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An unusual cause of urinary incontinence: Urethral coitus in a case of Mayer-Rokitansky-Kuster-Hauser syndrome

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Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a rare anomaly in women and is characterized by congenital aplasia of the uterus and vagina, with normal development of secondary sexual characteristics and a normal karyotype. We report a case of a 38-year-old women with MRKH syndrome that had experienced urethral sex for many years. She presented with urinary incontinence and dyspareunia. The patient's secondary sexual characteristics were normal, and examination revealed a widely open incompetent megalourethra and an absent vagina. Laboratory studies confirmed a 46, XX karyotype. Imaging included ultrasonography and magnetic resonance imaging, which indicated bilateral normal ovaries and a rudimental bicornuate uterus. After confirming the diagnosis of MRKH, the patient underwent urethroplasty by urethral plication, antiincontinence surgery by autologous fascial sling of the bladder neck, and the creation of a neo-vagina using a urethral flap. After 3 months, voiding cystourethrography and uroflowmetry confirmed normal voiding. There were no postoperative symptoms of urinary incontinence, and the patient was completely satisfied.

Keywords: Plastic surgery; Urethra; Urinary incontinence

INTRODUCTION

Urethral coitus is a rare condition and has been mostly reported in women with Müllerian anomalies. Urinary incontinence, dyspareunia, and infection are the most common presenting symptoms in patients with urethral coitus [1].

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is a disorder of the female genital tract and is characterized by the absence of the upper portion of the vagina, an absent or hypoplastic uterus, and normal or hypoplastic fallopian tubes [2]. These patients may also have associated urological or skeletal abnormalities. Women with MRKH syndrome have normal genotypes and phenotypes, and diagnosis is often made during adolescence in the context of primary amenorrhea with normal puberty [3].

Here, we report a case of previously undiagnosed MRKH syndrome with urinary incontinence as a result of urethral coitus and megalourethra.

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CASE PRESENTATION

A 38-year-old woman was referred to our clinic complaining of severe urinary incontinence that easily occurred during light, daily physical activities (walking and changing the position). Her medical history revealed that she had primary amenorrhea, difficulty with intercourse, and infertility.

Physical examination showed normal secondary female sexual characteristics, and a genital examination in the lithotomy position revealed a normal vulva, labia majora, labia minora, and clitoris. However, she had a wide, open megalourethra, which was incompetent with poor closure, and a finger easily entered the bladder (Fig. 1). Vaginal examination by speculum was impossible because of vaginal agenesis. Laboratory studies included chromosomal analysis, which revealed a 46, XX karyotype. Ultrasonography and magnetic resonance imaging showed normal bilateral ovaries, a rudimental bicornuate uterus with intrauterine myoma, and vaginal agenesis. Urodynamic study results revealed a stable bladder and a positive stress test, with a Valsalva leak point pressure of 20 cm H_2O and maximum urethral closure pressure less than 10 cm H_2O (intrinsic sphincter deficiency).

Cystourethroscopy confirmed a dilated urethra and an open bladder neck (Fig. 2). After confirming MRKH syndrome and severe stress urinary incontinence, the patient was scheduled for surgical repair.

SURGERY

The patient was placed in the dorsal lithotomy position

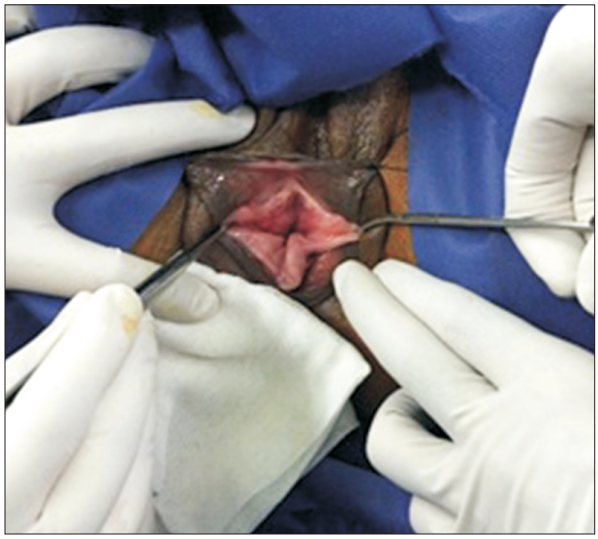

Fig. 1. Megalourethra in a case of Mayer-Rokitansky-Kuster-Hauser syndrome.

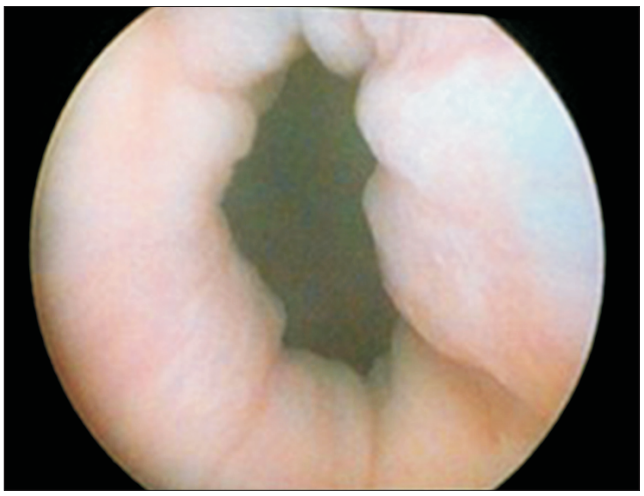

Fig. 2. Open bladder neck in cystourethroscopy.

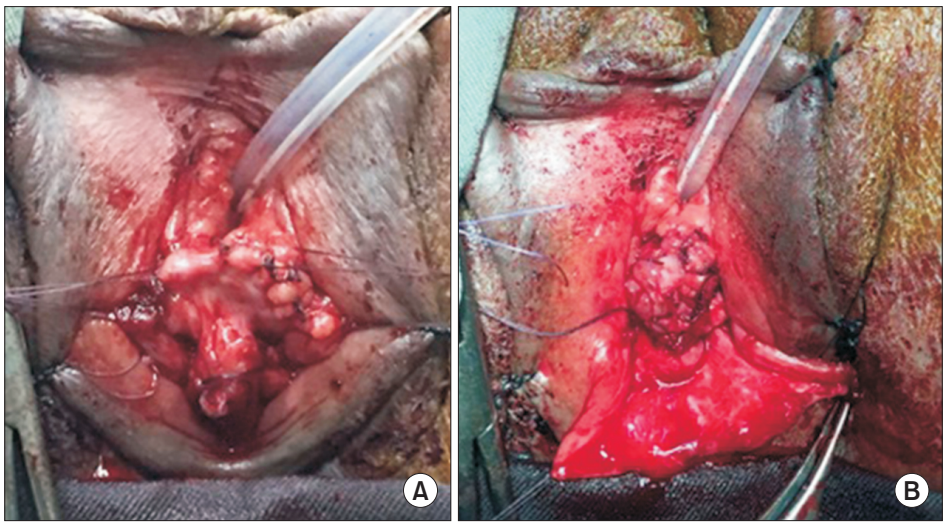

Fig. 3. (A) Urethral reconstruction: The superior mucosal layer of ventral lip of the urethral wall was plicated over the 16-Fr Foley catheter. (B) Inferior layer was prepared for vaginoplasty.

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under general anesthesia. A 16-Fr Foley catheter was transurethrally inserted, and an inverted U incision was made through the ventral lip of the redundant urethral wall. After dissecting the ventral aspect of the urethral wall from 2 to 10 o'clock and exposing the bladder neck, the urethral mucosa and submucosal tissues were tapered to a Foley catheter. Urethral plication was performed until the urethra fit over the 16-Fr Foley catheter (Fig. 3). The urethral wall was separately sutured on both lateral sides by 4-0 vicryl stitches over the catheter, from the meatus to the bladder neck. Then, the redundant wings were ventrally transferred and sutured over each other without incising the urethral lumen.

As there was no vaginal lumen, the dissection was carried out to free the urethra and to develop a narrow paraurethral space bilaterally. The suprapubic incision was made, and a rectus fascia graft (6 cm long) was harvested.

Stamey needles were passed (approximately 4 cm apart) through the retropubic space from the abdominal wound. Then, the endopelvic fascia was perforated, and the tips of the needles were passed into the paraurethral space. The arms of the fascia were sutured by 0 vicryl sutures. The sling sutures were connected to the tips of the needles, and the needles were brought out through the suprapubic incision. The sling was positioned at the level of the bladder neck and secured by suturing the sling to the periurethral fascia (Fig. 4). The sling sutures were tied in the suprapubic space without tension.

The remaining inferior mucosal layer of the ventral lip of the urethral wall was used to reconstruct the neo-vagina. After creating a space between the urethra and the rectum, the urethral flap was brought through the space and sutured (Fig. 5).

The patient was discharged from the hospital on

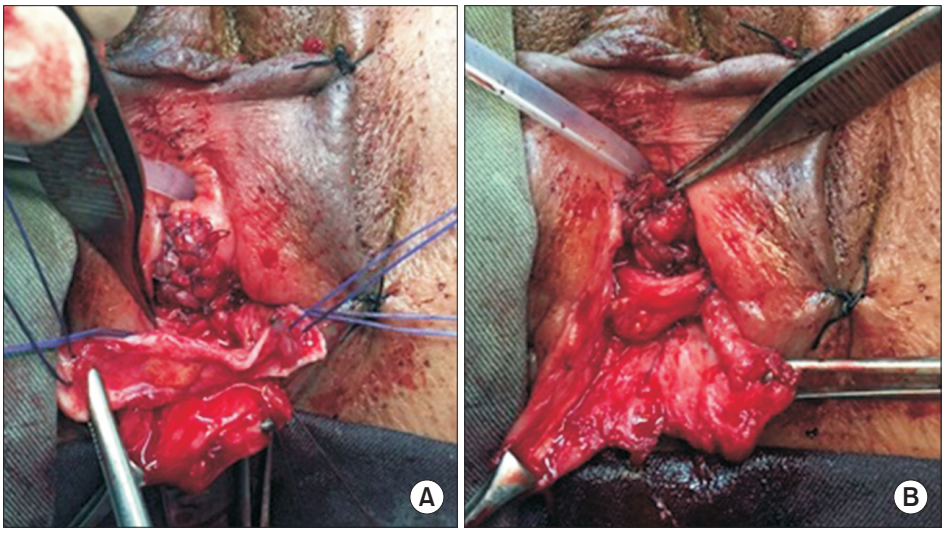

Fig. 4. (A) The rectus fascia graft was prepared for pubovaginal sling. (B) The sling was positioned at the level of bladder neck.

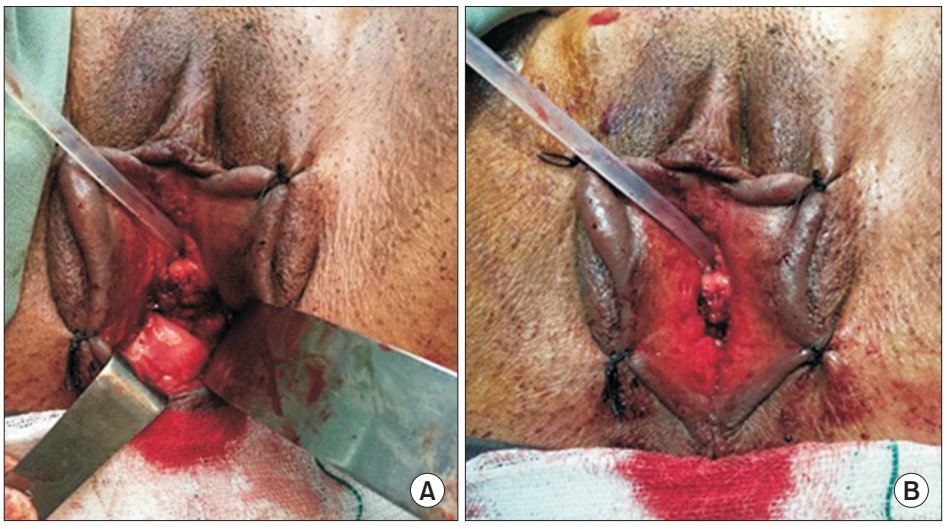

Fig. 5. (A) Creation of neo-vagina in the rectovaginal space that was lined by urethral wall flap. (B) Final result of urethroplasty and vaginoplasty.

postoperative day two, and the Foley catheter was removed three weeks after surgery. At the first trial to void, the patient voided freely with a postvoid residual urine of 30 mL.

Three months later, voiding cystourethrography and uroflowmetry were performed, and normal voiding was confirmed. At the 6-month follow-up, the patient was continent after the removal of the urethral catheter.

DISCUSSION

Urethral intercourse is a rare condition, mostly reported in Müllerian anomalies. These anomalies include Mayer-Rokitansky syndrome, a microperforated hymen, and a transverse vaginal septum, although there are some reports of urethral coitus despite normal genitalia [4].

Reported complications after urethral intercourse include urethral dilatation, trauma, urinary incontinence, and recurrent urinary tract infections; surprisingly, in some cases, there are no associated complications [5].

Aksakal et al. [1] described urinary incontinence due to urethral coitus in a multiparous woman who became continent after plication of the urethra. Zargham et al. [4] reported 2 cases of intra-urethral intercourse and stress urinary incontinence. One of these cases (a 21-year-old patient) had normal genitalia, and the other (a 19-year-old patient) had a vertical vaginal septum. Outcomes were successful in both patients after urethroplasty with paravaginal flap and classic sling.

In the case of stress urinary incontinence (SUI) and intrinsic sphincter deficiency, pubovaginal slings are more effective than retropubic colposuspensions, with outcomes comparable to those reported with midurethral slings. The artificial urinary sphincter provides long-term cure rates but is associated with considerable morbidity, whereas the clinical results of bulking agents are not sustained beyond 1 year [6].

Here, after urethroplasty with plication of the mucosal layer of the ventral urethra, we performed a pubovaginal sling procedure with rectus fascia for the treatment of SUI, and the outcome was favorable.

MRKH syndrome is the second most common cause of primary amenorrhea. Individuals with this syndrome will typically first visit a gynecologist at the age 14 to 15 when the absence of menstrual cycles becomes evident. Often, at this age, there is no examination because patients are too young, and they are given hormonal medications with the hope of triggering menstruation. Sometimes an inaccurate examination may lead to a mistaken diagnosis of imperforated hymen. Occasionally, older patients with the classic MRKH syndrome consult a gynecologist because of difficult or painful intercourse. Therefore, it seems that the initiation of sexual activity is a significant challenge for patients with this syndrome and may lead to abnormal coitus through the urethra, which has been presented in most case reports.

In a case report by Rouzi [7] in 2013, urethral intercourse was documented for the first time in a case of misdiagnosed MRKH syndrome. A 32-year-old woman presented with severe pain, dysuria, and mild hematuria after intercourse. Her examinations revealed an absent vagina and uterus and noticeably dilated urethra. The patient underwent a modified Vecchietti operation.

Bacopoulou et al. [5] in 2014 reported 2 cases of Müllerian agenesis with urethral coitus. A 19-year-old Caucasian woman and a 16-year-old Filipino girl presented with primary amenorrhea and a history of 'normal' vaginal coitus. Neither patient suffered significant urethral damage to require urethra reconstruction. However, the first adolescent suffered recurrent pyelonephritis and renal scars, and the second adolescent had dysuria. The first adolescent underwent a hernia repair and vaginoplasty, and the second adolescent underwent vaginal dilatations.

Management of vaginal agenesis in MRKH syndrome remains controversial. The choice of procedure depends on individual anatomy, fertility potential, and psychological and social factors [8].

There are 2 main types of procedures used. The first procedure consists of creating a new cavity and may be done surgically or nonsurgically. The second procedure is vaginal reconstruction with a segment of the bowel [9,10].

Currently available methods for vaginal reconstruction are the Vecchietti procedure, which consists of increasing the vaginal size by gradually applying traction to the vaginal wall, and the creation of a neo-vagina in the rectovaginal space that is lined by different flaps, such as skin, peritoneum, and intestine [11].

In our case, the patient had megalourethra with redundant wall, and a neo-vagina was constructed by using part of the inferior urethral wall as a flap for lining the vaginal cavity.

CONCLUSIONS

We reported a case of MRKH with severe SUI that was treated with plication of megalourethra and a pubovaginal sling procedure, as well as the creation of a neo-vagina, with acceptable result. In conclusion, careful assessment and

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physical examination of patients with primary amenorrhea is recommended in adolescents.

CONFLICTS OF INTEREST

The authors have nothing to disclose.

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