Antenatally diagnosed upper moiety hydronephrosis in a child with ectopic ureter: A case report

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ABSTRACT

An ectopic ureter is any ureter, single or duplex, that does not enter the trigonal area of the bladder. In a duplex system, the ectopic ureter is inevitably the upper pole ureter. In males, the ectopic ureter always enters the urogenital system above the external sphincter. The majority of ectopic ureters are detected through prenatal ultrasound imaging, even if the specific diagnosis is not made until after birth. Radionuclide renal imaging remains the gold standard for renal functional assessment, and this is usually best provided by dimercaptosuccinic acid imaging. The goals of therapy are the preservation of renal function; elimination of infection, obstruction, and reflux; and maintenance of urinary continence. We report a case of a 6-month-old male child presenting with urinary tract infection. The child was diagnosed to have had a cystic dilatation of the right upper moiety on antenatal ultrasonography imaging.

Key words: *Antenatal hydronephrosis, Duplex kidneys, Ectopic ureter, Obstructive uropathy, Prenatal ultrasound, Vesicoureteral reflux*

An ectopic ureter is defined as any ureter, single or duplex, that does not enter the trigonal area of the bladder [1]. In a duplex system, this ureter is inevitably the upper pole ureter, presumably because of its budding from the mesonephric duct later than the lower pole with incorporation into the developing urogenital sinus [1]. In males, the ectopic ureter always enters the urogenital system above the level of the external sphincter or pelvic floor and usually enters into the Wolffian structures, including vas deferens, seminal vesicles, or ejaculatory duct. Male patients do not present with incontinence but rather with infection and pain of the affected organs (testicles and epididymis) [1]. Be it a single or duplex system, an ectopic ureter invariably leads to severe hydronephrosis reflecting the distal obstruction. This may lead to impaired renal development even to the point that the affected segment may be nonfunctional on functional assessment [2].

Rarely bilateral single-system ureter could be ectopic and may be associated with a severely hypoplastic bladder and bilateral renal abnormalities, typically dysplasia [3-5]. Some of these children may be considered to have bladder agenesis owing to the absence of a recognizable bladder structure, presumably because of the absence of bladder work *in utero* [1]. The majority of ectopic ureter can be detected on prenatal ultrasound imaging, even if the specific diagnosis is not made. The abnormality on prenatal scans prompts postnatal imaging, which will invariably determine the specific cause, lead to further studies, and permit an adequate characterization of the condition [1]. We report a case of a unilateral ectopic upper moiety ureter in a male child identified on antenatal scans and managed postnatally.

CASE REPORT

A 20-year-old pregnant female presented to the department for a normal checkup. Antenatal abdominal sonography done in the female revealed a healthy fetus at the gestation age of 21 weeks. The head circumference, abdominal circumference, and the biparietal diameter were within normal range. Antenatal scan repeated at the gestation age of 36 weeks revealed a slowdown of the fetal growth and was on the 10th percentile. The amniotic fluid was normal. The right kidney showed a well-defined anechoic cystic area in the upper pole measuring 31 mm×31 mm×32 mm (Fig. 1). The bladder was overdistended, and the left kidney appeared normal. The mother was advised to have a post-delivery ultrasonography of the newborn and to consult a pediatrician.

Postnatal ultrasonography of the 1-month-old male newborn revealed a cystic swelling in the upper pole of the right kidney measuring 24 mm×23 mm×23 mm. The lower half of the right kidney appeared normal. Upper moiety ureter was markedly dilated right up to the bladder. The child was started on oral antibiotics by the consultant pediatrician. At the age of 3 months,

a voiding cystourethrogram was done on the advice of a pediatric surgeon, which revealed a Grade V vesicoureteral reflux (VUR) on the right side. The child was continued on oral antibiotics. The child developed two episodes of febrile urinary infection within the next 3 months.

The child was brought to the pediatric urology services of our hospital following the second episode of febrile urinary tract infection. The child was poorly nourished and anemic. He was admitted and started on injectable antibiotics. Magnetic resonance urogram was done which revealed a complete duplex system on the right side, with the upper moiety ureter opening beyond the bladder neck into the urethra (Fig. 2a and b). The upper moiety ureter was dilated and tortuous. The child underwent cystoscopy which revealed the right upper moiety ureter opening beyond the bladder neck, proximal to the verumontanum (Fig. 3a). The lower moiety ureter on the right side opened into the trigone of the bladder. Radionuclide studies revealed a functioning right upper moiety, and hence, it was decided to preserve the upper moiety. The child underwent open right ureteroureterostomy with excision of the lower one-third of the right upper moiety ureter (Fig. 3b). The post-operative period was uneventful.

DISCUSSION

Congenital anomalies of the genitourinary tract are the most commonly identified malformations on an ultrasonography examination, with an incidence of 1–4 in 1000 pregnancies [6]. They represent 15–20% of all prenatally diagnosed congenital anomalies [7], and obstructive uropathies account for the majority of cases. It is well known that prenatal diagnosis improves the outcome of the affected child because of early recognition and treatment of these anomalies, preventing further renal damage and loss of renal function. With the introduction of modern ultrasound screening programs, about 60% of children having surgery for renal or urinary tract problems in their first 5 years of life are identified by prenatal ultrasound [8]. Duplex kidneys occur when the kidney is divided into two separate pelvicalyceal systems, or moieties, with either complete or partial duplication of the ureters [9]. The upper pole ureter may end either in the bladder or ectopically into the vagina or urethra. Ectopic ureter is frequently associated with dysplasia of the upper pole. The lower pole ureter drains laterally into the trigone of the bladder and is more prone to develop VUR [10].

Since the natural history of antenatally diagnosed duplex kidneys is uncertain, it is common practice to prophylactically treat the infant with antibiotics, to potentially reduce the risk of complications, while the diagnosis is confirmed and severity of any renal dysplasia is assessed. Whitten *et al.* [9] reported a 75% accuracy of ultrasound to diagnose duplex kidneys in the fetus. The authors concluded that accurate sonographic diagnosis of duplex kidneys in the fetus was possible in a dedicated multidisciplinary setting and that antenatal counseling and planning of postnatal care for the infant could be made with a high degree of certainty.

Radionuclide renal imaging remains the gold standard for renal functional assessment, and this is usually best assessed

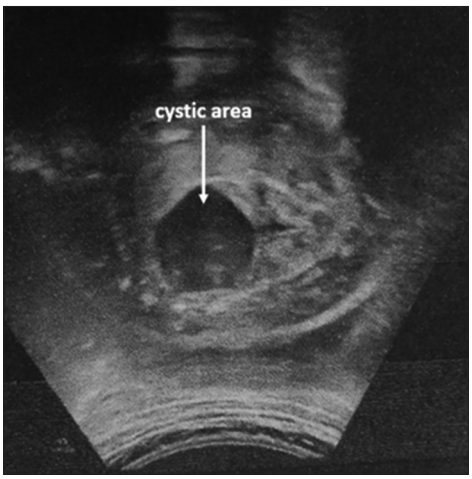

Figure 1: Antenatal ultrasonography at 36 weeks showing a well-defined cystic area in the upper pole of the right kidney measuring 31 mm×31 mm×32 mm

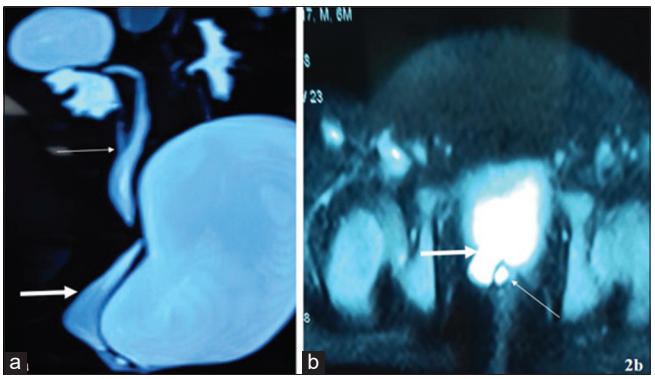

Figure 2: (a) Magnetic resonance (MR) urogram showing complete duplication of the right ureter, the thin arrow shows the normal sized lower moiety ureter, and the thick arrow shows the upper moiety dilated ureter, (b) MR program reveals bladder with the upper and lower moiety ureters at the level of trigone and bladder neck

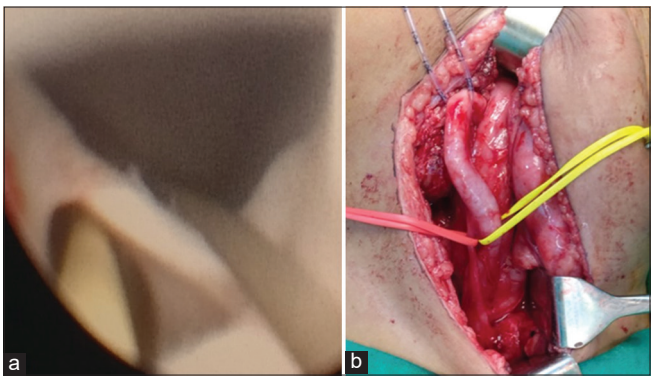

Figure 3: (a) Cystoscopy reveals the opening of the upper moiety ureter beyond the bladder neck. (b) Exploration reveals the normal sized lower moiety ureter and the dilated upper moiety ureter

by dimercaptosuccinic acid (DMSA) imaging [1]. The function of the affected upper pole is the principal focus; however, the function of the other renal moieties must also be determined, particularly if there is lower pole reflux or hydronephrosis of any unit [1]. The endoscopic evaluation of an ectopic ureter is a must as most cases of ectopic ureters would eventually come to operative intervention, and concurrent endoscopic evaluation

is important if not essential [1]. It may be difficult to localize an ectopic ureteral orifice endoscopically at times.

The goals of therapy should be clearly defined and factored into the clinical decisions. These goals are the preservation of renal function; elimination of infection, obstruction, and reflux; and maintenance of urinary continence [1]. For an ectopic ureter associated with a duplicated system, the primary concern is the preservation of functional renal parenchyma if at all possible. This goal is achieved by correcting obstruction and preventing reflux with its risks of renal parenchymal damage from infection [11]. Incase of an ectopic ureter, conservation of renal parenchyma and function is of prime importance and can be achieved by a common sheath reimplantation or ureteroureterostomy performed either at the upper end or lower end of the ureter. The decision-making for renal parenchymal preservation is largely empirical, and there are few objective criteria to indicate how much residual function is worth preserving [1]. The surgery can be performed both by open or laparoscopic means. Laparoscopic procedures do offer reduced morbidity with less post-operative pain, earlier return of gastrointestinal function, less hospitalization, and presumably a quicker return to work for the parents [12].

CONCLUSION

Radionuclide renal imaging remains the gold standard for renal functional assessment, and this is usually best provided by DMSA imaging. The goals of therapy are the preservation of renal function; elimination of infection, obstruction, and reflux; and maintenance of urinary continence. Upper pole removal using a partial nephrectomy or heminephrectomy of a duplex system is typically the preferred treatment when there is clearly no function in the upper pole and if there is concern about how effective a drainage procedure may be because of massive dilation.

REFERENCES

1. Dougal MC, Wein A, Kavoussi L, Partin A, Peters C. Campbell-Walsh Urology, 11th ed. Philadelphia, PA: Elsevier-Saunders; 2016. p. 3075.
2. Nerli RB, Ravish IR, Amarkhed SS, Reddy MR. Antenatally diagnosed unilateral hydronephrosis–long term follow-up. Indian J Urol 2005;21:59-63.
3. Koyanagi T, Tsuji I, Orikasa S, Hirano T. Bilateral single ectopic ureter: Report of a case. Int Urol Nephrol 1977;9:123-7.
4. Noseworthy J, Persky L. Spectrum of bilateral ureteral ectopia. Urology 1982;19:489-94.
5. Johnin K, Narita M, Jang KC, Wakabayashi Y, Yoshiki T, Okada Y. Bilateral single ectopic ureters with hypoplastic bladder: How should we treat these challenging entities? J Pediatr Urol 2007;3:243-6.
6. Grandjean H, Larroque D, Levi S. The performance of routine ultrasonographic screening of pregnancies in the eurofetus study. Am J Obstet Gynecol 1999;181:446-54.
7. Elder JS. Antenatal hydronephrosis. Fetal and neonatal management. Pediatr Clin North Am 1997;44:1299-132.
8. Bhide A, Sairam S, Farrugia MK, Boddy SA, Thilaganathan B, Bhide A. The sensitivity of antenatal ultrasound for predicting renal tract surgery in early childhood. Ultrasound Obstet Gynecol 2005;25:489-92.
9. Whitten SM, McHoney M, Wilcox DT, New S, Chitty LS. Accuracy of antenatal fetal ultrasound in the diagnosis of duplex kidneys. Ultrasound Obstet Gynecol 2003;21:342-6.
10. Mackie GG, Stephens FD. Duplex kidneys: A correlation of renal dysplasia with position of the ureteral orifice. J Urol 1975;114:274-80.
11. Churchill BM, Sheldon CA, McLorie GA. The ectopic ureterocele: A proposed practical classification based on renal unit jeopardy. J Pediatr Surg 1992;27:497-500.
12. Nerli RB, Vernekar R, Guntaka AK, Patil SM, Jali SM, Hiremath MB. Laparoscopic hemi/partial nephrectomy in children with ureteral duplication anomalies. Pediatr Surg Int 2011;27:769-74.

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