Endourology

Single-system orthotopic ureterocele with calculus masquerading as a bladder tumour – A case report and review of literature

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Introduction

As cystic dilations due to a congenital wall weakness of the distal ureter,¹ ureteroceles tend to be found in autopsy with an incidence of 1/500 to 1/4000. According to literature solitary stones within such a single-system ureterocele are quite common with a prevalence of about 4%–39%.² However the diagnosis of such a situation is largely based on typical diagnostic signs such as the cobra-head-sign or the changing size of the ureterocele during sonographic evaluation as well as the direct proof of the ureterocele in cystoscopy and MRI-/CT-imaging. Still there are rare situations in which the diagnostic findings aren’t as obvious to interpret as one might think. Such a case which is of interest to each clinically working urologist we present at hand. As to our knowledge there has only one such case been reported in literature so far.³

Case presentation

A 59-year old Patient was transferred to us by an outward Radiology after diagnosing a calciﬁed right-sided bladder tumour (approx. 2cm in diameter) with a consecutive ureteral obstruction and a therefore massively dilated right-sided distal ureter in a CT-scan. The patient himself initially consulted his primary physician due to right-sided lower abdominal pain so that the formerly mentioned CT-scan was performed (see Figs. 1 and 2). Further investigation by cystoscopy within our clinic conﬁrmed that the tumour was highly suspicious of being a urothelial carcinoma. Therefore a transurethral resection of the tumour was planned. Unfortunately no urinary cytology was acquired.

Intraoperatively cystoscopy once again confirmed the highly suspicious ﬁndings to most likely be an urothelial carcinoma and therefore transurethral resection was begun. However, after the ﬁrst slices were made, the tumour revealed itself to be an intravesically prolapsed ureterocele containing a calculus instead of being a urothelial carcinoma. The calculus was extracted under slight bipolar resection of the ureterocele-wall as well as lithotripsy and ﬁnally a Double-J-Stent was implanted.

In the postoperative follow-up period the Double-J-Stent was removed 4 weeks after surgery and sonographic as well as CT-based controls showed unsuspicious renal drainage on the right side with a residual dilatation of the distal right ureter and pelvis.

Histological analyses of the resected tissue revealed no malignancy and the analysis of the extracted calculus showed it to be 100% Calcium-Oxalate-Monohydrate (Whewellit).

Discussion

As the incidence of ureteroceles in adults is very low and diagnosis is often delayed, there is scarcely literature about an adequate diagnostic algorithm as well as the proper treatment available.⁴

Most recommendations derive from pediatric surgery and suggest an individualized diagnostic approach with e.g. MRI/CT-scans, cystoscopy, cystography, sonography and isotope renogram ﬁtted separately for each patient as there is no “one ﬁts them all”-approach.⁵ This is of special importance as typical diagnostic signs such as the cobra-head-sign or the changing size of the ureterocele during sonographic evaluation might not be present in every patient. As our case at hand shows, even a combined diagnostic approach with CT-scan, sonography and cystoscopy can lead each urological practitioner astray.

Depending on the diagnostic ﬁndings further treatment should be equally ﬁtted for each patient and focus on the prevention of further complications such as vesico-ureteral reﬂux, recurrent UTIs and loss of

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renal function. In more complex cases with duplex renal collecting systems including blindly ending ureters, hypoplastic renal segments which are drained by the ectopic ureter or infrasphincteric ureteral orifices ablative surgical approaches including nephrectomy, heminephrectomy and ureterectomy are mostly considered to be the therapy of choice. However, it is commonsense, that ureteroceles in adults tend to be less complicated and are more likely to be small and single-system with orthotopic ureteral orifices. The proper treatment, as it is in children with single-system orthotopic ureteroceles as well, is accordingly transurethral incision/resection of the ureterocele and stenting the ureter as we did with our patient.

Still the main complication of this technique seems to be vesico-ureteral reflux. We therefore recommend regular follow-up by urothrocystography, which we will perform on our patient during one of his next consultations. Furthermore follow-up after transurethral incision of a ureterocele should include CT-urographic controls as well as sonographic controls.

As it comes to the long-term outcome after endoscopic de-roofing or puncture of orthotopic and even complicated, ectopic, ureteroceles recent publications suggest that the paradigm shifts slowly towards endoscopic treatment as it is safe and long-term effective. Jawdat et al. just published a summary of 78 cases which had a median follow-up of 12 years after endoscopic puncture of orthotopic as well as ectopic ureteroceles. It shows that in the group of patients with orthotopic ureteroceles who had undergone an endoscopic puncture of the ureterocele, there was scarcely additional surgery necessary. Within the group of patients suffering of an ectopic ureterocele more patients required secondary surgery, but there still was no statistical significance between those two groups (p = 0.716) supporting Chowdhary et al.’s statement that even those patients can be managed endoscopically. Furthermore a significant number of patients who suffered of a new onset of vesico-ureteral reflux after the procedure showed spontaneous regression.

According to Jawdat and Chowdhary non-functional, undrained, renal segments as a complication of ectopic ureteroceles should no longer be an indication for major surgery such as (partial-)nephrectomy.

Conclusion

The case presented at hand shows that due to its rareness ureteroceles in adults pose a diagnostic and therapeutic challenge to each urologist and might be misinterpreted as a more common pathology such as a urothelial carcinoma of the bladder. The diagnostic algorithm should therefore be multimodal including sonography, CT-/MRI-scans,
cystoscopy and in some cases an isotope renogram and cystography. The therapy of choice for orthotopic single-system ureteroceles is transurethral incision, de-roofing and Double-J-Stenting of the ureter. As it comes to more complicated, ectopic, ureteroceles endoscopic incision should be evaluated as first-line treatment as well as the latest data suggest that major surgery even in these cases is rarely necessary and can therefore be avoided.

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### Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2018.09.002.

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