CASE REPORT

Obstructed Right Upper Moiety in a Bilateral Partial Duplex Renal System in an Adult

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Summary

Duplex renal system, a common congenital anomaly of the kidney and urinary tract, is preferably identified through screening. For obstructed symptomatic duplications, prompt relief of obstruction to forestall renal parenchymal loss is recommended. We present a case of neglected obstruction of the right upper moiety in an adult female with discordant bilateral renal duplex system. A 49-year-old female with a 10-year history of recurrent right flank pain and fever presented for clinical evaluation and treatment. She had no such symptoms in early childhood. A diagnosis was made of obstructed right upper renal moiety in bilateral discordant renal duplication with complete moiety parenchymal destruction. She had right renal exploration with complete excision of the hydronephrotic sac and the grossly dilated moiety ureter down to the obstruction at the fusion of both upper and lower moiety ureters. Her postoperative period was uneventful. She is symptom-free and her urinary tract is structurally intact. Hitherto asymptomatic renal duplication can become symptomatic in early adulthood from intrinsic moiety ureter obstruction, suggesting a need for lifetime monitoring of persons with duplex renal systems. Late presentation with neglected symptoms, and consequent renal moiety destruction as seen here, is a result of the absence of such monitoring.

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Introduction

Duplication of the kidney with or without the ureter is reportedly the most common congenital anomaly of the kidney and the urinary tract (CAKUT). It has an incidence rate of 0.8% in the healthy adult population and 2–4% in patients investigated for urinary tract symptoms (1,2). In 40% of cases, the duplicate kidney moieties drain through separate ureters down to the urinary bladder as a complete duplex system, while in about 60% of cases the duplicate units fuse at a point to drain through a common ureter to the urinary bladder as a partial duplex system (3,4). It is possible for a complete duplex system to exist with a partial duplex system on the contralateral side in one individual, but such discordant bilateral duplications are rare.

Generally, duplex renal systems are asymptomatic and of no grave consequence, and so are underreported (2). However, a number of disorders requiring treatment have been observed in association with duplex systems: obstruction, reflux disease, and urinary stones are not

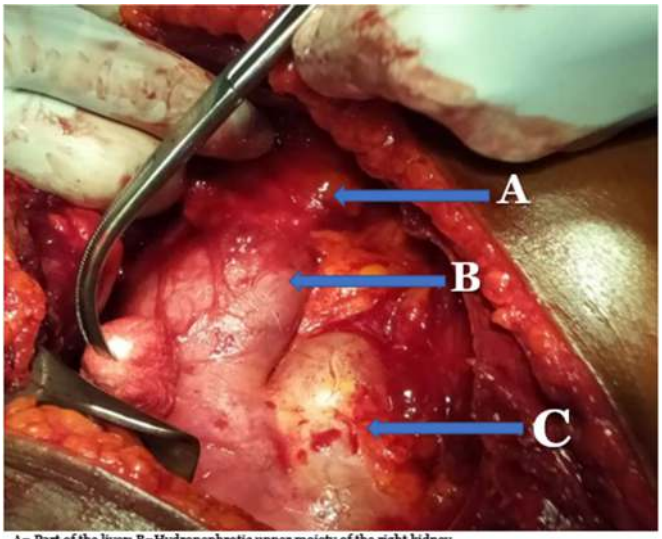
peculiar to but may be commoner in duplex systems (1,2). With respect to obstruction, the upper moiety may be at a greater risk (5). We present a case of neglected obstructed upper moiety of a right partial duplex system in an adult female with a discordant bilateral duplex renal system. The University of Nigeria Teaching Hospital Bioethics Committee approved this report (NHREC/05/01/2008B-FWA000024581RB00002323), and written informed consent for this report was obtained from the index patient.

Case report

Our patient was a 49-year-old woman who presented with a 10-year history of recurrent right flank pain. The pain was dull, intermittent, non-radiating, aggravated by coughing, and relieved temporarily by analgesics. She was able to perform her activities of daily living despite the episodes of pain. In addition, she experienced recurrent fever, but had no hematuria or flank swelling.

Clinical examination upon presentation revealed her body temperature was 36.5°C, blood pressure 110/80 mmHg, heart rate 72 beats per minute, and respiratory rate 20 cycles per minute. Renal angle tenderness and urine culture were positive, but her hemogram and serum levels of sodium, potassium, chloride, bicarbonate, and creatinine, as well as blood urea nitrogen levels were within normal limits. A transabdominal ultrasound scan showed bilateral duplex renal systems with hydroureteronephrosis of the right upper moiety. Computed tomography-urography corroborated the bilateral duplex renal systems with a grossly hydronephrotic, non-functioning, right upper renal moiety and a dilated right upper moiety ureter down to its fusion with the normal caliber lower moiety ureter just proximal to the urinary bladder. The left was a complete duplex system (Fig. 1).

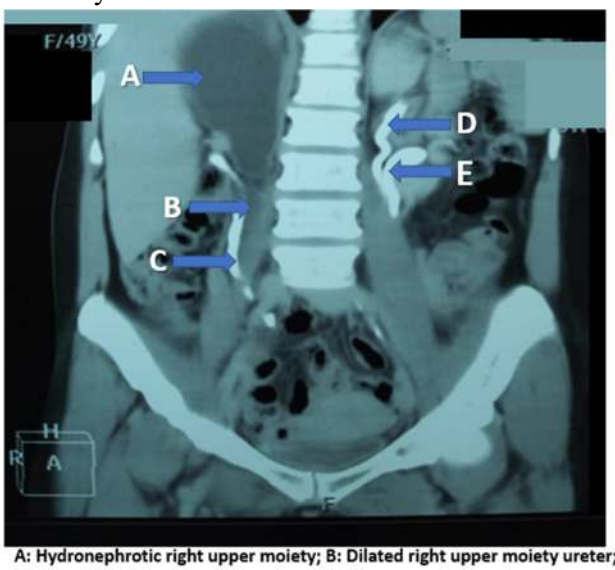
Cystoscopy revealed normal bladder mucosa with a single right ureteric orifice and double left ureteric orifices, confirming a complete duplex system on the left and a partial duplex system on the right.



A= Part of the liver; B=Hydronephrotic upper moiety of the right kidney
C= Normal lower moiety of the right Kidney

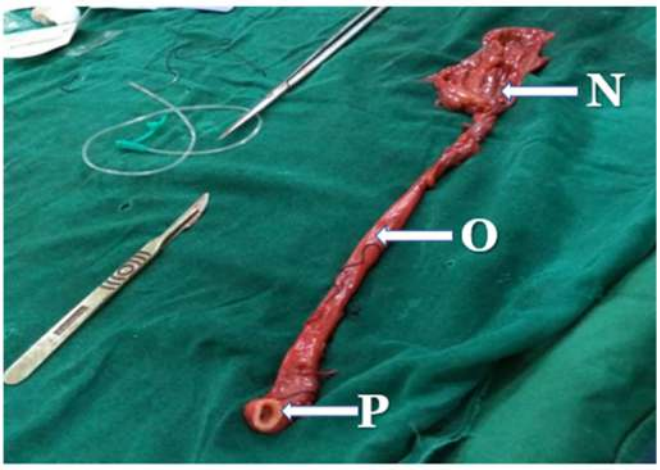
Figure 1. A complete duplex system on the left side

Surgical exploration showed the renal moieties on the right were within a single capsule (Fig. 2), supplied by single renal artery and vein. The hydronephrotic upper moiety was excised and the dilated upper moiety ureter was traced down to its fusion with the lower moiety ureter, ligated and excised. Histological report on the excised specimen (Fig. 3) was in keeping with the diagnosis. Her immediate postoperative period was uneventful. She has remained symptom-free for 2 years, and the other renal moieties have remained stable clinically.



A: Hydronephrotic right upper moiety; B: Dilated right upper moiety ureter;
C: Normal right lower moiety ureter; D: Left upper moiety ureter;
E: Left lower moiety ureter.

Figure 2. Renal moieties on the right within a single capsule



N= Hydronephrotic sac ;O= dilated ureter ; P= Hypertrophic ureteric muscle

Figure 3. Excised specimen

Discussion

Duplex renal system is a recognized developmental anomaly of the upper urinary tract. Usually asymptomatic and identified incidentally, it has no prognostic implications. However, when symptoms arise, they do so due to any or a combination of recognizable complications such as obstruction, reflux, and calculus formation, or of associated anomalies such as ureterocele and ectopic ureteric insertion (6).

Bilateral duplication of the renal system is less common than unilateral duplication, and a discordant bilateral duplication is even rarer (4,7). The index patient had a discordant bilateral duplication: while there is an asymptomatic complete duplex system on the left, the right has a symptomatic partial duplex system. This is in keeping with the hypothesis that the developmental processes culminating in duplication of the kidney and ureter are localized, although most probably on a background of chromosomal defect (8).

Non-calculous ureteric obstruction in a duplex renal system tends to occur at the confluence of the ureters or at the level of the insertion of the ureter in the urinary bladder (5). Such persistent significant obstruction usually causes progressive proximal dilation. Yo-yo refluxing between the two units of a partial duplication can also result in progressive proximal dilation (9). Absence of contrast in the ureter draining the non-functional upper renal moiety in the index patient excludes significant duplicate ureteric yo-yo refluxing. The observed obstruction in this patient must have been of long standing, considering the extent of destruction of the parenchyma of the corresponding renal moiety. Such late presentation with non-functioning corresponding renal moiety necessitating heminephrectomy has been reported in our environment, but mostly in children (10). In the absence of any significant renal tissue in the grossly hydronephrotic upper renal moiety, excision of the redundant hydronephrotic sac with the dilated ureter poses no major challenge. This procedure is better undertaken laparoscopically (11,12), but due to absence of facility and expertise, we adopted an extraperitoneal open-excision approach in this case.

Unilateral duplex renal systems are more commonly reported in females (7,13). Bilateral duplex renal systems may also be more common in females. When duplication occurs, observations suggest that the upper moiety may be at a greater risk of developing abnormalities than the lower moiety (5,12). Had the diagnosis been made earlier, this patient may have benefitted from dismembering the duplicate ureter and inserting the upper moiety ureter to the urinary bladder (5). However, her late presentation to appropriate care due possibly to a poor index of suspicion by her primary healthcare providers resulted in this unfortunate complete renal moiety parenchymal loss.

Conclusion

Hitherto asymptomatic duplex renal system may become symptomatic later in life. When identified without symptoms and without evidence yet of renal function compromise, the integrity of the renal moieties may have to be closely observed for life. Thorough assessment with a high index of suspicion is necessary in persons presenting with flank pains so as to promptly identify missed duplex renal systems, duplex system-associated disorders and thereby forestall the attendant renal moiety parenchymal loss.

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