Urethral sex in a woman with previously undiagnosed Mayer-Rokitansky-Küster-Hauser syndrome

A.A. Rouzi

Department of Obstetrics and Gynecology, King Abdulaziz University, Jeddah (Saudi Arabia)

Summary

Purpose: To report a case of urethral sex in a woman with previously undiagnosed Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome. *Materials and Methods:* A 32-year-old woman presented with severe pain, dysuria, and mild hematuria after each intercourse. Secondary sexual characteristics were normal. Vaginal and rectal examinations revealed an absent vagina and uterus. Further investigations showed a normal hormonal profile, a 46 XX karyotype, and a normal intravenous pyelography. Pelvic ultrasonography and magnetic resonance imaging (MRI) confirmed the absence of a uterus and the presence of bilateral ovaries. *Results:* A diagnosis of MRKH syndrome was made and the patient underwent a modified Vecchietti operation for the creation of a new vagina. The urethral meatus was noticeably dilated. Postoperatively, the vaginal length was six to seven cm. Long-term follow-up revealed that she was able to have normal and satisfactory vaginal intercourse without any problems. *Conclusion:* Urethral intercourse is documented here for the first time in a case of misdiagnosed MRKH syndrome.

Key words: Urethra; Sex; Mayer-Rokitansky-Küster-Hauser syndrome.

Introduction

A review of the published literature shows that urethral sex has never before been reported. A unique case of urethral intercourse in a woman diagnosed with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is presented.

Materials and Methods

This was an apparently healthy 32-year-old woman who sought medical advice for severe pain, dysuria, and mild hematuria after each intercourse. She had no history of urinary incontinence. At time of presentation, she had been married for two years to a 65-year-old man. Intercourse was infrequent and occurring twice every month. She was seen by other physicians more than ten years prior for primary amenorrhea and was reassured. She had never had any menses. On examination, her weight was 111 kg and height was 168 cm. Secondary sexual characteristics were normal. Vaginal and rectal examinations revealed an absent vagina (Figure 1) and uterus. Investigations showed a normal hormonal profile, a 46 XX karyotype, and normal intravenous pyelography. Pelvic ultrasonography and magnetic resonance imaging (MRI) confirmed the absence of the uterus and the presence of bilateral ovaries. A diagnosis of MRKH syndrome was made and she underwent a modified laparoscopic Vecchietti operation for the creation of a neovagina as previously reported [1]. The urethral meatus was noticeably dilated (Figure 2).

Results

After five postoperative days, the vaginal length was six to seven cm. Vaginal dilators were used after discharge from the hospital. She was able to have vaginal intercourse three weeks after the surgery. Long-term follow-up revealed a normal and satisfactory sexual relationship.

Discussion

MRKH syndrome is the second most frequent cause of one in 1,500 to one in 4,000 female births [2]. It is associated with normal ovarian function, normal female karyotype, and presence of secondary sexual characteristics. The syndrome is due to congenital aplasia of the Müllerian ducts. Congenital anomalies of the upper urinary tract may occur in 30%-40% of cases. Structural anomalies of the vagina and urogenital system are often challenging to diagnose and treat [3]. This is illustrated in the present case, where the diagnosis of MRKH syndrome was missed when the patient first complained of primary amenorrhea. Women with MRKH syndrome may also present with concerns of dyspareunia or the inability to have intercourse. However, in this cultural context, sexual issues are rarely revealed and discussed. In addition, sexual ignorance, as in the current case, is another contributing factor for the missed diagnosis. With respect to the treatment of MRKH syndrome, many non-surgical and surgical approaches exist. The aim is to create a neovagina of adequate size to allow normal sexual intercourse. Gradual dilatation with the use of dilators by the patient was described by Frank in 1938 [4]. Surgical corrections include a list of different techniques described in the literature [5]. The McIndoe procedure uses skin graft to cover a mold inserted into a surgically-created space between the bladder and rectum [6]. Recent evidence suggests that the modified laparoscopic Vecchietti operation is simple, safe, and effective [7]. In conclusion, urethral sex is documented for the first time in a case of previously undiagnosed MRKH syndrome.

Revised manuscript accepted for publication September 1, 2012

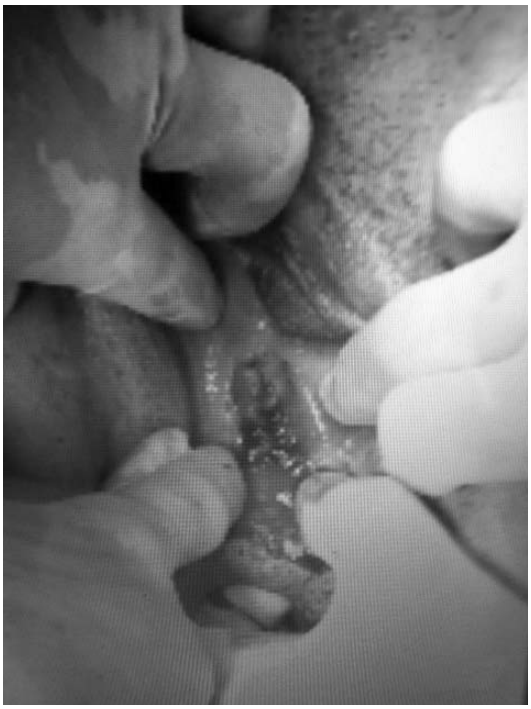

Figure 1. — Absent vagina.

Figure 2. — Urethral dilatation and a dilator in the created neovagina.

References

[1] Brucker S.Y., Gegusch M., Zubke W., Rall K., Gauwerky J.F., Wallwiener D.: "Neovagina creation in vaginal agenesis: development of a new laparoscopic Vecchietti-based procedure and optimized instruments in a prospective comparative interventional study in 101 patients". *Fertil. Steril.*, 2008, *90,* 1940.

[2] Aittomaki C., Eroila H., Kajanoja P.: "A population-based study of the incidence of Mullerian aplasia in Finland". *Fertil. Steril.,* 2001, *76*, 624.

[3] Klingele C.J., Gebhart J.B., Croak A.J., DiMarco C.S., Timothy G., Lesnick T.G., Lee R.A.: "McIndoe procedure for vaginal agenesis: Long-term outcome and effect on quality of life". *Am. J. Obstet. Gynecol*., 2003, *189,* 1569.

[4] Frank R.T.: "The formation of an artificial vagina without an operation". *Am. J. Obstet. Gynecol.*, 1938, *35,* 1053.

[5] Fedele L., Bianchi S., Zanconato G., Raffaelli R.: "Laparoscopic creation of a neovagina in patients with Rokitansky syndrome: analysis of 52 cases". *Fertil. Steril*., 2000, *74,* 384.

[6] McIndoe A.H., Bannister J.B.: "An operation for cure of congenital absence of the vagina". *J. Obstet. Gynaecol. Br. Emp.*, 1938, *45,* 490.

[7] Fedele L., Bianchi S., Frontino G., Fontana E., Restelli E., Bruni V.: "The laparoscopic Vecchietti's modified technique in Rokitansky syndrome: anatomic, functional, and sexual long-term results". *Am. J. Obstet. Gynecol*., 2008, *198,* 377.

Address reprint requests to:
A.A. ROUZI, M.D.
P.O. Box 80215
Jeddah, 21589 (Saudi Arabia)
e-mail: aarouzi@gmail.com