Functional medicine

Renal duplication with ureter duplex not following Meyer-Weigert-Rule with development of a megaureter of the lower ureteral segment due to distal stenosis – A case report

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ABSTRACT

Meyer-Weigert-Rule predicts the draining pattern of duplex ureters in bipolar renal duplications. The upper pole is normally seen as ectopic and therefore dysplastic due to obstruction, whereas the lower pole is related to vesicoureteral reflux. In our case, this rule is violated with uncrossed ureter duplex and a dysplastic lower pole in connection with obstruction.

Introduction

Duplication of the upper urinary tract belongs to the most common congenital urological anomalies. Diagnosis of bipolar duplex kidneys with an upper pole and a lower pole is usually made either on prenatal sonograms or after birth during urologic evaluation. The ureteral drainage pattern is normally governed by the Meyer-Weigert rule, predicting that the ureter of the upper part usually drains inferomedially into an ectopic location, which commonly leads to obstruction. The lower ureter part usually drains superolaterally to its normal insertion in the trigon. This is commonly associated with vesicoureteral reflux as a result of a lack of sufficient submucosal tunnel that leads to pyelonephritis, scarring and a so-called “drooping lily” appearance on IVPs. Deformation of a duplicated upper urinary tract is based on the development of two ureteral buds. The constant relationship between the upper (ectopic) and lower (orthotopic) part was discovered by Carl Weigert (1877) and Robert Meyer (1946). As part of the embryologic development, the ureters and their orifices complete a 180-degree clockwise rotation in their longitudinal axis. The proximal bud, which was higher and draining the upper system, is now below the first bud – this way the two ureters cross each other (Meyer-Weigert-Rule). This paper aims to present an unreported case, in which the lower renal duplication was not associated with a vesicoureteral reflux as the Meyer-Weigert rule requires. Contrary, the lower renal part was obstructive.

Case presentation

The female 23-year-old patient suffers from symptoms of recurrent urinary tract infections as well as left sided sporadic flank pain for at least three years. Unfortunately, the patient does not know whether prenatal and/or postnatal diagnoses have been performed. Furthermore, extended diagnosis of the flank pain was not made. Due to the unknown variables, MRI of the abdomen was performed first (Fig. 1), which revealed a duplication of the left kidney with ureter duplex. In this case, the upper renal part showed a normal configuration with no obstruction of urine drainage, whereas the lower renal part presented a megaureter and a dilated renal pelvis. In micturating cystogram (MCU) a typical bladder contour was observed with no vesicoureteral reflux (Fig. 2).

The findings were discussed with the patient and afterwards, the decision was made to perform a left lower pole heminephroureterectomy. Beforehand, a Cystoscopy revealed a normal configured right ostium but there was no ostium detectable on the left side. In the estimated area of the right ureter orifices only a mucosal wrinkle could be detected most likely due to the imprint of the megaureter. Since heminephroureterectomy of the lower pole was already indicated a forced diagnostic was not performed. Intraoperatively, it became obvious that the lower part was dysplastic, which was most likely due to the ongoing elevated pressure. Furthermore, in open surgery the lower part inserted inferolaterally into the bladder.

Follow up was performed six month after surgery including medical history, physical examination and ultrasound of the abdomen. No

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Further events of urinary tract infections as well as sporadic flank pain were reported. Ultrasound and physical examination showed a primary wound healing and a normal configured upper moiety without signs of urinary retention.

Discussion

This unusual case demanded a literature review of the following topics: renal duplication, duplex ureter and the Weigert-Meyer rule or Meyer-Weigert law with only one adequate match. Therefore, to the best of our knowledge, we are presenting this case in which the ureter of the lower renal part inserted inferolaterally without vesicoureteral reflux. A small number of exceptions to the Meyer-Weigert rule have been published that deal with a more caudal insertion of the ureter of the lower renal pole.\textsuperscript{4,5}

Jain et al. presented a case report of a 10-year-old male with intermittent periumbilical pain. The lower pole was dysplastic and the ureter was massively dilated, whereas the upper pole presented no dilatation. Intraoperatively the lower part could not be cannulated which leads to the assumption of a distal stenosis.\textsuperscript{3} In our reported case, the upper pole ureter was at the right place, and the lower pole ureter presented as a megaureter as described by Jain et al. So far there is no adequate explanation for those two rare cases.

Fig. 1. MRI of the abdomen of a twenty-three-year-old patient born with renal duplication of the left side and ureter duplex. A Megaureter of the lower renal moiety (*). B Normal configured upper renal moiety (white arrow) with no drain obstruction. C and D demonstrate a different view of the left Megaureter (white arrow) and the normal configured upper renal moiety (*).

Fig. 2. Micturating cystogram (MCU) showing an even delimited bladder without any sign of vesicoureteral reflux.
Conclusion

Although the Meyer-Weigert rule is applicable to most cases of bipolar renal duplication, several case reports point out exceptions to this rule, especially with focus on the lower renal part. Pediatric radiologists and urologists should be alerted as this newly introduced variation may require a tailored management, if the lower renal part is obstructive.

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