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Blind-ending bifid ureter – a case report of rare congenital anomaly and its sonographic appearance

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Keywords

ureter; ultrasonography; urinary tract

Abstract

Aim of the study: We report a case of a blind-ending bifid ureter in a 67-year-old woman with ascites initially diagnosed with B-mode and Color Doppler ultrasonography and afterwards verified with contrast-enhanced abdominal computed tomography. A literature review of the pathogenesis, sonographic appearance with differential diagnoses and clinical significance is also presented and discussed. Case description: The patient was referred for an abdominal ultrasound due to enlarged abdomen circumference. Ultrasound revealed signs of chronic pancreatitis with cavernous transformation of the portal vein and large ascites resulting in bilateral pelvicalyceal system dilatation. Additionally, we have preliminarily diagnosed right-sided, dilatated blind-ending bifid ureter with associated contralateral complete duplication of the ureter and the collecting system. These findings, initially revealed with ultrasound, were confirmed with contrast-enhanced abdominal computed tomography. Conclusions: To our knowledge, this is the first detailed description of sonographic appearance of blind-ending bifid ureter.

Introduction

A blind-ending bifid ureter is a congenital anomaly of the urinary system. There has been over 200 reported cases in the literature since the first description by Herbert in 1904⁽¹⁾. The exact incidence of this variant is unknown since patients are typically asymptomatic. However, they may present with abdominal pain, hematuria or urinary tract infection. It appears that the condition is more frequent in women and affects predominantly the right side⁽²⁾. Most cases were diagnosed with intravenous or retrograde pyelography⁽³⁾, computed tomography (CT) urography⁽¹⁾ or ureteroscopy⁽²⁾.

Case report

A 67-year-old woman was admitted to our hospital with ascites. She was referred for an abdominal ultrasound, which revealed significant ascites, features of chronic pancreatitis and cavernous transformation of the portal vein.

Furthermore, there was moderate pelvicalyceal system dilatation on the left side with clearly visible ureter duplication (Fig. 1). The collecting system of the right kidney and right ureter were also dilated (Fig. 2). Additionally, a small (18 × 18 mm) anechoic, cystic structure was visualized near the upper pole of the right kidney (Fig. 3). The lesion continued into a tubular, 8 mm diameter anechoic structure which showed no signal on Color Doppler and followed the course of the proper ureter (Fig. 4, Fig. 5). Due to the large ascites, the urinary bladder and distal parts of ureters were not visible on ultrasound. The patient denied any urinary symptoms and her urinalysis was unremarkable.

Contrast-enhanced abdominal CT was performed, which confirmed the ultrasonographic conclusions. There was large ascites constricting the distal part of ureters and the urinary bladder was almost empty. A transverse section shows bilateral duplication of ureter (Fig. 6). On the left side, both ureters continued superiorly into the duplicated collecting system of the left kidney. However, on the right

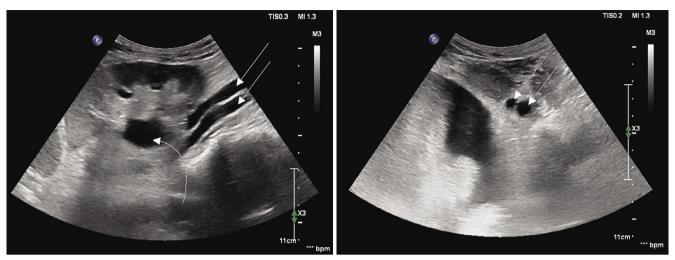


Fig. 1. Left kidney collecting system dilatation (curved arrow) with ureteral duplication (straight arrows) in longitudinal (left) and transverse (right) scans

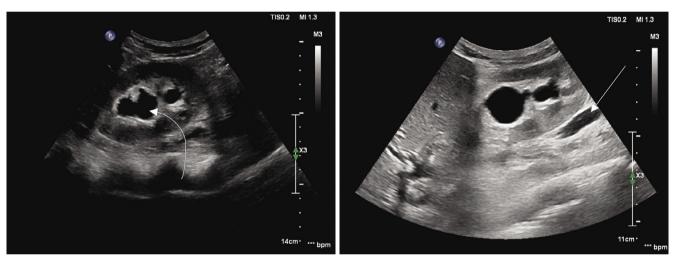


Fig. 2. Right kidney collecting system dilatation (curved arrow, left) with visible distention of the right ureter (straight arrow, right)

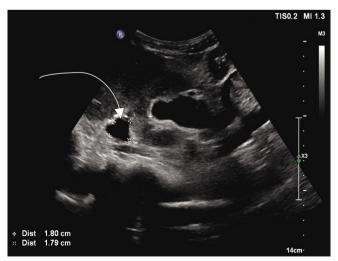


Fig. 3. A small anechoic, cystic structure (curved arrow) superior to the upper pole of the right kidney represents the proximal dilatation of the blind branch of bifid ureter

side one branch running superiorly dilatated into a cystic lesion measuring 17×12 mm, located near the upper pole of the right kidney (Fig. 7, curved arrow), while the other one joined the proper collecting system of the right kidney. Distally, the two ureteral branches conjoined on the right side while they remained separate on the left side (Fig. 8).

In the late CT phase, the contrast was excreted by both kidneys and was clearly visible in the pelvicalyceal systems (Fig. 7). However, the cystic lesion described above in both ultrasound and CT examinations contained no contrast, which means that there was no functional renal tissue surrounding the proximal bulbous dilatation of one of the ureter branches on the right side. All findings implied the diagnosis of a blind-ending branch of the right ureter.

Discussion

A blind ending-bifid ureter is an uncommon anatomical variant that results from abnormal embryogenesis.

J Ultrason 2022; 22: e191-e195

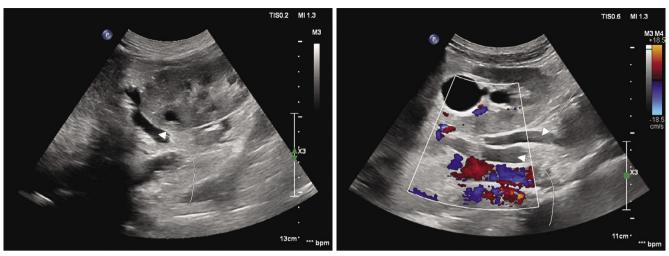


Fig. 4. Dilatated blind branch of the bifid ureter (curved arrow), following the course of the proper ureter (straight arrow), both showing no signal on Color Doppler (right)



Fig. 5. Transverse view of two branches of the bifid ureter (straight arrows) running in the retroperitoneal space. Ascites is also visible



Fig. 6. A transverse section from abdominal CT shows two branches of the ureter bilaterally (straight arrows)



Fig. 7. A transverse section from abdominal CT (inferior to Fig. 6) shows fusion of two branches into a single ureter on the right side (curved arrow). Ureteral branches on the left side remain separate (straight arrows)

At around 28 days of gestation, the ureteric bud arises from the mesonephric duct and starts growing cranially to reach the metanephric mesenchyme. The ureteric bud normally develops into ureter, renal pelvis, renal calices and collecting tubules, while the metanephric mesenchyme forms nephrons including glomeruli, proximal convoluted tube, loop of Henle and distal convoluted tubule.

If the ureteric bud divides into two branches which afterwards reach separately the metanephric mesenchyme, it results in a complete duplication of the renal collecting system. Premature division of the ureteral bud with subsequent failure of one branch to reach the metanephric mesenchyme produces a blind-ending bifid ureter.

According to the definition established by Culp, it is a blind-ending hollow structure whose lumen joins that of the ureter at a distinct angle, whose wall presents the same histologic coats as the ureter and whose length is more than twice its greatest diameter⁽⁴⁾. Three subtypes of this variant can be distinguished: proximal, middle and distal, depending on the origin of the blind branch⁽²⁾.

J Ultrason 2022; 22: e191-e195

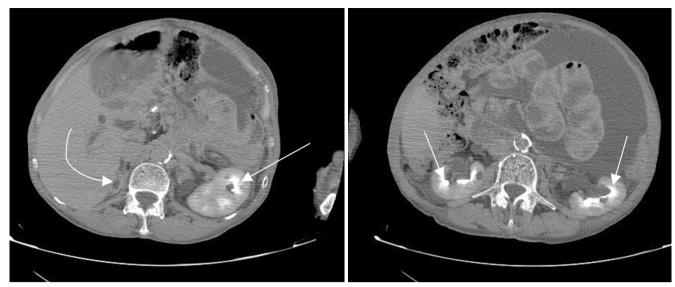


Fig. 8. A transverse section from late phase contrast-enhanced abdominal CT shows contrast in pelvicalyceal system of both kidneys (straight arrows). The proximal bulbous dilatation of the blind-ending branch of the right ureter contains no signs of contrast enhancement (curved arrow)

Typically the blind limb has a bulbous proximal dilatation, which is most likely due to uretero-ureteral reflux. The mechanism of the reflux includes asynchronous peristaltic waves: an antegrade peristalsis in the proper ureter which, after reaching the bifurcation, proceeds as a retrograde peristalsis into the blind branch. The blind branch may be entirely dilated which is thought to be due to partial stenosis at the level of the Y junction⁽⁵⁾.

Some authors claim that a cap of nephrogenic tissue representing the remnant of the metanephric blastema^(6,7) or a fibrous band connecting the upper blind extremity with a metanephric remnant⁽³⁾ may be present, while others have not reported such observations⁽⁸⁾.

Differential diagnosis of blind-ending bifid ureter involves primarily its bulbous proximal dilatation and includes adrenal cysts and other retroperitoneal cystic lesions. In the case of ureter duplication seen on ultrasound or CT, both branches should be followed proximally to assess if they are connected with the collecting system (ureter duplex with duplication of the renal collecting system) or if one of them passes by the kidney ending blindly. Both branches should be also followed distally in an attempt to assess if they unite (bifid ureter) or reach the urinary bladder separately (complete ureter duplication).

The diagnosis of this anomaly can be initially suspected with ultrasonography and should be confirmed with intravenous or retrograde pyelography, CT urography or ureteroscopy. The key role of ultrasound in the diagnostic process is to visualize the lesion, characterize it using B-mode and Color Doppler options, suggest an initial diagnosis and refer for necessary further imaging mentioned above. Procedures with intravenous contrast may be less sensitive in detection of the blind branch when there is no uretero-ureteral reflux mentioned above. However, they allow to exclude

the presence of functional renal tissue surrounding the suspected blind-ending branch.

Blind-ending bifid ureter may cause complications such as abdominal pain, hematuria⁽¹⁾ and recurrent urinary tract infections⁽³⁾. Ureterolithiasis⁽⁹⁾ or even transitional cell carcinoma⁽¹⁰⁾ might develop in the blind-ending branch.

Surgical removal of the blind-ending branch should be considered in symptomatic patients and, according to previously published reports, it alleviates the symptoms⁽¹⁾.

Summary

We reported a case of a blind-ending bifid ureter associated with contralateral ureteral duplication, initially suspected in an abdominal ultrasound examination and afterwards verified in contrast-enhanced abdominal CT. Pathogenesis, sonographic appearance with differential diagnoses and clinical significance of this congenital anomaly are reviewed.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations which might negatively affect the contents of this publication and/or claim authorship rights to this publication.

Author contributions

Original concept of study: WŁ, BM. Writing of manuscript: WŁ, BM. Final acceptation of manuscript: WŁ, BM. Collection, recording and/or compilation of data: WŁ. Critical review of manuscript: BM.

e194 J Ultrason 2022; 22: e191-e195

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J Ultrason 2022; 22: e191-e195