Case Report

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The Weigert – Meyer Law of Ureteral Duplication– A Rare Pathological Entity

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ABSTRACT

Ureteral duplication is a developmental anomaly leading to many complications in children. The Weigert-Meyer law states that 'In a complete ureteral duplication, the ureter whose orifice is more medial and caudal reaches the upper moiety and the other ureter whose orifice is more lateral and cephalad reaches the lower renal moiety'. This law has been observed universally in cases of ureteral duplication. The possible complications of double ureter and duplex system include obstruction, lithiasis, ureterocele and vesico-ureteral reflux. Hence it is important to recognize this entity for early intervention and to prevent complications. We are publishing one such rare case in a 10 month old child with review of literature.

Keywords: *Weigert-Meyer Law, Ureteral Duplication, Ectopic Ureter*

Introduction

The Weigert-Meyer law was given the name following discovery by Carl Weigert and later Meyer. They observed this law to be true in all cases of ureteral duplication. [1] The Weigert- Meyer law is applicable to a completely duplicated collecting system. It states that the ureter arising from the lower pole moiety runs a short intramural course and inserts into the urinary bladder more superiorly and laterally. This predisposes the ureter to reflux.[2,3] The other ureter arising from the upper pole of the kidney inserts more inferiorly and medially and is prone for ureterocele and obstruction.[2]

The prevalence of double ureter is said to be 0.1% to 3%.[4] It is observed that due to this malformation, the complications are more common in girls than in boys. The entity is underreported with four cases from older literature and seven recently added cases.[1]

Case Report

A 10 month old female child was brought to the hospital by her mother with complaints of frequent urinary tract infections and failure to thrive. On external examination, the external genitalia were normal. There was no suprapubic tenderness on abdominal examination.

Routine laboratory investigations showed anemia of microcytic, hypochromic type, raised total leucocyte at 14,300 cells/cumm normal serum creatinine at 0.5 mg/dl. Urine routine and microscopy showed no abnormality.

Ultrasound of the abdomen showed a severely dilated and tortuous right ureter. The caliber of the distal ureter was 1.5 cm. Severely dilated pelvicalyceal system was seen in the upper pole region of the right kidney which was likely a separate upper moiety. The distal opening of the dilated right ureter was seen beyond the urinary bladder likely in the vestibule of the vagina or the urethra. The calyceal system was not dilated in the lower moiety. The left pelvicalyceal system and urinary bladder was normal.

MRI Urography showed a duplex kidney on the right side with duplication of the renal calyces, pelvis and the entire ureters. The upper renal moiety drained into the dilated ureter which measured 1 cm in diameter. The dilated ureter had an ectopic opening below the level of the urinary bladder into the vaginal introitus. There was significant pelvicaliectasis of the upper renal moiety with thinning of the renal cortex, measuring 3.5 mm in maximum thickness.

The lower renal moiety appeared normal with the ureter of the same draining into the urinary bladder. The left kidney and ureter appeared normal.

Furthermore, a dimercaptosuccinic acid (DMSA) scan was done which showed both the kidneys in normal location. The cortical function of the upper moiety of the right kidney was significantly reduced (7%) while the lower moiety of the right kidney showed good cortical function (93%).

Right heminephrectomy was performed and the gross examination showed right kidney with attached ureter together measuring 4 x 3 x 2.5 cm. External examination of the kidney was unremarkable.

Cut surface of kidney showed loss of normal cortico-medullary differentiation and dilated pelvi-calyceal system suggestive of hydronephrosis. Ureter measured 5.5 cm in length, showed features of hydroureter. (Fig 1).

Microscopic sections from the kidney revealed thickened capsule, with few normal glomeruli and some showing features of focal segmental glomerulosclerosis. Tubules were atrophic and decreased in number.

Interstitium showed lymphoid aggregates and lymphoid follicles with germinal centres. Blood vessels were normal. (Fig 2). Pelvis and ureter showed thinning and loss of transitional lining epithelium suggestive of hydroureter. (Fig 3).

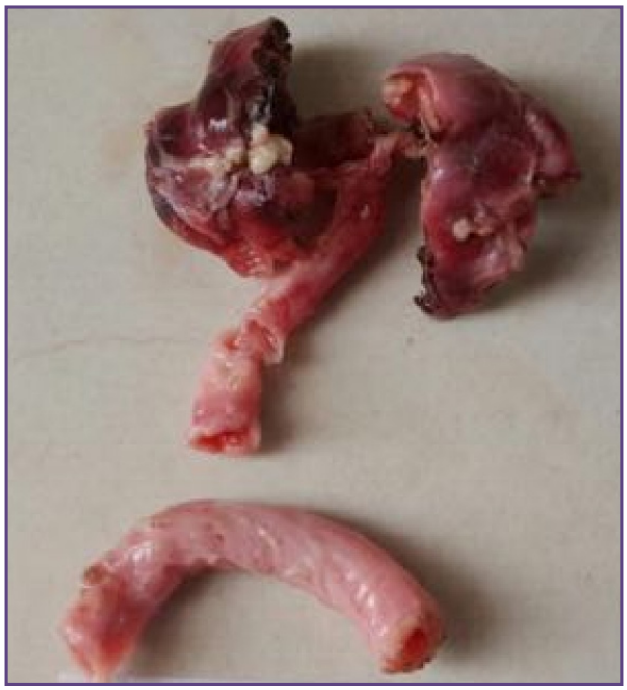

Fig. 1: Gross of right heminephrectomy showing loss of cortico-medullary differentiation and dilated pelvi-calyceal system with hydroureter.

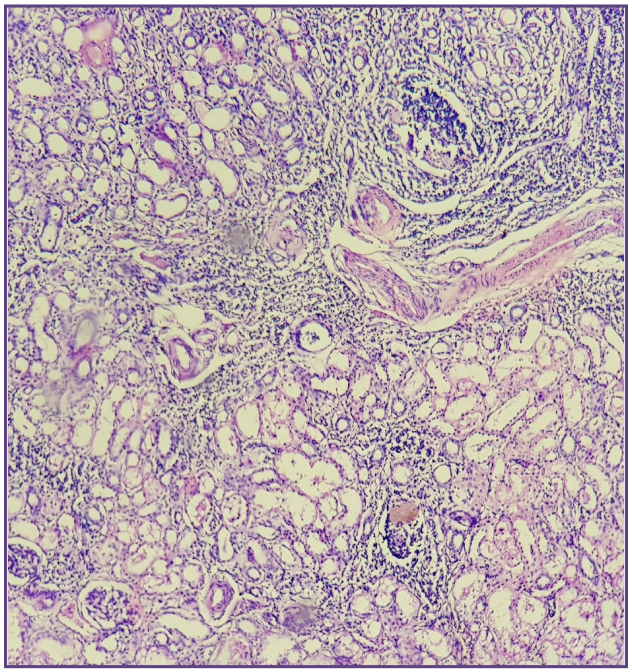

Fig. 2: H & E, 10X, kidney showing features of interstitial nephritis.

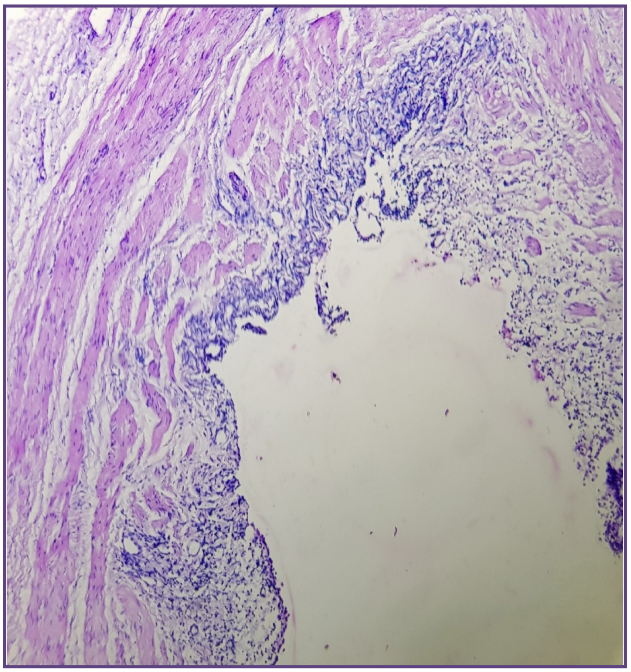

Fig. 3: H & E, 10X showing dilated ureteral cavity, lined by thinned out urothelium.

Discussion

Renal duplication or ureteral duplication is the most common urinary tract anomaly.[3] According to the Weigert-Meyer rule, the orifices of the ureters draining the upper pole of the kidney open inferior and medial to the orifice draining the lower pole of the kidney. This was seen in 85% of subjects with complete ureters.[3] The same finding was observed in our study.

During embryogenesis and development, formation of 2 ureteral buds followed by failure of contact of one of the buds with the metanephrogenic blastema results in the double ureteral orifice formation. This ureteral duplication may be complete or incomplete and is genetically determined by an autosomal dominant trait with incomplete penetrance.[3]

At about 4-6 weeks of gestation, the distal mesonephric duct gets absorbed into the urogenital sinus which results in the separation of the ureteral bud and the mesonephric duct orifices.[2] It is reported that this condition is seen in females more commonly with the female to male ratio of 6:1.[2]

In such cases of duplex kidney with double ureter, the lower moiety is most commonly affected by vesico-ureteral obstruction and reflux, whereas the upper moiety is most commonly affected by hydronephrosis. The same finding was observed in our case.

Normally, the ureteral orifices migrate laterally and cranially while the mesonephric duct orifice migrates downward and medially.[2] In case the ureteral bud has had a cranial origin, it migrates downward and medially along the with mesonephric duct and opens along the course of the mesonephric duct.[2] Continuous incontinence occurs if this opening is distal to the internal ureteral sphincter.[2]

It is important to know whether the ectopic ureter shows reflux before planning a partial nephrectomy since it is difficult to dissect the ureter distally if it is inserting beyond the internal ureteral sphincter.[2] Bladder neck diverticula are commonly associated with duplex moieties but they usually disappear by eight years of age.[2] In our case, the ureter of the upper renal moiety appeared dilated while the ureter of the lower renal moiety appeared normal.

It was stated by Curry NS that reports of ectopic duplicated ureters draining into diverticula were likely to be partially filled dilated distal ureters.[2] A case report of a bladder diverticulum arising from the lateral wall of the bladder adjacent to an ectopic ureter was reported by Michelotti B et al.[2] On urogram, prevalence of partial duplication of ureters is three times more common than complete duplication of ureters.[2]

According to Hascalik and associates, ureteral duplication could be genetically determined by an autosomal dominant trait with incomplete penetrance.[4] While on the other hand, Bruno and colleagues said that ureteropelvic obstruction is more common in the presence of a duplex kidney and this could be inherited as an autosomal dominant pattern.[4]

The lower pole system is dominant in most of the cases of double ureter, hence pelvi-ureteric junction obstruction is affecting the lower moiety as compared to the upper moiety.[4] This can be associated with anomalies of the renal system.[4]

Maldevelopment of the valve mechanisms results in vesicoureteral reflux involving the lower pole of the duplex system.[4] On the other hand, hydronephrosis is seen involving the upper pole of the duplex system.[4]

In a duplex renal system, massive dilatation of the upper pole moiety is either due to vesicoureteral reflux or error in development as suggested by Mahajan and associates.[4] However, in our case, the massive dilatation was due to hydronephrosis.

Conclusion

The Weigert-Meyer Law is an unusual developmental anomaly and has to be considered in the differential diagnosis in children presenting with repeated UTI for timely intervention and management.

References

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