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Duplicated collecting system of a kidney complicated with hydronephrosis – diagnostic methods review: A case report

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Summary

- Background:** Duplicated collecting system of a kidney occurs in 1.7–4.2% of the population. It is a complex, unilateral or bilateral, congenital abnormality of the pyelocalyceal system and the ureter. The 2 ureters fuse to form a single ureteral orifice or empty separately into the bladder (ureter duplex). Duplicated collecting systems with complete ureteric duplication may lead to developing vesicoureteral reflux, hydronephrosis, and urinary infection.
- Case Report:** This article presents a case of a 49-year-old woman with duplicated collecting system and hydronephrosis in the upper pole. The anomaly was diagnosed using urography, ultrasonography and computed tomography examination.
- Conclusions:** The best method for diagnostics of the duplicated pyelocalyceal system complicated by hydronephrosis is computed tomography examination, especially multislice computed tomography. The authors present also the options for therapy.
- Key words:** urogenital system defect • ultrasonography examination • computed tomography examination
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Background

Duplicated collecting system of a kidney is a congenital anomaly affecting the pyelocalyceal system and the ureter that occurs in 1.7–4.2% of the population and is bilateral in ca. 20% of cases [1]. The duplicated ureters either fuse to form a single orifice to the bladder (partial duplication), or empty separately into the bladder (complete duplication of the ureter). Depending on the anatomic level of their anastomosis, two types of junctions may be distinguished: extravescical (Y-shaped, more common) and intravesical (V-shaped). In very rare cases, the ureter, initially single, divides in its distal portion, assuming the shape of inverted Y. Complete duplication occurs, when the two ureters have separate orifices into the bladder. The ureter draining the lower pole of the kidney most often enters the bladder slightly laterally to the normal site, which is associated with shortening of the segment running within the bladder wall and worse “anchorage” in the bladder triangle. As a result, there is an increased

risk of vesicoureteral reflux, leading with time to distension of the urinary tract and in the later period to scar-like lesions in the inferior calyces. The orifice of the upper ureter is often narrowed and located more medially and lower than the normal site (Weigert-Meyer rule). The upper ureter may also empty to the seminal vesicles, posterior urethra, vaginal vestibule or rectum – the location of the orifice is then referred to as ectopic, and it may lead to outflow disturbances and urine retention in the upper urinary tract. If the course and the orifice of the duplicated ureter are normal, the anomaly is generally asymptomatic and is detected accidentally during imaging examinations performed for other reasons. The clinical significance of renal collecting system duplication is associated with the possibility of complications such as urolithiasis, inflammations of the urinary tract and renal parenchyma, or hydronephrosis, developed as a result of vesicoureteral reflux or cysts of the ureteral orifice [2–4]. Ureterocele is a cyst-like distension of the final segment of the ureter within the wall of the urinary bladder with

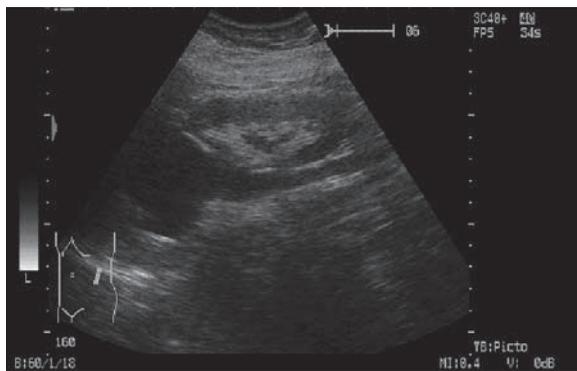


Figure 1. Ultrasonography examination of the left kidney shows a hypoechoic lesion representing hydronephrosis in the upper part of duplicated kidney collecting system. The demonstration that dilated ureter exits from the upper pole mass established the diagnosis of duplication of the renal collecting system.

submucosal protrusion into the vesical lumen. There are 2 types of cysts: simple and displaced (ectopic). Simple cysts usually affect a single ureter and are located at the site of its normal orifice. Ectopic cysts, frequently accompanying duplicated ureters, affect the ureter draining the upper part of the renal collecting system and are found in the vicinity of the bladder neck or in the internal orifice of the urethra. Early diagnosis of a cyst of the ureteral orifice makes it possible to institute less invasive treatment and is very important for the prognosis [5,6].

Case Report

A 49-year-old woman complaining of transient painful symptoms in the left lumbar region of a few years' duration was referred to the Department for computed tomography to verify the unclear findings concerning the left kidney in USG and urography. Abdominal USG revealed an anechoic structure of 54×45 mm dimensions in the upper pole of the left kidney, which might correspond to distended upper portion of the collecting system, drained by a distended left ureter. Distension of the lower portion of the collecting system was slight. The USG image suggested the presence of duplicated collecting system (Figure 1).

Urography revealed bulging of the upper pole of the left kidney, with deformation of the pyelocalyceal system and the superior calyces and the renal pelvis compressed from above and displaced. The radiological findings were unclear and required differentiation between a cyst and a neoplastic lesion (Figure 2aB).

The patient underwent abdominal CT performed using a helical technique with a single-row Siemens apparatus, 5 mm slice thickness for the abdominal cavity and 3 mm slice thickness for the pelvic cavity, TF 5 mm, RI 3 mm, with 100 ml intravenous contrast medium bolus administered using an automatic syringe. Image acquisition was carried out before and after contrast administration. The abdominal organs were examined with 30 s and 180 s delay, and the pelvic ones with a delay of 40 s and 15 min. The assess-



Figure 2AB. Urography examination. Excretory urogram demonstrates inferior and lateral displacement of the opacified lower part of the duplicated left kidney collecting system caused by the dilated, obstructed system of the nonopacified upper part of the duplicated system. The right collecting system is normal.



Figure 3A. Computed tomography examination of the abdomen, nephrographic phase, axial scan on upper pole of kidney level. A cystic lesion in the upper pole of the left kidney with peripheral contrast enhancement and irregular shape.



Figure 3B. Computed tomography examination of the abdomen, excretory phase, coronal reconstruction. Cystic lesion represents hydronephrosis in upper part of duplicated left kidney collecting system; widened proximal part of upper ureter is visible. Cortex atrophy in upper part of the left kidney. Thin parenchymal rim with contrast enhancement. Lower part of collecting system with normal surrounding renal cortex, renal pelvis and ureter with contrast inside.

ments were carried out on axial cross-sections and on multiplanar reconstructions (MPR) based on them.

In the vicinity of the upper pole of the left kidney, a cyst-like lesion of ca. $60 \times 45 \times 40$ mm dimensions, with contrast enhancement of the peripheral part of up to 2 mm thickness, was visualized. The lower pole of the described lesion was narrowed in funnel-like manner. A duct up to 15 mm in diameter, extending to the urinary bladder along the normal ureter, arose from it. The two ducts crossed in their middle segment (Figure 3A,B).

In the topography of the paravesical segment, structures saturated with calcium salts of max. size ca. 13×5 mm, which could correspond to concrements, were visible (Figure 4AB).

The overall radiological presentation suggested duplication of the pyelocalyceal system of the left kidney and the ureter with the presence of concrements in the paravesical seg-

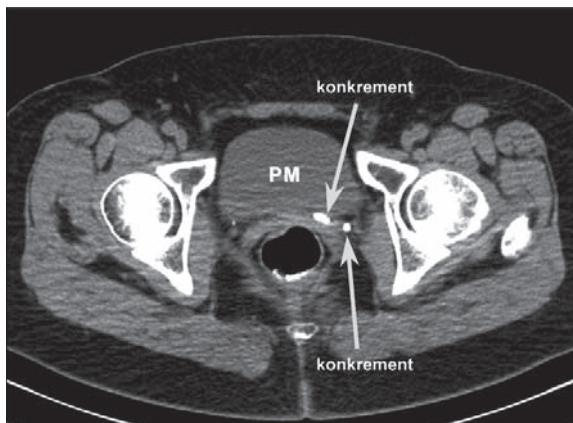


Figure 4A. Computed tomography examination of the pelvis, before contrast medium injection. Axial scan: near the urinary bladder (PM) are shown calcified structures (arrows), which may represent concrements in the left ureter.

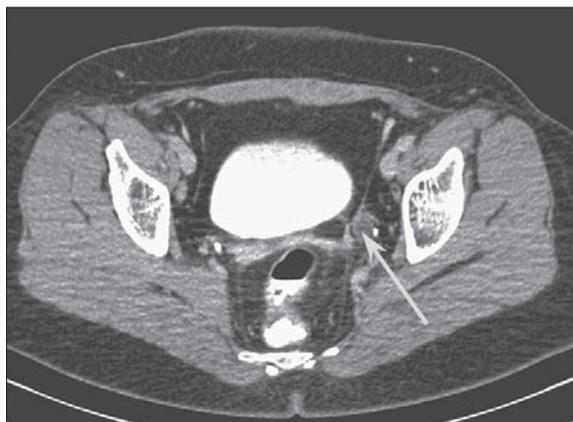


Figure 4B. Computed tomography examination of the pelvis, excretory phase, axial scan: image demonstrates two left ureters. One of them (arrow) is dilated – diameter max. 15 mm and nonopacified.

ment of the ureter arising from the upper part of the duplicated collecting system, which, impairing urinary outflow, led with time to chronic urine retention.

Discussion

The diagnostics of duplication of the collecting system of the kidney utilizes such imaging methods as: ultrasonography (USG), urography, computed tomography (CT), sometimes miction cystoureterography. The essential and initial examination is abdominal USG, because of its common availability. Duplication of the collecting system is indicated in USG by the presence of echo of two separate sinuses („split sinus” sign). As this sign is not always seen, ultrasonography is a specific method, but of low sensitivity [1,3,7]. The results of USG can be verified by urography or CT. In urography, attention should be paid to asymmetric size of the kidneys and the pyelocalyceal systems. The distended collecting system and the ureter draining the upper pole of the kidney may compress and displace laterally the lower pyelocalyceal system giving the „drooping lily” sign [8,9]. It should be realized that impairment of the renal secretory function may lead to unclear urographic findings, like

in the presented case. In such a situation, the respective collecting system shows no contrast enhancement, and the surrounding parenchyma is invisible („Nubbin sign”) [10]. It may mimic a tumor or postinfarction parenchyma atrophy. Differential diagnosis should also take into consideration postinflammatory atrophy, postoperative scarring or hypoplasia. Reduction of the number of calyces, visualization of a duplicated collecting system and the presence of well-defined borderline of normal kidney parenchyma make it possible to differentiate the anomaly from a renal tumor. In order to visualize the ectopic location of the ureteral orifice, miction cystoureterography can also be performed, and in patients with hydronephrosis the second ureter and the level of its orifice can be visualized by pyelography.

At present, multislice computed tomography (MSCT), enabling multiplanar reconstructions and volume rendering, plays an increasing role in diagnostics of the anomaly. A typical CT presentation in case of nonobstructed duplication of the pyelocalyceal systems is a „faceless kidney” sign. The transversal plane image presenting the borderline between the upper and lower part of the duplicated collecting system shows the renal parenchyma without the hilus („face”) [9,11]. Computed tomography is a method more precise than urography and USG in the diagnostics of duplicated pyelocalyceal system complicated by obstruction, especially in the cases when the renal secretory function is impaired significantly or completely and parenchyma atrophy occurs. MSCT can be useful also to visualize ectopic ureteral orifices. On the other hand, a disadvantage of this method is the possibility that the presence of hypertrophic renal columns can sometimes lead to incorrect diagnosis of duplication of the ureters – by a false positive presentation of the “faceless kidney” sign.

Besides MSCT, magnetic resonance plays an important role in urological diagnostics. MR urography includes two methods of imaging of the urinary tract:

MR hydrography (static MR urography – sMRU), utilizing T2-weighted images without contrast enhancement, in which solid organs and mobile fluids are hypointense, and static, or slowly moving fluids – hyperintense. Three types of sequences are used: RARE, FSE and HASTE.

sMRU is a sensitive method of detection of hydronephrosis, assessment of its severity and also of the level of urinary outflow obstruction. Because of no necessity of contrast administration, puncture of the urinary tract and exposure to ionizing radiation, it may be used in children, pregnant women, patients hypersensitive to contrast agents and after kidney transplantations.

MR urography with contrast enhancement, utilizing T1-weighted images with contrast administration, used

for anatomical and functional assessment of the kidneys and the urinary tract, especially the lower segments of the ureters. This technique allows to differentiate acute and chronic obstruction of the urinary tract, but has a lower value in detection of its causes [12,13].

Duplication of the renal collective system with chronic retention should be first of all differentiated from cyst-like forms of renal tumors. In case of cystic lesions, thickened walls and irregular outlines, as well as the presence of contrast enhancement, arouse the suspicion of a dysplastic process. The following tumors of the kidney may develop in cystic forms: multilocular cystic nephroma, neural cell carcinoma and Wilms tumor. Multilocular cystic nephroma is a rare, nonhereditary benign kidney tumor, usually localized unilaterally in the lower pole. Plain radiography of the abdominal cavity and urography usually visualize such a tumor as a soft tissue mass (especially if it is large and displaces the adjacent structures). USG reveals a large, well-delineated cyst-like mass. The presence of a narrow parenchymatous margin can arouse doubts in differentiation between that tumor and duplication of the renal collecting system with chronic urine retention. Visualization of hyperechogenic partitions between the cysts and the collecting system – the renal pelvis continuous with the ureter – becomes important. In contrast-enhanced CT images, a multilocular cystic nephroma is visualized as a solid-cystic multilocular tumor indenting into the renal hilus. Other tumors may yield similar images. Differential diagnostics should also take into consideration renal column hyperplasia and polycystic dysplasia. Surgical treatment of this anomaly involves resection of the inactive upper system together with maximally long ureteral segment from lumbar approach. In case of a ureterocele with retained parenchymal function within the upper collecting system, endoscopic cystotomy can be performed to obtain decompression of the upper collecting system.

Conclusions

1. In cases of uncomplicated duplication of the renal collecting system, urography is usually sufficient to establish the diagnosis.
2. Complicated cases may arouse doubts in differential diagnostics.
3. Multi-row computed tomography with multiplanar reconstructions and volume rendering is nowadays increasingly available and popular method applied in the diagnostics of renal collective system duplication accompanied by urine retention and impaired secretory function of the kidney.

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