Case Report

Surgical Management of Complete Procidentia in a Female Patient with Bladder Exstrophy-epispadias Complex: Case Report and Literature Review

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Abstract

We herein describe the operative approach of a postmenopausal woman with a history of surgically corrected congenital bladder exstrophy-epispadias who presented with long-standing complete procidentia. The patient was initially treated by laparoscopic sacral colpopexy in conjunction with a modified Elevate mesh kit anterior vaginal repair with and posterior vaginal wall repair in the form of native tissue suture plication repair. Her prolapse recurred 8 months' later due to a detachment of the mesh at the level of the promontorium. During the second-look laparoscopy, a resuspension of this mesh was deemed unsatisfactory; therefore, with patients' consent, a successful colpocleisis was performed. This case report emphasizes the complexity of pelvic organ prolapse (POP) in the context of a bladder exstrophy-epispadias complex. These women are more likely to fail the more conventional current surgical treatments for POP, coercing to revert to colpocleisis.

Keywords: Bladder exstrophy, colpocleisis, pelvic floor, procidentia, prolapse recurrence

Introduction

Bladder exstrophy-epispadias complex (BEEC) is a rare congenital malformation which originates from a failure of midline closure during fetal development, leading to the absence of the anterior bladder wall and consequent protrusion of the posterior bladder through the anterior abdominal wall. Failure of midline pubic symphysis fusion often coexists with pubic diastasis, bifid clitoris, anterior displacement of internal genitals and labia majora, and an absent or attenuated urethrovesicular sphincter and urethra.[1,2] The associated congenitally weakened cardinal and uterosacral ligaments, anatomically modified levator ani muscles along with the pubic diastasis gives rise to an increased risk of pelvic organ prolapse (POP), which is reported to emerge in approximately 18%–30% of women with bladder exstrophy.[2,3]

These anatomic changes along with potential adhesions from previous reconstructive surgery, make the operative management of POP in bladder exstrophy patients extremely challenging.[4]

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Case Report

A 57-year-old parous woman with severe complete procidentia was referred to our urogynecological unit for management of her uterovaginal prolapse. Aside from the prolapse, she suffered from recurrent cystitis, urgency but no stress urinary incontinence. Bowel motions were normal. In her medical history, we note hypertension and type 2 diabetes mellitus. During early childhood, she was diagnosed with bladder exstrophy-subsymphyseal epispadias complex and a reconstruction of the bladder, urethra, and the external genitalia was performed along with a rectus sheath fascial sling operation to correct stress urinary incontinence at the age of 6 years. The latter was complicated by the formation of a fistula draining urine through the suprapubic wound, which was surgically corrected. Although she noted improvement

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of the stress incontinence, it had not completely resolved until 6 months' later, when a subsequent urethroplasty was carried out.

The patient had two uneventful pregnancies both delivered by cesarean section at term. During the postpartum, her prolapse deteriorated and became symptomatic. The congenital malformation of her pelvic outlet and absent pelvic floor support made the use of a pessary impossible and no further action was taken upon.

Physical examination revealed a complete procidentia and a wide genital hiatus, due to absent pubic rami and absent symphysis. POP Quantification (POPQ) was Aa + 3, Ba + 12, C + 12, Gh 10, Pb 3, TVL 16, Ap + 3, Bp + 16, D + 16.

On reduction of the prolapse occult stress incontinence was revealed. A postvoid residual of 100 ml was detected. The urethral meatus was pinpoint and malpositioned toward the right, and the vaginal epithelium was hyperkeratinized [Figure 1].

Umbilical hernia and scarring from previous midline incision were noted. The patient declined having had vaginal intercourse for over two decades.

After multidisciplinary consultation, a surgical correction of the prolapse with the postoperative assessment of the urinary incontinence and potential second-stage management was advised.

A laparoscopic subtotal hysterectomy and sacrocolpopexy in combination with anterior vaginal mesh repair and posterior colpoperineoraphy were planned.

First, an anterior vaginal wall repair with Elevate Anterior mesh (AMS, Minnetonka, USA) was performed. The anterior vaginal wall was incised vertically and vaginal skin reflected off the bladder. The Elevate mesh was inserted and lower lateral aspect of the mesh secured to obturator internus muscle bilaterally in the usual manner. The apical aspect of the mesh was not secured to the sacrospinous ligament as per usual insertion, but an extension of a Gynecare Gynemesh PS Prolene mesh (Ethicon, J and J) was added, rolled up, and positioned high in the vesicovaginal space to facilitate later laparoscopic retrieval to form the anterior mesh arm of sacrocolpopexy. The intention was to achieve a continuous mesh support to the entire anterior vaginal wall from Level 1 to Level 3. After closure of the vaginal incision, the laparoscopic component of the repair was commenced. Palmer's point entry technique was judged to be safest in the context of previous abdominal surgeries and anticipated intra-abdominal adhesions. On entry dense, omental adhesions to the suprapubic midline became apparent. Furthermore, the bladder was adherent to the abdominal wall and to the uterus. After extensive adhesiolysis, the anatomy could be examined. Except for a bicornuate fibroid uterus and a right hydroureter, possibly from long-standing ureterovesical reflux, no other intra-abdominal anatomical changes were observed. Subtotal hysterectomy and bilateral salpingo-oophorectomy were executed with an advanced bipolar instrument, followed by in-bag morcellation of the uterus. Next, the bladder was reflected downward and the anterior Elevate mesh with the GyneMesh extension was retrieved. The mesh was unrolled and used as anterior vaginal arm of sacrocolpopexy. Consequently, the pouch of Douglas was opened and the peritoneum reflected off the posterior vaginal wall to approximately 3 cm from the perineal body.

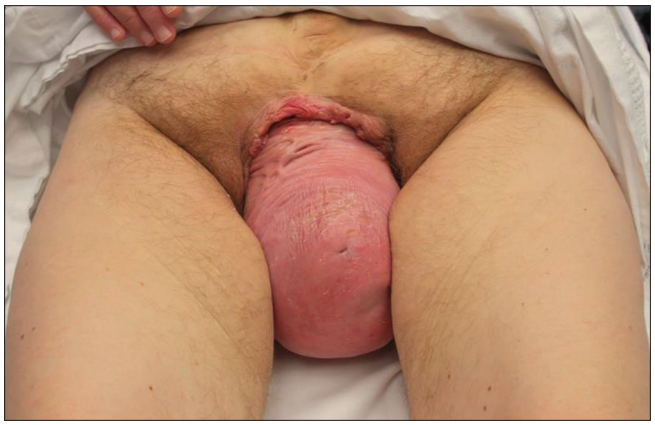

Figure 1: Complete procidentia in a patient with bladder exstrophy-epispadias complex

Then, the Gynemesh was cut to size and sutured to posterior vaginal wall and cervical stump (2.0 PDS, p-dioxanone, Ethicon, J and J). The anterior and posterior meshes were joined with a nonabsorbable suture (2.0 Prolene, Ethicon, J and J) superior to the cervical stump. Subsequently, the presacral area was opened and the mesh secured by tacking (ProTack™ 5 mm Fixation device, Medtronic) to the sacral promontorium after which the peritoneum was closed over the mesh.

Finally, classical colpoperineoraphy was performed to reduce the rectocele and very widened genital hiatus.

The overall postoperative course was uneventful. The patient displayed a satisfying anatomic correction (POPQ Aa-1, Ba-1, C-8, Gh 7, Pb 3, TVL 9, Ap-2, Bp-2, and D-9) and no urinary, bowel, or prolapse symptoms. Unfortunately, 8 months' later, she developed recurrence of almost the same degree of prolapse noted preoperatively. A laparoscopic revision of her sacrocolpopexy was planned and consent for colpocleisis was obtained. Laparoscopic inspection revealed detachment of the mesh from the sacral promontory. Resuspension of the proximal end of the mesh, however, did not deliver a satisfying result. Therefore, a colpocleisis was deemed the treatment of choice. Postoperatively, the patient was subjectively satisfied with normal urinary continence. On examination, an obliterated vagina without prolapse was noted [Figure 2].

Discussion

The relationship between BEEC and POP has been well documented. The earliest case report was described by Ismael in 1939.[5]

Precipitating factors contributing to POP and procidentia in these patients are congenitally weakened cardinal-uterosacral

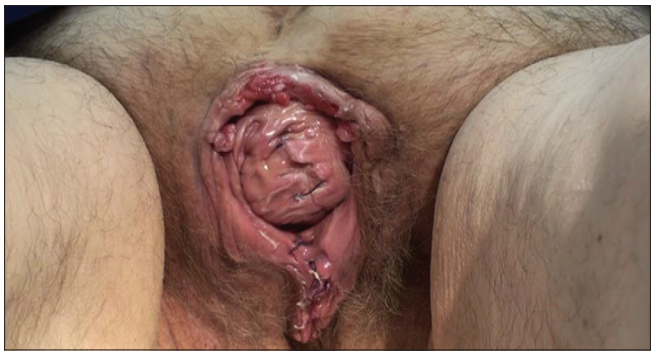

Figure 2: Status postcolpocleisis

ligamental (level 1) support and the reduction in level 2 and 3 support due to the absence of anterior bony pelvis and distorted muscular support of the anatomically altered levator ani muscles.[1,2] Pregnancy may aggravate symptoms in this prolapse-prone population. Burbige *et al*. reported on uterine prolapse complicating pregnancy in 7 out of 11 patients with bladder exstrophy.[6] Similarly, Giron *et al*. described 14 pregnant bladder exstrophy patients,[7] half of which suffered from genital prolapse. Our patient, although having preexisting prolapse since adolescence, developed symptomatic prolapse after her second delivery at age 36, which was initially managed conservatively.

Both the anatomical changes as well as potential adhesions from previous reconstructive surgery make the operative management of POP in these patients challenging. Hence, several surgical techniques to manage POP have been described.[4]

A vaginal approach, such as sacrospinal hysteropexy or colpopexy which addresses level 1 support has been reported to have successful outcomes[1] and can be a valuable treatment option. Abdominal procedures such as sacral colpopexy and hysteropexy are considered to have more durable results in bladder exstrophy patients and may be preferred, especially in setting of recurrent prolapse.[4] However, one has to be aware that cervical elongation may occur after sacral hysteropexy, mimicking uterine prolapse recurrence, as described by Kwong *et al*.[8]

We opted to manage the procidentia with laparoscopic sacral colpopexy, since this is considered the gold standard in addressing apical prolapse[9] and because of the great level of expertise in our center regarding this technique. Unfortunately, the patient relapsed 8-month postoperatively due to detachment of the mesh at the level of the promontory. Vault recurrences after laparoscopic sacral colpopexy may be attributed to detachment of the mesh at the level of the vaginal vault, cervical remnant, or at the promontory. Nygaard *et al*. showed a 5% surgical retreatment for recurrent prolapse in his long-term follow-up study after abdominal sacrocolpopexy.[10] We used 2 Protacks (nonabsorbable) to attach the mesh to the promontory, but nonabsorbable sutures have been equally described for this purpose.

We managed the prolapse recurrence after laparoscopic sacrocolpopexy with a colpocleisis, an obliterative procedure, which is a reasonable option in managing prolapse recurrence after laparoscopic sacral colpopexy in nonsexually active patients.

Conclusion

This case describes the complexity of the surgical management of procidentia in a patient with BEEC. Laparoscopic sacral colpopexy is considered the gold standard for apical prolapse. Failure of this treatment in a BEEC-patient is extremely challenging and consequent management must be tailored to the individual patient. One has to take into account the surgical risks, coital activity, and vaginal anatomy. In this case, a colpocleisis was considered a reasonable option to manage the prolapse recurrence.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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