Case report

PENOPUBIC EPISPADIAS IN A 40YEAR OLD MAN: A CASE REPORT

ABSTRACT

Epispadias is part of the exstrophy-epispadias complex; it has a defective dorsal wall of the urethra with potential incompetence of urinary continence mechanism1. Isolated male epispadias is due to failure of the urethral plate to tabularize on the dorsum of the penis. It is rare with an incidence rate of 1 in 117,000 live males2. The severity depends on the position of the urethral opening and ranges from peno-pubic to penile and glandular2. Epispadias is usually repaired within the first year of life though some patients have presented in their second decade of life 1-3. Epispadias is classically associated with bladder extrophy in over 90% of the cases while isolated epispadias with continence is very rare constituting less than 10% of cases4. Separation of pelvic bones is seen in 70% of peno-pubic epispadias affecting the bladder neck and external sphincter leading to incontinence and stress urinary dribbling3. The goals of repair include; achieving a cosmetically acceptable and functional penis, which is straight and adequate in length, enabling penetrative sexual intercourse and urinary continence 1,2. There has been a report of isolated epispadias in adults but none was as old as 40 years. This patient was married for 20 years but had been unable to impregnate his spouse. He had modified Cantwell-Ransley procedure after a detailed clinical and psychological evaluation to achieve the goals of repair. Adults presenting with isolated continent peno-pubic epispadias are rare. Presenting at this age and after having been married for two decades could have a derogatory effect on body image, self-confidence, psycho-sexual and reproductive life. Surgical correction of this anomaly as we did has the potential to lead to acceptable outcomes as demonstrated in this index case.

KEYWORDS

Urethra, continent, penile, exstrophy, isolated, anomaly

INTRODUCTION

Epispadias is part of the exstrophy-epispadias complex; it has a defective dorsal wall of the urethra with potential incompetence of urinary continence mechanism[1]. Isolated male epispadias is due to failure of the urethral plate to tabularize on the dorsum of the penis. It is rare with an incidence rate of 1 in 117,000 live males[2]. Severity depends on the position of the urethral opening, from penopubic to penile and glandular[2]. Epispadias is usually repaired within the first year of life though some patients have presented in their second decade of life 1-3. The goals of repair include; achieving a cosmetically acceptable and functional penis, which is straight and adequate in length, enabling penetrative sexual intercourse and urinary continence [1,2].

There have been reports of isolated epispadias in adults but none was as old as 40 years. This patient was married for 20 years but had been unable to impregnate his spouse. He had modified the Cantwell-Ransley procedure after a detailed clinical and psychological evaluation to achieve the goals of repair.

CASE SUMMARY

Mr Y.D. is a 40year old married man who presented to our clinic on account of an abnormally formed penis since birth. Growing up he noticed his penis looked different from that of his friends and he urinated from a dorsal and proximal point making his urine splay. He gets penile erections which tend to curve towards the anterior abdomen wall.

No urinary incontinence or gait abnormality was noted. After being married for 20 years his wife convinced him to seek proper evaluation and treatment. The marriage was said to have been consummated however no deposition of semen into the vagina during ejaculation. There was no proper medical care in childhood as he grew up an orphan with his older sibling being the caregiver.

Examination revealed an abnormally formed penis, twisted about 45 degrees to the left with a ventral hood, dorsal chordee, penopubic meatus, and a supple and splayed penile urethra with the urethral plate measuring 4cm in width. Figures 1 and 2. Scrotal examination revealed normal testes bilaterally. Abdominopelvic sonography and pelvic X-rays revealed no abnormality. Figure 3. His complete blood count was normal with a pack cell volume of 47% and normal kidney function.

After adequate counseling, consent was obtained and he had modified Cantwell Ransley repair under combined spinal and epidural anesthesia. Meatal advancement and glanuloplasty were done using the Heineke-Mikulicz technique. The penis was de-gloved down to its root. Parallel incisions were made on the sides of the urethral plate to mobilize it from the corpora cavernosa. Figure 4. The urethral plate was tabularized over a stent (size 16Fr silicon catheter) using continuous submucosal using monocryl 4/0 suture. Caverno-cavernostomy was done on both sides of the corpora allowing the tubularized urethral to be repositioned ventrally before suturing the caverosa together above the urethral. Figure 5. A W-flap was raised in the pubic region and the suspensory ligament was released to give length to the penile shaft. The penile skin was then apposed with vicryl 3/0 sutures and the skin flap closed. We left a supra-pubic catheter in place for temporary urinary diversion to rest the urethral repair.

Postoperatively he received parenteral anti-biotics and analgesics. He was commenced on oral feeds and medication including tolterodine, antibiotics and haematinics. He was discharged home on the 9th postoperative day with the urethral stent and suprapubic catheter in situ. Eight days later he developed a superficial surgical site infection which we treated using out-patient daily dressing with povidone iodine. The suprapubic and urethral catheters were removed by the 4th post-operative week following a normal urethrogram. Figure 6. He voided satisfactorily and is happy with the outcome. Figure 7.

DISCUSSION

Isolated epispadias with continence is very rare constituting less than 10% of cases of epispadias.[4] It is classically associated with bladder extrophy in over 90% of cases.[5] Repair of epispadias is usually done in the first year of life. Presentation in adulthood is not common. The site of the urethral opening varies depending on the severity, from peno-pubic to penile and glandular[2]. This is characterized by aplasia of the dorsal part of the urethra.[6]

There may be varying degrees of incontinence. Treatment is surgical and aimed at giving the patient normal urinary control, and a straight, cosmetically and functionally acceptable penis. Surgical techniques used in the repair of epispadias include the Modified Cantell Ransley technique and Mitchell-Bagli technique[2]. Mitchel and Bagli introduced the complete penile disassembly technique. There is complete splitting of the corporal bodies and hemi-glans into separate halves with complete dissection of the urethral plate off the corpora. This makes for proper ventralization of the tubularized urethra. Cantwell proposed a technique based on complete mobilization of the urethral plate that was then tubularized and transplanted ventrally between the corpora. Young

modified it by avoiding complete urethral plate mobilization for better preservation of its vascularity thus decreasing the risk of urethral fistula formation.[4]

Our patient, who was married for 20 years, presented to us at the age of 40 following persuasion from his wife. There was no record of medical consultation during his young age as he began as an orphan very early in life and the caregiver did not make sufficient effort toward having the abnormality treated. [7]

Isolated epispadias with continence is rare comprising <10% of all epispadias cases4. Over 90% of the cases present with bladder extrophy[5]. Separation of pelvic bones is seen in 70% of peno-pubic epispadias affecting the bladder neck and external sphincter leading to incontinence and stress urinary dribbling[3]. The index patient did not present with any of these and he had a one-stage repair.

CONCLUSION

Adults presenting with isolated continent peno-pubic epispadias are rare. Presenting at this age and after having been married for two decades could have a derogatory effect on body image, self-confidence, psycho-sexual and reproductive life. Surgical correction of this anomaly as we did has the potential to lead to acceptable outcomes as demonstrated in this index case.

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Figure 1. Clinical picture of peno-pubic epispadias before surgery

Figure 2. Plain Pelvic X- ray showing normal anatomy

Figure 3. Clinical picture showing degree of chordee in epispadias

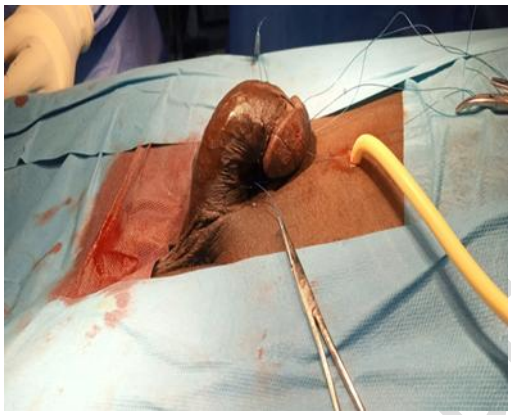

Figure 4. Separation of the urethral plate from the cavernosa

Figure 5. Immediate post-operative clinical picture

Figure 6. Post-operative urethrogram showing the normal reconstructed urethra

Figure 7. Clinical picture 2 months after repair