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CASE REPORTS

URETHRO-FISTULAR CALCULUS AFTER EXSTROPHY-EPISPADIAS SURGERY

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A very unusual case of a huge calculus, extending from the urethra into a dorso-lateral urethro-cutaneous fistula is reported in an 8-year-old African boy who underwent primary bladder closure for exstrophy at the age of 16 months, followed by epispadias surgery 6 months later. The patient was lost to follow-up after exstrophy-epispadias surgery for 6 years. At 8 years of age, he presented with a fistulized distal urethra forming a diverticulum-like structure containing a round stone with a diameter of 28 mm. The fistula was opened, the stone removed and urethroplasty was performed. Such urethral calculus has, to our knowledge, not been previously reported in exstrophy-epispadias (EE) surgery. This case shows the necessity and importance of careful long-term follow-up, as such a severe complication may develop well after primary surgery.
Key words: child, urethro-cutaneous fistula, calculi, exstrophy, epispadias.

INTRODUCTION

Distal urethral calculi, although fairly common in children from developing countries[1], are rarely larger than 10 mm. Such calculi are mostly composed of struvite and uric acid[2]. We herein report the case of an 8-year-old Senegalese boy.

CASE REPORT

A Senegalese boy underwent primary bladder closure for exstrophy at the age of 16 months, followed by Cantwell-Ransley epispadias repair[3] 6 months later at our institution. At that time the epispadias repair was complicated by a urinary tract infection associated with a dorso-lateral penile urinoma evolving into a urethro-cutaneous fistula which closed spontaneously.

Six years later, the patient was referred for planned surgical management of bilateral asymptomatic vesicoureteral reflux (VUR) diagnosed on routine follow-up. Urinary tract infection was never observed. Local examination revealed an asymptomatic voluminous hard mass on the right dorso-lateral side of the penis, 3 cm proximal to the meatus, associated with a fistula. On voiding cystourethrogram a large fistulizing diverticular structure containing a calcified mass could be observed in the distal urethra (Fig.1). Abdominal ultrasound demonstrated normal sized kidneys and isotope renography showed adequate renal function.

Preoperative urethroscopy showed mild distal urethral narrowing with a urethral calculus partially impacted within the urethral fistula (Fig. 2). Surgical opening of the cutaneous side of the fistula allowed the removal of a second intrafistular calculus measuring 28 mm in diameter (Fig. 3, 4). Beyond the diverticulum-like pouch, and after progressive fistula dissection towards the urethra, the smaller calculus of 11 mm in diameter already seen at urethroscopy was extracted and the urethra reconstructed in two layers over a Ch. 10 urinary catheter with absorbable 7/0 suture. The mineral composition of the calculi was 40% ammonium urate and 60% struvite.

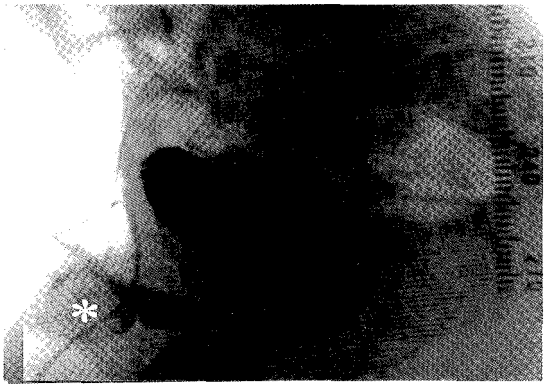

Fig. 1: Preoperative voiding cystourethrogram showing urethro-cutaneous fistula with a diverticulum-like structure containing a calcified mass (*).

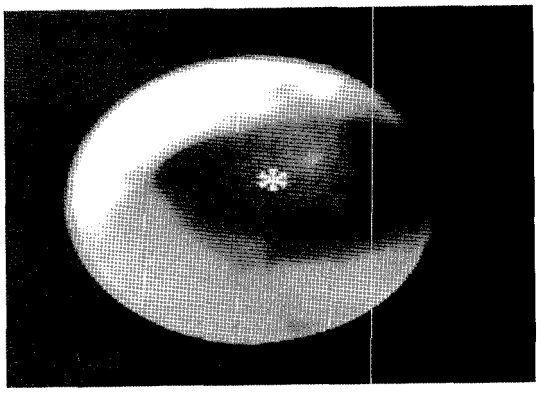

Fig. 2: Preoperative urethroscopy showing a smaller calculus (*) impacted in the fistula.

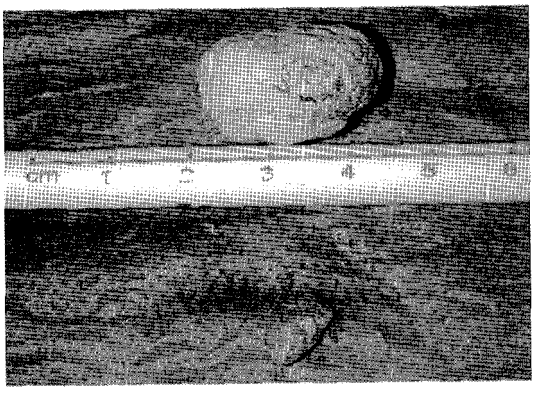

Fig. 3: Macroscopic aspect of the 11 and 28 mm sized ammonium urate - struvite stones.

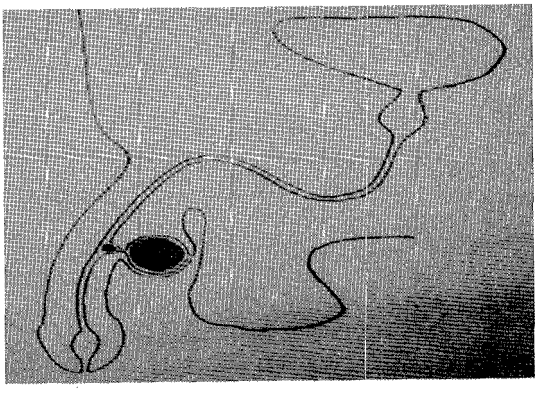

Fig. 4: Schematic drawing of intraoperative status.

At 8 months follow-up, the patient was asymptomatic and urethrocystoscopy demonstrated the absence of urethral stricture or fistula. Furthermore, the earlier diagnosed bilateral VUR was surgically corrected, bladder augmentation and continent Mitrofanoff stoma were performed.

DISCUSSION

Urethral lithiasis is a classical complication in patients operated for EE[4,5]. In children with EE, postoperative indwelling urinary catheters and urinary stasis due to a tightly reconstructed bladder neck are well-known causes of infection-linked calculi[4,6]. Congenital urethral diverticulum typically occurs in the distal ventral urethra, but it is an uncommon location for stone formation[7]. Acquired urethral diverticulum is thought to be frequently due to intrauterine urethral stricture or anterior urethral valves leading to obstructive uropathy[8].

In the present case, epispadias repair was complicated by a dorso-lateral urinoma and consecutive urethro-cutaneous fistula formation. The fistula closed spontaneously thereafter. Upon admission of the patient for elective reflux surgery, a dorso-lateral urethral diverticulum-like structure with 2 calculi was observed. As suggested for congenital urethral diverticulum, the etiology of the reported diverticulum associated with EE may also be due to the abnormality of the urethral plate with focal limitation of spongiosum development[6,7]. Nevertheless the most likely origin of this pathology is a

partial urethral stenosis with subsequent urinoma and fistula development, following surgical reconstruction of the urethral tube. Furthermore, urolithiasis affects patients with bladder exstrophy approximately 160 times more often than the general population[4]. Its location is primarily vesical or in the upper urinary tract, but calculi are found to be urethral in 8% of these patients[4].

Distal urethral calculi are reported to have a mean diameter of 5-7 mm[1,9]. In our patient the size of 28 mm for a single stone is very uncommon. The absence of classic clinical presentation such as pain and/or obstructive voiding symptoms, post-void dribbling[10], and the fact that the diverticulum was not hindering the urinary flow prevented early diagnosis and certainly favored the stone's excessive growth.

In conclusion, endoscopic management and/or lithotripsy can be done in most cases of small urolithiasis[1,9]. In case of a calculus located in a large diverticulum-like fistula, open surgery and extraction of the stones followed by urethroplasty is mandatory. A congenital or acquired distal urethral diverticulum remains exceptional even in patients with EE. Symptomatology and treatment of urethral fistula and calculi depend widely on their shape, size and location.

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RESUME

CALCUL DE LA FISTULE URETRALE APRES CHIRURGIE D'EXTROPHIE-EPISPADIAS

Nous rapportons le cas très peu commun d'un calcul énorme, s'étendant de l'urètre dans une fistule urétro-cutanée dorso-latérale chez un garçon africain de huit ans qui avait subi la fermeture primaire de vessie pour extrophie à l'âge de 16 mois, suivi de chirurgie d'épispadias 6 mois plus tard. Le patient a été perdu de vue au suivi après chirurgie d'exstrophie-epispadias pendant 6 années. À 8 ans, il s'est présenté avec une fistule de l'urètre distal formant un diverticule, la structure contenant un calcul rond avec un diamètre de 28 millimètres. La fistule a été ouverte, le calcul enlevé et une urétroplastie a été exécutée. Un tel calcul urétral, à notre connaissance, n'a pas été précédemment rapporté parmi les cas des chirurgies pour extrophie-épispadias (EE). Ceci montre

la nécessité et l'importance du suivi à long terme, comme une complication si grave peut se développer bien après une chirurgie primaire.
Mots clés : enfant, diverticule, calcul, extrophie, épispadias.

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