AN UPPER RENAL PELVIS TUMOR ARISING IN INCOMPLETELY DUPLICATED COLLECTING SYSTEM – A CASE REPORT

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ABSTRACT

Congenital abnormalities of the kidney and urinary tract are common. In particular, the duplicated collecting system is one of the most common kidney abnormalities, but most are asymptomatic. The pathogenesis of these abnormalities is also known very little and some pathogenic genes are thought to be related. The transitional cell carcinoma arises in the renal pelvis in incompletely duplicated collecting system is very rare. The report presents a clinical case: a 37-year-old female patient hospitalized with hematuria. CT scan of the abdomen discovered. The right kidney incompletely duplicated collecting system, and an upper renal pelvis tumor # 2.2 x 3.5 cm, spread to the renal pelvis and the head of the ureter. Purpose: Report on a rare clinical case: "an upper renal pelvis tumor arising in incompletely duplicated collecting system" to supplement to the current literature.

Keywords: incompletely duplicated collecting system, not complete, upper urinary tract tumor, transitional cell carcinoma.

I. INTRODUCTION

Congenital abnormalities of the renal and urinary tract are common, the frequency of approximately 1/500 fetal ultrasound examinations [1]. In particular, duplicated collecting system is one of the common renal abnormalities, occurring in the general population, nearly 10% of all human beings are born with congenital abnormalities of the genitourinary system and the duplicated collecting system occurs in approximately 1 in 160 individuals [2].

However, coexistence between the incompletely duplicated collecting system and upper renal pelvis tumor is very rare. In a case, the report aims to present a rare clinical case a right upper renal pelvis tumor arising in the incompletely duplicated collecting system. Renal pelvis tumor with pathology is transitional cell carcinoma. Recurrence of tumors is often noted, so it should be diagnosed early and directed towards treatment [3].

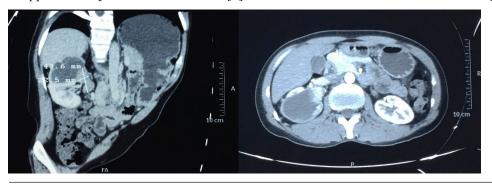


Figure 1:
The right kidney
has an incompletely
double pelvicalyceal
lobulated, the upper
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3.5 cm.

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II. A CASE REPORT

A 37-year-old female patient, hospitalized for intermittent hematuria that began 3 months ago with right hip pain, did not spread with several times of clot colic and intermittent hematuria. Recently, symptoms of hematuria often, anxiety should be admitted to hospital. Hematological investigations within normal limits. Urine analysis: revealed 50 red blood cells. Abdominal ultrasound and abdominal computed tomography showed that: The right kidney has an incompletely double pelvicalyceal lobulated, the upper pole has a soft tissue density, irregular margin, size # 2.2 x 3.5 cm, infuse contrast after injection, spread to the renal pelvis and the head of the ureter, local hydronephrosis (grade III). The lower pole is not stone, not hydronephrosis. Lower pole function is normal. The left kidney is not stone, not hydronephrosis in the renal pelvis, ureters. Normal kidney function (Figure 1).

A right nephrectomy was submitted for pathologic examination.

Protocol surgery:

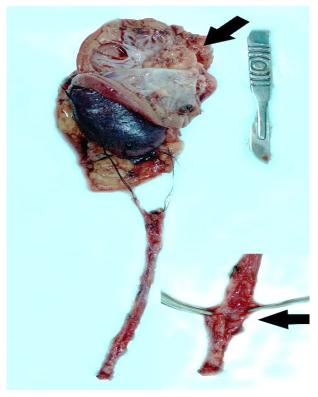


Figure 2: Two right ureters merge into one before emptying into the bladder wall about 1 cm

1 Slitting the right flank side # 10cm, dissected the anatomical layers into the retroperitoneal cavity. Dissection of the fat layer around the kidney, revealing 01 renal artery, 01 renal vein with 02 ureters on the right side of the kidney. Use hemalock to clamp the ends of arteries, veins, and ureters and remove them while taking a right kidney out (Figure 2). Investigating the tumor on the upper pole of the right kidney, spreading to the renal pelvis. To insert the drainage, close the anatomical layers, transfer the position of the supine patient.

2. Slitting Right Gibson line, dissected muscle layers, revealed the right ureter. Dissection of the right ureter, up to the bladder, detects that the two right ureters merge into one before emptying into the bladder wall about 1 cm. Cut the ureters and cut off a part of the bladder wall (Figure 2). To insert the drainage, close the incision. To send a specimen for pathological department.

Pathology: The specimen measured 11x7x4 cm and weighed 210 gm. The upper pole was expanded externally, the tumor lies in the renal pelvis spreading to the right ureter. A cut section revealed a tumor on the upper pole measured 1.5x1.5x2 cm that was circumscribed and grey-white to gray-brown color, with areas of hemorrhage and necrosis. The rest of the kidney parenchyma is well preserved.

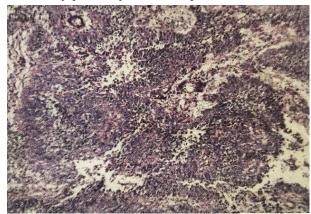
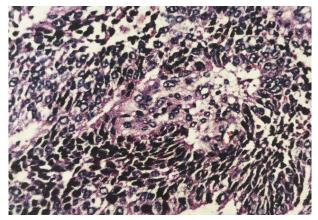


Figure 3: Large, layered tumor cells with papillae structure with blood-related axes. The nucleus is big, sharp, and not visible. Cells proliferate in many rows, basement membranes are destroyed, invade stromal tissue and blood vessels. Conclusion: TCC arising in the kidney with a duplication of the pelvicalyceal system and upper ureter.

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Biopsy tumor: large, layered tumor cells with papillae structure with blood-related axes. The nucleus is big, sharp, and not visible. Cells proliferate in many rows, basement membranes are destroyed, invade stromal tissue and blood vessels. However, there was a noteworthy duplication of the pelvicalyceal system and double upper ureters. There was no pelvicoureteric obstruction, hydrone-phrosis. The ureters, kidney vessels are normal and the fat around the kidneys have no tumors. Conclusion: Transitional cell carcinoma, arising in the kidney with a duplication of the pelvicalyceal system and upper ureter was reported

The postoperative period is stable and discharged after 7 days. The patient is uncomplicated and has remained stable for a month after surgery.

III. DISCUSSION

The kidney - urinary tract duplication

The kidney-urinary tract duplication is associated with a variety of abnormalities, so it is important to use nomenclature or terminology [4]. A duplex system is a renal unit in which the kidney consists of two pelvicalyceal systems incompletely, partially or completely duplication of the ureters. **Partial duplication** occurs when two bifid ureters join before emptying into the bladder, as in the above case. **Complete duplication**: two ureters completely separate and each ureter drains a separate pelvicalyceal system and opens separately into the urinary or genital tract. In the case of double ureters, each ureter will drain a separate renal pelvis system and empty into the urinary tract.

Duplex renal systems are commoner in females (65% of cases) and occurs bilaterally in 20% of cases, as in the above case, female and one side. A total of 60% of the kidney-urinary tract duplication is expressed, 40% represents complete ureteral duplication [4].

Embryologically, a single ureteric bud can be divided before fusing with the mesenchyme to form a bifid ureter and a duplex kidney. Complete ureteral duplication will occur if the two ureter buds arise close together in the normal position on the mesonephric duct [5]. The ontogenic mechanism for the variety of congenital abnormalities of the kidney and urinary tract shows that the initial budding of the ureter is important, as it can produce different clinical features simultaneously [6]. Several gene mutations have been identified so far that cause congenital abnormalities of the kidney and urinary tract in humans. Genes include paired box gene 2 (PAX2), a gene for X-linked Kalman syndrome (KAL), a human homolog of the drosophila 'eyes absent' gene (EYA1), angiotensin II receptor type 2 (AGTR2) and hepatocyte nuclear factor 1β (HNF-1 β) [7]. In many people with.

The kidney-urinary tract, duplication is asymptomatic and may still not be diagnosed throughout their lives, but some cases will appear in childhood with complications, such as urinary tract infections [5]. Reflux of the bladder - ureter (approximately 70%) in infected patients. Usually, only the lower pole of the ureter are related to reflux in 90% of cases due to the misplaced position, and in the remaining cases, the upper or lower poles are involved if the upper ureter is misplaced position. Reflux can occur in 1 in 5 patients.

Transitional cell carcinoma of the kidney – urinary tract system.

The development of a tumor in a duplicated collecting system is very rare. The presence of upper urinary tract cancer in a duplicated collecting system has been reported previously [3]. And according to our research, there are about 15 cases recorded. Therefore, the pathogenesis of this relationship

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with genetic and molecular basis takes a long time to further analyze.

Our patients exhibited hematuria with a dull pain on the right hip. Hematuria is a common symptom, sometimes with clot colic or patients may detect a mass in cases of renal cell carcinoma [8]. In men, a rapidly developing varicocele is rare, often on the left and is an impressive sign. In 25% of patients, there are no local symptoms. Some patients have secondary manifestations in the bones or lungs. Fever, anemia, polycythemia, constipation symptoms and, rarely, nephrotic syndrome and paraneoplastic syndrome can be presented. With the development of equipment and technological advances in prenatal ultrasound diagnosis, congenital abnormalities of the kidney and urinary tract can be detected in the uterus so that the postnatal management can be planned [9].

Primary tumors of the renal pelvis or urinary tract are relatively uncommon compared to renal parenchymal tumors, representing about 5%-10% of all renal tumors [10] and 1% of all genitourinary tumors [11]; Bladder tumors, renal pelvis, and ureter tumors appear in proportion 51: 3: 1 [7]. While TCC only 1% to 3% of upper urinary tract tumors [12]. Transitional cell epithelial types are most frequently diagnosed (85% to 95% of cases)

[13]. Then, the upper urinary tract tumor encountered in this abnormality is very rare.

A total nephroureterectomy with excision of the bladder cuff has been the standard treatment for upper urinary tract tumors. In a retrospective study of 198 patients with transitional cell carcinoma of the upper urinary tract, postoperative recurrence rate with symmetrical tumor growth accounted for 2.5%, with a ureteral stump after cutting. With conservation treatment accounted for 19% and in the bladder was 36.4% [12]. Because the rate of recurrence of the tumor is high in the ureter stump position after local resection, a total nephroureterectomy seems to be a better treatment choice. A heminephroureterectomy may be considered only in cases of complete duplication, or when the opposite-side kidney is nonfunctioning.

IV. CONCLUSION

The right upper renal pelvis tumor arising in incompletely duplicated collecting system is very rare. Diagnosis is often based on the symptoms of hematuria combined with abdominal computed tomography. A total nephroureterectomy with excision of the bladder cuff has been considered the standard of treatment for this abnormality to avoid recurrence of the tumor.

REFERENCES

- Colodny AH. Antenatal diagnosis and management of urinary abnormalities. Pediatr Clin North Am 1987;34:1365-81.
- 2. Atwell JD, Cook PL, Howell CJ, Hyde I, Parker BC. Familial incidence of bifid and double ureters. Arch Dis Child 1974;49:390-3.
- 3. Chen KS, Chuang CK, Wu CH, Liaw CC, Lee N. Upper urinary tract tumor in a duplicated collecting system: report of three cases and review of the literature. Chang Gung Med J 2003;26:377-82.
- 4. Schlussel RN, Retik AB. Anomalies of the ureter. In: Walsh PC, Retik AB, Vaughan ED, Wein AJ eds. Campbell's Urology. 7th ed., vol. 2.WB Saunders: Philadelphia, PA; 1998:1814-57.

- 5. Whitten SM, Wilcox DT. Duplex systems. Prenat Diagn 2001;21:952-7.
- Miyazaki Y, Ichikawa I. Ontogeny of congenital anomalies of the kidney and urinary tract, CA-KUT. Pediatrics International 2003;45:598-604.
- Nakanishi K, Yoshikawa N. Genetic disorders of human congenital anomalies of the kidney and urinary tract (CAKUT). Pediatrics International 2003;45:610-6.
- Fowler CG. The kidneys and ureters. In: Williams NS, Bulstrode CJK, O'Connel PR eds. Bailey & Love's Short practice of surgery. 25th ed. Edward Arnold Ltd: London; 2008:1285-92.
- 9. Tsuchiya M1, Hayashida M, Yanagihara T, Yo-

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- shida J, Takeda S, Tatsuma N et al. Ultrasound screening for renal and urinary tract anomalies in healthy infants. Pediatrics International 2003;4:617-23.
- 10. Cotran RS, Kumar V, Robbins SL. Robbins' pathologic basis of disease (ed 5), Chap 20. WB Saunders Company, 1994, pp 987-8.
- 11. Richie JP. Carcinoma of the renal pelvis and ureter, in Skinner DG, Lieskovsky G (eds): Diagnosis and management of genitourinary cancer.

- WB Saunders Company, 1989, pp 323-36.
- 12.Krogh J, Kvist E, Rye B. Transitional cell carcinoma of the upper urinary tract: prognostic variables and postoperative recurrence. Br J Urol 1991;67:32-6.
- 13.Bennington JL, Beckwith JB. Tumors of the kidney, renal pelvis, and ureter. In: Atlas of Tumor Pathology, Series 2, fascicle 12. Washington, DC: Armed Forces Institute of Pathology, 1975; 308.