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Female Hypospadias and Urethral Stricture Disease in a Circumcised Postmenopausal African Woman: Diagnosis and Management

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ABSTRACT

Globally, both female hypospadias and female urethral stricture are uncommon conditions. Female genital mutilation, on the other hand, is a common practice in the West African sub-region, with up to half of the female population circumcised. We report a case of female hypospadias with stricture in an elderly West African lady who had also been subjected to female genital mutilation during childhood. Urethral dilatation with long-term clean intermittent self-catheterization may suffice in these situations, with scarred vaginas avoiding the need for invasive, cumbersome, and difficult surgical reconstruction of the urethra.

INTRODUCTION

Both female hypospadias and female urethral stricture are uncommon urologic conditions [1,2]. Female hypospadias refers to an abnormality of the urethra with the external meatus located on the anterior vaginal wall within the introitus, and it is often associated with recurrent lower urinary tract infections [1]. Its diagnosis, though simple, is often overlooked and usually made after difficult urethral catheterizations. Female urethral stricture occurs in 4 to 18% of women with bladder outlet obstruction, and, unlike extensively researched male urethral stricture disease, only limited studies on small series are reported in literature [2,3]. The most common etiologies of female urethral stricture are traumatic urethral injury, iatrogenic urethral injury, and inflammatory urethral disease; its optimal management is still under evaluation [4].

Female genital mutilation is an abhorrent traditional practice not uncommon in the West African sub-region, often resulting in scarring of the female external genitalia and life-long morbidity [5]. Here we describe our management of hypospadias with urethral stricture in an elderly West African lady who additionally had been previously subjected to female genital mutilation during childhood.

CASE REPORT

A 68-year-old widowed West African lady presented to our Emergency Department with chronic urinary retention. She had a preceding 10-year history of worsening obstructive lower urinary tract symptoms but no hematuria or necroturia. She also had multiple episodes of cystitis over several decades; mostly post-coital. Her first pregnancy 40 years prior ended in obstructed labor and caesarean section delivery of a live baby; however, she subsequently had six uneventful vaginal deliveries. The patient had been subjected to female genital mutilation during early childhood, which is a tradition practiced by her West African Yoruba tribe. Physical examination revealed painless suprapubic swelling over 20 weeks and a hypertrophic midline infraumbilical scar. Examination of the external genitalia revealed an amputated rudimentary clitoris and defects in and scarring of the labia minora with vagina atrophy; the dimple of the urethral meatus was seen not at its normal anatomic position but on the anterior vaginal wall juxtaposed to the edge of the vaginal introitus (Figure 1 and Figure 2). The opening could not admit a 12 Fr urethral catheter. Serum assays showed deranged electrolytes with hyperkalemia, with elevated urea and creatinine. An abdominopelvic ultrasound revealed bilateral hydronephrosis and a grossly distended

KEYWORDS: Female hypospadias; female urethral stricture; female genital mutilation

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Figure 1. Scarred and atrophic vagina.

Figure 2. Female external genitalia: An amputated rudimentary clitoris, scarred labia minora, and an atrophic vagina with the external urethral meatus located on the anterior vaginal wall juxtaposed to the edge of the vaginal introitus.

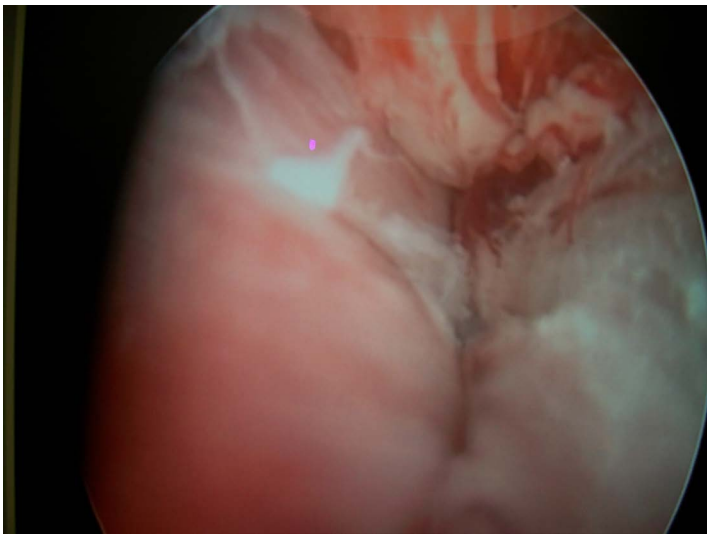
bladder with 0.5 cm thickened walls. She had a suprapubic cystostomy done, and an indwelling suprapubic catheter was placed for continuous drainage. Post obstructive diuresis occurred and resolved after 4 days, with normalization of the deranged electrolytes. A cystogram showed evidence

Figure 3. Cystogram: Evidence of long-standing bladder outlet obstruction with extensive trabeculation of the bladder (irregular walls). An attempt at obtaining a micturating film failed, as the bladder neck did not open and the urethra was not outlined despite a distressing urge to micturate and straining.

of extensive trabeculations in the bladder and an attempt at obtaining a micturating film failed, as the bladder neck did not open and the urethra was not outlined despite distressing urge (Figure 3). A 6 Fr feeding tube was successfully negotiated through the dimple of the external urethra under fluoroscopic guidance, and urethral dilatations to 20 Fr were done easily over a guide wire. This made urethrocystoscopy possible, which revealed fibrosis and fixed narrowing of the distal third of the urethra (Figure 4), a normal proximal urethra, extensive trabeculation, and sacculations in the bladder. The urethra was subsequently dilated to 30 Fr, an indwelling 16 Fr Foley urethral catheter was placed, and the suprapubic catheter removed. The urethral catheter was removed after a week and she voided satisfactorily with a Qmax of 20 ml/sec. She was taught clean

intermittent self-catheterizations with an18 Fr catheter and instructed to do it thrice weekly for the first month; this was reduced to once weekly during the second month, as she was voiding satisfactorily with no evidence of residual stricture. At the sixth month of follow-up, the patient still voids satisfactorily with no new episodes of symptomatic urinary tract infection; she empties her bladder with only insignificant post-void residual urine volume (40 ml) seen on ultrasonography and the previous hydronephrosis noted has resolved. Uroflowmetry revealed normal flow, with a Qmax of 25 ml/sec and a voiding volume of 520 ml.

Figure 4. Urethroscopy: Urethroscopic view showing scarring and fibrosis of the distal urethra.



DISCUSSION

Female hypospadias, though often asymptomatic, may be associated with bothersome symptoms of vaginal voiding, post-coital cystitis, or urethral syndrome [1]. Treatment involves surgical reconstructions of the hypospadias; two common techniques in use include vaginal flap urethroplasty and urethrolysis with meatal transposition (with or without submucosal tunneling) [1,6]. To our knowledge, stricture of the female hypospadiac urethra has not been previously reported. We suspect the stricture in the index case may be due to periurethral fibrosis resulting from recurrent lower urinary tract infections commonly associated with the anomaly [1,6]. At present, there are no defined diagnostic criteria for female urethral stricture; however, lower urinary tract symptoms and difficulty in placing a 12 Fr urethral catheter or larger is suggestive [7].

Smith et al. demonstrated the efficacy of urethral dilatation and long-term clean intermittent self-catheterization (CISC) for female urethral stricture, and its success was shown to hold off the need for major reconstructive surgery; this is particularly true for patients compliant with the CISC regime, though the best size of catheter or the best regime for CISC remains unknown [8]. This option is attractive for those patients desiring less invasive treatment and motivated to perform CISC. It may also prove useful for the patient with a scarred vagina in whom urethral reconstruction may be difficult and prone to failure. In our region (sub-Saharan Africa), there are additional issues of cost of and compliance with CISC to consider. The index patient is well motivated, and the empirically prescribed CISC frequency of once weekly lessens the cost burden. A recent report with larger series and longer follow-up than that reported by Smith et al. described a poorer outcome following urethral dilatation for female urethral stricture [3]; however, the patients in the series did not receive long-term CISC, which may have been responsible for the observed lack of sustained response.

Various urethroplasty techniques for female urethral strictures have been described [6,9-12]. A vaginal inlay flap is simple and involves the advancement of an inverted U-shaped flap of anterior vaginal mucosa into a posterior stricturotomy of the diseased urethra; the technique has shown satisfactory long-term results [6]. A Y-shaped vaginal vestibular flap, which is rotated and interposed into an anterior stricturotomy, is a slightly more complex procedure but has also shown acceptable results [9]. Dorsal and ventral onlay urethroplasty using buccal, lingual, and vaginal mucosal grafts has also been reported with durable results [10-12].

Surgical repair of female hypospadias with stricture may be more complex, as the stricture would further shorten the already short hypospadiac urethra. Reconstruction may involve a combination of urethrolysis, urethral transposition, and urethroplasty, or some other cumbersome amalgam of procedures that may be especially daunting in a scarred vagina from FGM, as is the case in the index patient. Urethral dilatation with long-term CISC appears to be a reasonable alternative in this situation.

Female genital mutilation is not uncommon in West Africa; though overall prevalence appears to be decreasing, up to half of all females are circumcised, and a quarter of the young girls are still being circumcised today [5]. Just as some male urethral reconstruction surgery is potentially made more difficult following circumcision, female genital mutilation may result in more difficult female urethral and genital reconstructive surgery, and conservative treatment may be a reasonable alternative for these patients.

CONCLUSION

Patients with female hypospadias have recurrent lower urinary tract infections, which may lead to periurethral fibrosis and urethral stricture disease. Surgical reconstruction may be cumbersome and difficult, especially in a scarred vagina. Urethral dilatation with long-term CISC may be reasonable and effective in these situations, especially in the compliant patient, thus avoiding the need for invasive and difficult surgical reconstruction of the urethra.

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