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Case Report

Anatomy Section

Unilateral Bifid Ureter with Persistent Left Superior Vena Cava: A Cadaveric Case Report

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ABSTRACT

This is a case report of unilateral bifid ureter with Double Superior Vena Cava (DSVC) which was observed during the routine dissection on a 60-year-old male cadaver. The finding is of interest due to the presence of urogenital and vascular anomalies. The case belonged to the category of incomplete double renal pelvis and ureter along with a Persistent Left Superior Vena Cava (PLSVC) with an enlarged coronary sinus and a normal Right Superior Vena Cava (RSVC). Incomplete double ureter with the angled point of junction of the two ureters creates a narrowing that can further obstruct normal flow, predisposing to uretero-ureteric reflux and associated complications such as hydronephrosis, as well as increases the possibility of ureteral injury during surgery. PLSVC is usually an insignificant finding, however, its presence makes it difficult for insertion of central venous catheter via left internal jugular vein and is important to surgeons and interventionalists.

Keywords: Double superior vena cava, Incomplete double ureter, Uretero-ureteric reflux

CASE REPORT

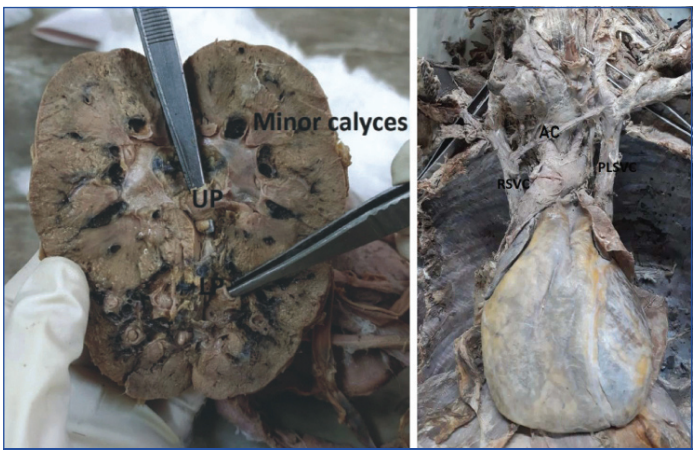
A rare case of unilateral bifid ureter along with duplex collecting system and PLSVC is presented in this case report. The anomaly was detected in a 60-year-old male cadaver during routine dissection in Medical College, Kolkata, India. The left kidney was 12.4 cm in length with a width of 9.9 cm at its maximum, while the right kidney measured 11.2 cm in length and 7 cm in breadth at its maximum. The present case belonged to the category of incomplete double renal pelvis and ureter. The right kidney had double ureters arising from the superior and inferior poles of the renal pelvis that united with each other to form a single ureter (Y-shape) distally before opening into the urinary bladder. At its origin, two separate limbs of the right-side ureter could be differentiated. The two limbs coursed down upon the posterior wall of the abdomen and later joined with each other beneath the level of pelvic brim. From the hilum to the point of junction, the superior limb measured 27.7 cm while the inferior limb measured 26.5 cm. These limbs united 2.3 cm above the urinary bladder and formed a single ureter, that opened into the bladder wall by a single aperture.

The duplicated ureters ran side-by-side; the laterally placed ureter arising from the inferiorly placed renal pelvis and the ureter placed medially, beginning from the renal pelvis situated above the inferior one. Both the ureters had almost equal diameter. There was no abnormality in the opening of the ureter into the bladder [Table/Fig-1].

[Table/Fig-1]: Right bifid ureter.
UU: Upper ureter; LU: Lower ureter; RV: Renal vein

On the left side, the ureter was single, normal measuring 28 cm in length from the hilum to the bladder wall and width of 1.5 cm was noted. The relation of left renal vein, artery with left pelvis was maintained in before backward direction. In the right pelvis, the upper ureter passed behind the right renal artery and vein while the lower ureter passed below the right renal vein. In the coronal section, duplication of pelvicalyceal system including two separate renal pelvis on the right side, one above the other, each giving rise to a ureter was observed in right kidney [Table/Fig-2]. There were five minor calyces in the upper pelvis and three in the lower pelvis.

In the same case, presence of both left and right superior vena cava was found, PLSVC with an enlarged coronary sinus and normal right SVC [Table/Fig-3]. These were equal in size. The left SVC crossed lateral to aortic arch, continued into the coronary sinus and the coronary sinus opened into the right atrium. A communication between left and right SVC was also present. Based on the anatomical classification of whether the azygos vein and the innominate anastomosis is present or not [1], the case reported here belonged to the Type I classification (presence of innominate anastomosis between the left and right SVC).



[Table/Fig-2]: Coronal section of right kidney showing duplication of pelvicalyceal system. **[Table/Fig-3]:** Persistent left superior vena cava with normal right superior vena cava having communication between them.
UP: Upper pelvis with 5 minor calyces; LP: Lower pelvis with 3 minor calyces; RSVC: Right superior vena cava; PLSVC: Left superior vena cava; AC: Anastomosing channel. (Images from left to right)

DISCUSSION

Case reports demonstrating the presence of unilateral bifid ureter and PLSVC are rare. A cadaveric study in Japan had reported similar findings; however, the present case report is unique in the fact that a communication was found between right and left superior vena cava [2]. Congenital renal anomalies, including double ureter, constitute nearly 20-30% of all prenatal abnormalities [3]. Bifid ureter, which is one of the variations related to congenital anomalies of urinary system, usually presents as an incidental finding and may be associated with other congenital anomalies [4]. The incidence of double renal pelvis and ureters have been recorded variably in the range 0.23-3% [2,5]. Early splitting of the ureteric bud results in duplication of ureter. There may be partial or complete splitting, resulting in division of metanephric tissue into two parts, each having its own renal pelvis and ureter [6]. This explains the phenomenon leading to a duplex collecting system. Around the 5th week of intrauterine life, the two sources from which permanent kidney develops are: ureteric bud and metanephric blastema. Into the metanephric blastema, the uretric bud grows and branches, that leads to the formation of adult kidney while the ureter is formed from the stalk of the ureteric bud. Sometimes, however the ureteric bud divide before penetrating metanephric blastema which may then give rise to a bifid ureter [7].

During open or laparoscopic surgical procedures of abdomen and pelvic region, the possibility of ureteral injuries is increased in the presence of an incomplete double ureter [2]. Although bifid ureter remains largely asymptomatic, certain complications such as frequent urinary tract infection, uretero-ureteric reflux (most common in incomplete), vesico-ureteric reflux (common in complete), ureteric stenosis, urinary lithiasis and pyelonephritis have been reported [8,9]. In cases of incomplete double ureter having angled point at union of the two ureters, a fourth constriction is formed that further obstructs the normal flow, leading to flow of urine in reverse direction causing complications such as hydronephrosis [3]. PLSVC is the most common congenital abnormality of the venous system of thorax with an overall occurrence of around 0.5% and as high as 10% among persons affected with congenital cardiac disorders. It is generally asymptomatic and detected only incidentally during imaging or surgery of cardiovascular region, cannulation of left subclavian vein or device implantation. In most of the cases, where congenital heart disease is absent, it opens into the right atrium via coronary sinus [10].

As per anatomical classification of PLSVC based on whether the azygos vein and innominate anastomosis is present or absent, four types have been described [1]:

a. **Type I-** Left and right SVC anastomosis through innominate venous trunk.

b. **Type II-** Completely separated left and right SVC.

c. **Type III-** Absence or atrophy of the right SVC, blood drainage being realised through the left SVC.

d. **Type IV-** Separated left and right SVC, each one presenting its own correspondent azygos vein.

In some cases, drainage of PLSVC into the left atrium was due to the failure of formation of coronary sinus; this may be associated with other congenital heart diseases [1]. Owing to the clinical implications, a dilated coronary sinus on echocardiography, raise the suspicion of a PLSVC [11]. A double SVC is characterised by persistence of Left Anterior Cardinal Vein (LACV). The persistent LACV, the left SVC, drains into right atrium via coronary sinus [6]. The co-existence of PLSVC and RSVC is also called DSVC [12]. In a majority of cases, PLSVC appears as a duplicated superior vena cava (left and right). In this situation, the innominate vein connecting them is present in 30% of cases [3]. This finding is of importance to surgeons and interventionists as the placement of a central venous access device via left internal jugular vein can be rendered difficult [7].

CONCLUSION(S)

Prior knowledge about anomalies of the urinary system is useful for surgeons, as co-existing anomalies such as ureteral duplication, may increase further the morbidity of affected individuals. It is also useful for radiologists to familiarise themselves with complete/ incomplete double ureter and its subtypes as these may lead to misinterpretation of radiological images.

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