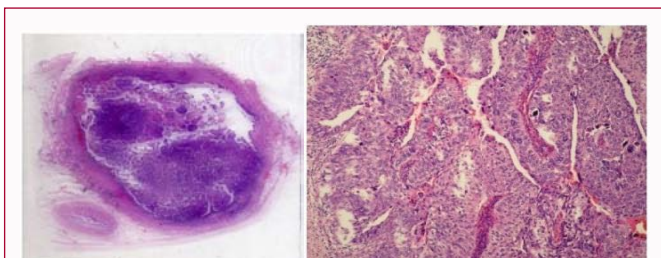


Figure 8: Low magnification and 100x magnification image show high-grade invasive papillary urothelial carcinoma, confined to the ureter.

The postoperative convalescence was uneventful and the patient showed good recovery. At 2-year follow-up there were no signs of local and distal recurrence or metachronous urothelial tumors.

Results and Discussion

Bladder cancers are the most common tumors of the urinary tract, and transitional cell tumors are the most common type. Renal and ureteral transitional cell tumors are relatively rare, accounting for only 5% of tumors. Of this percentage, more than 90% of tumors are located within the renal pelvis, while ureteral tumors are extremely rare [23-25]. The distal ureter is the most common location of tumors but multifocal implants may also occur [26]. As urinary collecting system duplication is found in 0.8% of the population, the coexistence of the two entities is expected to be very rare [1,2,10]. Duplicated collecting system very often remains undiagnosed as a large number of patients are asymptomatic, so diagnosing cancer in this situation is challenging. There is a small number of reports of ureteral cancer in duplicated collecting system in English literature [2,8-17,22]. Three reports of ureteral cancer in megaureter were found [18-20], written in Japanese and abstracted in English. Urinary duplication is frequently associated with other malformations, such as ureterocele in our case. Etiologic relationship between urinary duplication and ureteral cancers is not proven, but preferential development of cancer in the lower part of the duplicated ureters might suggest that recurrent infections, urinary stasis and reflux, and lithogenesis may be oncogenic [15]. Chronic irritation of ureteral mucosa caused by prolonged vesicoureteric reflux or urinary stasis due to ureterocele, could make ureter more susceptible to dysplasia and malignancy [11,15,27]. Similarly, increased frequency of neoplastic lesions was observed in patients with horseshoe kidneys, ureteral pseudodiverticulosis [28] and bladder diverticula [27-29]. Development of ureteral tumor in megaureter associated with ureterocele in our patient corroborates the hypothesis. Hydronephrosis is a common imaging finding in ureteric tumors, and hydroureter is often seen proximal to the point of obstruction with the lesion [30,31]. In our patient, however, megaureter which contained the neoplasm did not show proximal

dilatation, as the tumor growth did not obliterate the relatively wide lumen. Even if complete occlusion had occurred, the proximal dilation would not have developed, because the atrophic upper pole of the kidney did not produce urine. It is not fully clear whether moderate dilatation and impaired secretory function of lower moiety unit reflects reflux uropathy, which can be presumed in this type of anomaly, or is caused by compression with megaureter. The tumor is less probable cause of obstruction, as the tumor was located 2 cm apart from the point where the ureters crossed each other, and extraureteral infiltration was excluded both by imaging and histology. In the diagnostic work-up MSCTU was done as only imaging method after the tumor was suspected at cytology. In addition to the standard MSCTU protocol, the delayed excretory phase scan after 4 h was done to completely depict lower moiety collecting system, and finally exclude macroscopic lower moiety ureter infiltration. Although arterial phase scans might be omitted in the interest of decreasing radiation exposure [31,32], in our case these scans were useful to demonstrate renal vascular anatomy and relationship of duplicated renal vascularization and concomitant abdominal aortic aneurysm, the information helpful in planning surgery. Early contrast enhancement helped in detection of the tumor, which was nearly isodense to bloody content of megaureter on precontrast scans. As the upper moiety of the kidney is non-functioning, the megaureter was not opacified with contrast urine, and the tumor was not outlined as contrast filling defect, neither on standard nor on delayed excretory scans. As a consequence, multifocality could not be consistently excluded by use of MSCTU, but this was of little concern because the radical surgery was done. Considering that the tumor was histologically described as high-grade invasive urothelial carcinoma, the lack of periureteral fat infiltration and distant metastases are favourable for the patient prognosis. As the tumor was presumed as early-stage cancer confined to the collecting system wall, the total left nephroureterectomy with excision of both ipsilateral ureteral orifices and a contiguous cuff of bladder tissue was definitive treatment option. Considering that high-grade tumors of upper urinary tract can be associated with greater incidence of metachronous bladder recurrences [33], strict follow-up after radical nephroureterectomy is mandatory in our patient. Generally, ultrasound, intravenous urography, antegrade and retrograde ureteropyelography are used in detecting collecting system tumors. In our patient, previously performed ultrasound could not show cancer due to growth in the distal third of the ureter which was obscured by meteorism, whereas there was no hydronephrosis in the kidney itself due to partial renal parenchyma atrophy and poor urine production. Conventional excretory urography would not be a valuable initial imaging method in our case because of impaired function of the duplicated kidney. On the contrary, MSCTU with delayed excretory phase scans readily detected the tumor, enabled staging by exclusion of periureteral infiltration, lymph nodes enlargement and metastatic involvement of parenchymal organs and offered good depiction of complex left urinary system anatomy and concomitant aortic aneurysm.

Conclusion

In summary, we presented a case of high-grade urothelial cancer confined to megaureter associated with ureteral duplication, ureterocele, double renal arterial supply and aortic aneurysm. The patient was successfully diagnosed with MSCTU with delayed excretory scans in spite of impaired function of the left kidney and uncommon pattern of ureteral dilatation. The postoperative outcome was beneficial for the patient.

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