Laparoscopic sacrocolpopexy in a patient with vault prolapse of the sigmoid stump after vaginoplasty in Mayer-Rokitansky-Küster-Hauser syndrome: A case report

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Abstract

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a rare congenital anomaly that results in Müllerian agenesis that affects the uterus and upper two-thirds of the vagina. Sigmoid vaginoplasty is a surgical treatment option; however, vaginal prolapse may result as a complication of the sigmoid neovagina. There are no standards for treatment due to the rarity of this condition. We present the case of a 59-year-old woman with a history of sigmoid vaginoplasty who underwent laparoscopic sacrocolpopexy (LSC) for grade IV sigmoid stump prolapse. The patient had a successful outcome and no evidence of recurrent prolapse. This clinical case reveals the feasibility of LSC as a surgical treatment for sigmoid stump prolapses in patients with MRKH syndrome.

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1. Introduction

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a disorder that presents as Müllerian aplasia, agenesis, or vaginal agenesis, with an incidence of 1 per 4500–5000 females [1]. Müllerian agenesis is caused by embryonic underdevelopment of the Müllerian duct, resulting in agenesis, atresia of the vagina, uterus, or both. Primary vaginal elongation by dilation is the appropriate first-line approach in most patients, with a success rate of 90–96% [2]. Surgical treatment is preferred in patients for whom vaginal dilation is unsuccessful.

The surgical creation of a neovagina is performed in either late adolescence or young adulthood. Several surgical techniques may be used to create a neovagina; however, vaginoplasty has more complications than vaginal dilation, such as bladder or rectal perforation, graft necrosis fistula, diversion colitis, and adenocarcinoma. Prolapse of an artificial vagina derived from the sigmoid colon is rare [3].

We present a case of sigmoid neovaginal prolapse treated with laparoscopic sacrocolpopexy (LSC).

2. Case Presentation

A 59-year-old woman complained of the sensation of vaginal bulge for 5 years. She had a history of MRKH syndrome and had undergone sigmoid neovaginal construction at the age of 27 years. She was subsequently diagnosed with grade IV apical prolapse of the sigmoid stump (Aa 3, Ap 4, C 4, gh 3, pb 3, tvl 5, Ba 1, Bp 2), according to the pelvic organ prolapse quantification (POP-Q). She had not previously undergone any surgery for prolapse. To achieve the best surgical outcome and reduce the risk of recurrence, LSC was chosen as the surgical technique.

2.1. Surgical Procedure

The patient was kept in a 25-degree Trendelenburg position under general anesthesia. Four abdominal trocars were inserted at the infraumbilicus (10 mm), halfway between the umbilicus and the pubic symphysis (5 mm), and the left iliac as well as the right iliac fossa (5 mm each). The peritoneal and abdominal adhesions involving the bowels and abdominal wall near the trocar insertion were lysed.

The sigmoid neovagina was represented by thin-walled bowel tissue 7 cm in length (Fig. 1). The neovagina was located at the backside of the band of the uterus, which was attached to the atrophic ovaries (Fig. 1B). The peritoneum on the anterior neovaginal wall was dissected from the apex to the deepest part between the neovagina and uterus band tissue.

Abbreviations: MRKH syndrome, Mayer-Rokitansky-Küster-Hauser syndrome; LSC, laparoscopic sacrocolpopexy; NTR, native tissue repair.

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A self-cut strip of polypropylene mesh (Polyform®; Boston Scientific, Marlborough, MA) was inserted, and the end of the mesh was attached to the deepest part of the anterior neovaginal wall with three stitches with 3–0 absorbable suture. Additional stitches with 3–0 absorbable suture were placed to facilitate fixation. Fig. 1D: Posterior mesh fixation. Mesh was fixed to the lateral part of fat of the neovagina using 3–0 polyester suture bilaterally. Fig. 1E: The mesh was fixed at the promontory of the sacrum using 1–0 polyester suture.

Fig. 1. A: Vaginal speculum was inserted to facilitate recognition of the neovagina (arrow). The band of the uterus was grasped by forceps. Fig. 1B: Arrow shows both ovaries and the band of the uterus. Fig. 1C: Anterior mesh fixation. Additional stitches with 3–0 absorbable suture were placed to facilitate fixation. Fig. 1D: Posterior mesh fixation. Mesh was fixed to the lateral part of fat of the neovagina using 3–0 polyester suture. Fig. 1E: Anterior and posterior meshes were connected using 3–0 polyester suture bilaterally. Fig. 1F: The mesh was fixed at the promontory of the sacrum using 1–0 polyester suture.

The total operative time was 224 min, while the duration from dissection to peritonealization was 90 min. There were no immediate intraoperative complications. The estimated blood loss was 5 mL. The patient was discharged on the 5th postoperative day without any complications.

2.2. Follow-Up

Follow-up at 2 months, and at 1, 2, and 3 years showed no evidence of the recurrence of pelvic organ prolapse or any mesh-related complications. The patient did not present with any voiding or defecatory problems at the follow-up visits. POP-Q at 3-year follow-up was at stage I (Aa -3, Ap -3, C -8, gh 2, bp 3, trv 8, Ba -2, Bp -2).

3. Discussion

The vaginal canal in a patient with MRKH syndrome is either absent or markedly shortened and blind-ended. The initial approach, as considered by the American College of Obstetricians and Gynecologists (ACOG), is a vaginal elongation by dilation [4]. For patients with unsuccessful vaginal dilation, there are several surgical methods for creating a neovagina, such as sigmoid vaginoplasty. Edmond et al. described, in a review of 11 papers, the outcome of 179 patients with sigmoid neovagina, and of those, nine patients had a subsequent neovaginal prolapse [5]. However, to date, there has been no optimal treatment for neovaginal prolapse due to the rarity of the cases.

The etiology of neovaginal prolapse is unknown. Normally, the vagina is supported at three levels: the apex, middle, and lower parts, which consist of the ligaments, fascia, and muscles [6]. However, it has been hypothesized that patients with MRKH syndrome have congenital absence of apical and lateral anatomic support. The lack of suspensory or lateral support for the neovagina can result in vaginal prolapse. Sexual activity may also result in a progressive lengthening of the neovagina, which may disrupt the neovaginal fibrotic adhesions, and the absence of good anchorage may lead to prolapse [7].

Native tissue repair (NTR) has been attempted for the treatment of neovaginal prolapse, but the precise modalities differ in the literature. Freundt et al. described the suspension of the neovagina to the Cooper ligament in three cases; however, only one of the cases was treated successfully with this method alone [8]. Calcagno et al. reported a good anatomical and functional outcome of vaginal sacrospinous ligament suspension in the early stage of neovaginal prolapse after 20-month follow-up [9]. Similarly, Yokomizo et al. reported two patients with prolapse of sigmoid neovagina. One underwent resection of the redundant sigmoid and an abdominal suspension procedure, but there was a recurrence. The other patient was treated with the removal of an entire sigmoid neovagina and subsequent reconstruction used a pudendal thigh flap. No recurrence of prolapse was reported in the latter case [10]. Zhu et al. reported the outcome of bilateral iliacococcygeus neovaginal apical suspension. There was no recurrence of prolapse two years after surgery, but the patient had a low PISQ-12 (Prolapse/Urinary Incontinence Sexual Questionnaire-12) score [11].

NTR for general vaginal prolapse is reported to have failure rates as high as 30% [12]. Therefore, in an attempt to improve anatomical cure rates, synthetic mesh grafts have been developed. LSC has a better anatomical outcome and long-term success rate [13]. Recently, we showed a good outcome of LSC, with low rates of subjective and objective recurrence and of surgical complications. Our LSC for general vaginal prolapse is standardized on the use of double mesh fixation due to the systematic treatment of the three compartments of the pelvis (anterior, apical, and posterior) [14]. In our case, the patient had a grade IV neovaginal prolapse and she underwent LSC using separate anterior and posterior mesh fixation, which is similar to the case reported by Popov et al. [15].

In a systematic review, Kondo et al. reported successful outcomes of LSC as a treatment for recurrent sigmoid neovaginal prolapse. At 6-month follow-up, no recurrence was reported and the patient had a satisfactory sex life. Contrary to our case, Kondo et al. advocated the use of an anterior mesh only, in order to avoid mesh-related complications.
such as erosion and stricture. They also mentioned that the vascularization of the neovagina was supplied from its posterior aspect, which is why they chose to avoid the placement of a posterior mesh [16]. Although separate anterior and posterior meshes were used in the present case, no mesh-related problems were observed in the patient after 3 years of follow-up. The authors plan to follow up the patient for several years further to assess the long-term outcomes (prolapse symptoms and mesh-related complications).

Contributors

Manisha Yadav drafted the manuscript and contributed substantially to revision of the manuscript.

Tokumasa Hayashi drafted the manuscript and contributed substantially to revision of the manuscript.

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Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

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References

[1] L. Fontana, B. Gentilini, L. Fedele, C. Gervasini, M. Miozzo, Genetics of Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, Clin. Genet. 91 (2) (2017) 233–246, https://doi.org/10.1111/cge.12883.
[2] C.P. Roberts, M.J. Haber, J.A. Rock, Vaginal creation for müllerian agenesis, Am. J. Obstet. Gynecol. 185 (6) (2001) 1349–1352, https://doi.org/10.1067/mob.2001.119073.
[3] ACOG Committee Opinion Number 728, Müllerian agenesis: Diagnosis, management, and treatment, Obstet. Gynecol. 131 (1) (2018) e35–e42, https://doi.org/10.1097/AOG.0000000000002456.
[4] Committee opinion: no. 562: müllerian agenesis: diagnosis, management, and treatment, Obstet. Gynecol. 121 (5) (2013) 1134–1137, https://doi.org/10.1097/01.AOG.0000429659.93470.ed.
[5] D.K. Edmonds, C.I. Rose, M.G. Lipton, J. Quek, Mayer-Rokitansky-Küster-Hauser syndrome: a review of 245 consecutive cases managed by a multidisciplinary approach with vaginal dilators, Fertil. Steril. 97 (3) (2012) 686–690, https://doi.org/10.1016/j.jfertstim.2011.12.038.
[6] J.O. DeLancey, Anatomic aspects of vaginal eversion after hysterectomy, Am. J. Obstet. Gynecol. 166 (8 Pt 1) (1992) 1717–1724, https://doi.org/10.1016/0002-9378(92)90315-3.
[7] P. Oppelt, S.P. Renner, A. Kellermann, S. Brucker, G.A. Hauser, K.S. Ludwig, P.L. Strissel, R. Strick, D. Wallwiener, M.W. Beckmann, Clinical aspects of Mayer-Rokitansky-Küster-Hauser syndrome: recommendations for clinical diagnosis and staging, Hum. Reprod. 21 (3) (2006) 792–797, https://doi.org/10.1093/humrep/dei381.
[8] I. Freundt, T.A. Tooolenaar, H. Jierkel, A.C. Drogendijk, F.J. Huikeshoven, Prolapse of the sigmoid neovagina: report of three cases, Obstet. Gynecol. 83 (5 Pt 2) (1994) 876–879.
[9] M. Calcagno, M. Pastore, F. Bellati, F. Ploiti, D. Maffucci, T. Boni, P.B. Panici, Early prolapse of a neovagina created with self-dilatation and treated with sacrospinous ligament suspension in a patient with Mayer-Rokitansky-Küster-Hauser syndrome: a case report, Fertil. Steril. 93 (1) (2010) https://doi.org/10.1016/j.fertnstert.2009.10.010.267.e1–4.
[10] R. Yokomizu, T. Murakami, H. Nairou, A. Yamada, Treatment for prolapse of the sigmoid neovagina in Mayer-Rokitansky-Küster-Hauser syndrome, Obstet. Gynecol. 100 (5 Pt 2) (2002) 1085–1087, https://doi.org/10.1016/S0029-7844(02)00720-2.
[11] L. Zhu, N. Chen, J. Lang, Vault prolapse of sigmoid neovagina 26 years after vaginoplasty in Mayer-Rokitansky-Küster-Hauser syndrome: a case report, Int. Urogynecol. J. 24 (1) (2013) 179–180, https://doi.org/10.1007/s00192-012-1755-6.
[12] A.L. Olsen, V.J. Smith, J.O. Bergstrom, J.C. Colling, A.L. Clark, Epidemiology of surgically managed pelvic organ prolapse and urinary incontinence, Obstet. Gynecol. 89 (4) (1997) 501–506, https://doi.org/10.1016/S0029-7844(97)00058-6.
[13] D. Sarlos, L. Kots, G. Ryu, G. Schaer, Long-term follow-up of laparoscopic sacrocolpopexy, Int. Urogynecol. J. 25 (9) (2014) 1207–1212, https://doi.org/10.1007/s00192-014-2369-y.
[14] Y. Sawada, Y. Kitagawa, T. Hayashi, S. Tokiwa, M. Nagae, A.R. Cortes, M. Nomura, Clinical outcomes after laparoscopic sacrocolpopexy for pelvic organ prolapse: a 3-year follow-up study, Int. J. Urol. 28 (2) (2021) 216–219, https://doi.org/10.1111/jiu.14435.
[15] A. Popov, D. Guma, K. Mironenko, B. Skibodyanyuk, T. Mananikova, A. Fedorov, S. Tyurina, A. Koval, Laparoscopic sacrocolpopexy in a patient with vault prolapse of the sigmoid stump, Int. Urogynecol. J. 27 (2) (2016) 315–316, https://doi.org/10.1007/s00192-015-2766-x.
[16] W. Kondo, R. Ribeiro, K. Tsunamunuma, M.T. Zomer, Laparoscopic promontofixation for the treatment of recurrent sigmoid neovaginal prolapse: case report and systematic review of the literature, J. Minim. Invasive Gynecol. 19 (2) (2012) 176–182, https://doi.org/10.1016/j.jmig.2011.12.012.