Unusual Pattern Of Pyeloureteral Duplication Associated With Vaginal Ectopic Ureter In A 12-year Old Girl Identified By Magnetic Resonance Urography: A Case Report

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Article ID: WMC001155
Article Type: Case Report
Submitted on: 12-Nov-2010, 01:48:58 PM GMT Published on: 13-Nov-2010, 05:44:10 PM GMT
Article URL: http://www.webmedcentral.com/article_view/1155
Subject Categories: UROLOGY
Keywords: ectopic ureter, urinary incontinence, voiding dysfunction, pyeloureteral duplication, pediatric urology, magnetic resonance urography


Source(s) of Funding:
No funding was obtained during preparation of this manuscript.

Competing Interests:
The authors declare that they have no competing interests.
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Abstract

Introduction: The prevalence of ectopic ureter (EU) is low and it is usually associated with urinary system duplication. In such cases, incontinence is a major complaint. Case Presentation: We present a case of a female patient that was diagnosed with ectopic ureter opening at the posterior vaginal fornix along with a duplicated pyeloureteral system after a regular diagnostic work-up that turned out inconclusive and several years of observance. Magnetic Resonance Urography (MRU) consisted of a left tortuous ectopic ureter opening at the posterior vaginal fornix. The pattern of drainage was oddly inverted and influenced the chosen treatment, which consisted of transureteroanastomosis together with ligation of the ectopic ureter. Incontinence fully resolved after surgery. Conclusion: Even though Intravenous Pyelogram (IVP) is highly sensible to detect renal duplication when an ectopic ureter is draining a dysplastic upper segment of a duplex kidney, Magnetic Resonance Urography should be regarded as a better diagnostic tool in such cases.

Case Report(s)

A 12-year-old girl was referred to us for an urologic evaluation because of urinary loss despite normal voiding function. The continued losses had begun since the age of 3, when the patient first developed voiding control. At the beginning the family thought it would disappear, but as symptoms failed to improve, they searched for medical care at the age of 6. An ultrasound study was performed to investigate any urinary tract malformations, but at the time it did not reveal any abnormalities. A cystoscopic examination was also performed and was completely normal. Blood analysis, urinalysis and microbiologic cultures were unremarkable. Renal function markers were within limits of normality.

The ureter misplacement is usually deviated laterally to the normal insertion site, even though it can be located anywhere in the pelvis. When the misplacement is located underneath the sphincter, urinary incontinence is usually associated and consists of constant dripping of urine, with or without an associated pyuria. Possible locations are at the bladder neck, in the urethra or through the remains of the Wolf ducts (ejaculatory duct, seminal vesicles, vas deferens) in men or Müller ducts (urethra-vaginal septum vestibule, vagina, uterus) in women. The insertion site of the ureter has a good correlation with the degree of renal dysplasia, so that the further from the trigone the ureter is located, the more renal dysplasia is found.

As the symptoms were minor and did not interfere much with the patient’s development and quality of life, clinical observation was chosen, hoping that this affection would be psychologically related. Two years later, however, it seemed that as the patient grew older the anxiety grew bigger and they were willing to have this problem solved at any cost. A Intravenous Pyelogram (IVP) (illustration 1) was performed to evaluate the upper urinary tract but it turned out without significant findings. An
Eletroneuromyography along with an Urodynamic Study were carried out in order to identify some sort of neurological dysfunction, but they were again unrevealing. Finally, a Magnetic Resonance Urography (MRU) (illustrations 2 and 3) was performed and revealed a left tortuous ectopic ureter opening at the posterior vaginal fornix.

A surgical intervention was then planned in order to reinsert the ectopic ureter in the bladder, stopping the vaginal urinary loss, once no double system was observed. However, during the procedure, a radioscopic image was obtained (illustration 4) and a duplicated left ureter was accidentally found. A further manual evaluation (illustration 5) revealed that the medial ureter originated from the upper kidney pole and was draining in a tortuous tract into the vagina, whereas the lateral ureter had its origin in a mildly atrophic lower pole and had a normal insertion at the vesical trigone. Even though it was quite unusual, it was in accordance with the previous normal exams.

A transureteroanastomosis was then pursued and the ectopic ureter was ligated. The patient was left with vesical stenting the first day after surgery because mild hematuria was noted. The complaints of urinary loss vanished immediately. At the third post-operative day the stenting was withdrawn and the patient was completely satisfied. At a 6-month follow-up routine examination the patient appeared healthy and the symptoms never recurred.

Discussion

Urinary incontinence in girls due to ectopic ureter is an uncommon disease and most cases are associated with peloureteral duplication. The classical presentation of this affection is continuous urine loss together with normal micturition. It is established that in order to cause urinary incontinence, the ureter must insert below the external urethral sphincter in positions such as the vagina, perineum or introitus. (4) In our case it was located at the posterior vaginal fornix, which is in accordance with the findings of urinary incontinence.

When a ureter has a displaced insertion leading to incontinence, it is expected that it should be draining a kidney, or segment of a kidney, that is hypoplastic and dysplastic. This association is believed to reflect failure of an abnormal ureteral bud to induce differentiation of the metanephric blastema into functional renal tissue (3), however exactly the opposite was observed in this reported case. In our literature review we did not find any similar case nor explanations for such findings.

The diagnosis of EU is usually made by a typical incontinence history associated with signs of renal exclusion at IVP. (1) Even though IVP is highly sensible to detect renal duplication when an ectopic ureter is draining a dysplastic upper segment of a duplex kidney by showing displacement of the ipsilateral lower pole, in our patient it did not occur. This may be explained by the fact that only a mild degree of hypoplasia at the lower pole was observed and it had a normal functioning ureter.

Other diagnostic tools as ultrasonography and DMSA renal scans may be useful in some cases, however the former fails to show the system duplicity and the latter usually fails to detect dysplastic renal tissue. (4),(5) Contrast CT Urography with 3-D reconstruction of the images has also been applied for this matter, however, it has the disadvantage of very high radiation exposure and risk of allergic reaction to the contrast agents. (6),(7)

Despite the lack of consent of which method is the most cost effective tool for EU diagnosis, we believe that MRI is the best modality of investigation in such cases because it’s a single radiation-free method that is able to detect at once the presence of dysplastic tissue, duplicated urinary system and sometimes the tortuous ureteral tract. (8) The disadvantages are the high cost and the lack of broad access to it. However it is our perspective the MRU will be considered the gold standard diagnostic tool for EU in the near future.

Once the diagnosis is confirmed and the opposite kidney is normally functioning, the procedure of choice is to remove the dysplastic kidney or segment and the ectopic ureter. (2) Nevertheless, in our case the chosen treatment was transureteroanastomosis and ligation of the ectopic ureter, because the degree of hypotrophy found in the lower unit was mild and a normal ureter was draining from it. Furthermore, we proposed a long-term follow-up to determine whether the dystrophy will evolve and bring any consequence, but as symptoms immediately resolved, the case was successfully solved and no recurrence is expected.

Conclusion

Even though IVP has a high sensibility in detecting renal duplication when an ectopic ureter is draining a dysplastic upper segment of a duplex kidney, MRU should be considered the best investigation tool in such cases, especially when the initial diagnostic workup turns out unrevealing and symptoms fail to
improve.

Abbreviations(s)

- Ectopic ureter (EU)
- Magnetic Resonance Urography (MRU)
- Intravenous Pyelogram (IVP)
- Computed Tomography (CT)

Acknowledgement(s)

Authors contribution(s)

EPM conducted the patient, participated as the first assistant during the surgical procedure and was a major contributor in writing the manuscript. FECF help conducting the patient, participated as the second assistant during the surgical procedure and was a major contributor in writing the manuscript. BRSF participated as second assistant during the surgical procedure and was responsible for collecting data and consent from the patient. MAS was responsible for the literature review and was a major contributor to writing the manuscript. AGS was the intellectual mentor, the surgeon who led the team and the reviser of the manuscript and all collected material. All authors read and approved the final manuscript.

References

Illustrations

Illustration 1

Normal Intravenous Pyelogram

Illustration 2

MRU revealing a left tortuous ectopic ureter opening at the posterior vaginal fornix
Illustration 3

MRU revealing a left tortuous ectopic ureter opening at the posterior vaginal fornix

Illustration 4

Radioscopic image performed during surgery
Illustration 5

Manual exposure of the duplicated ureteral system
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