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Case Report

Laparoscopic uteroneovaginal anastomosis in Mayer-Rokitansky-Kuster-Hauser syndrome with functioning horns: a case report

Polly Chatterjee[1], Avishek Bhadra[2], Abhinibesh Chatterjee[1]*, Rumela Biswas[3]

[1]Department of Gynaecology and Obstetrics, Eskag Sanjeevani Baghbazar, Kolkata, West Bengal, India
[2]Department of Gynaecology and Obstetrics, R. G. Kar Medical College and Hospital, Kolkata, West Bengal, India
[3]Department of Gynaecology and Obstetrics, Medical College and Hospital, Kolkata, West Bengal, India

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***Correspondence:**
Dr. Abhinibesh Chatterjee,
E-mail: acbb258@gmail.com

ABSTRACT

Background: Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is characterized by congenital absence of the upper two-thirds of the vagina and a rudimentary or absent uterus, resulting in primary amenorrhea while maintaining normal secondary sexual characteristics. This condition arises from the agenesis or hypoplasia of the mullerian ducts and has an incidence of 1 in 4,000 to 5,000 female births. While most patients possess a non-functioning rudimentary uterus, a minority have functioning endometrium, often leading to complications such as pelvic pain due to hematometra or endometriosis. Surgical interventions typically involve the excision of rudimentary uterine structures, which may relieve pain but eliminate menstruation and the potential for pregnancy. This case details a novel two-stage laparoscopic technique to restore utero-vaginal continuity, thereby enabling menstruation and potential future pregnancies. A 28-year-old woman with MRKH syndrome presented with cyclic abdominal pain since adolescence. Imaging revealed bilateral rudimentary uterine structures with functioning endometrium. A two-stage laparoscopic procedure was performed: initially creating a neovagina and anastomosing the uterine components, followed by re-anastomosis and placement of a copper T multiload 375. The patient experienced her first menstruation 20 days post second surgery, achieving regular cycles thereafter. Follow-up ultrasounds confirmed normal uterine morphology and endometrial thickness. This case underscores the importance of a holistic management approach for MRKH syndrome, and introducing innovative surgical techniques. Our reproducible laparoscopic procedure demonstrates promising outcomes for restoring menstrual function and reproductive potential in patients with a functioning endometrium.

Keywords: Mayer-rokitansky-kuster-hauser, Laparoscopy, Restoration, Functional endometrium, Congenital mullerian anomalies

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome (mullerian agenesis) is a malformation complex characterized by congenital absence of the upper two-thirds of the vagina and an absent or rudimentary uterus in women who present with primary amenorrhea having normal development of secondary sexual characteristics (functioning ovaries) and a normal female karyotype. It results from the agenesis or hypoplasia of the mullerian (paramesonephric) ducts. It has an incidence of 1 in 4000-5000 female births.[1] Abnormalities of the kidneys and other organ systems are often associated. Majority of the patients have a rudimentary, non-functioning uterus, but 2% to 7% have a uterus with functioning endometrium.[2] The latter condition is usually detected at puberty when patients present with cyclic or chronic abdomino-pelvic pain secondary to hematometra, hematosalpinx or endometriosis. In patients having a uterus with a functioning endometrium one available surgical treatment is radical excision of the rudimentary uterus(es), which successfully relieves the pain but renders the patient

unable to menstruate or carry a pregnancy.[2] Since 1938, when it was first reported, many operative techniques of continuity reestablishment have been proposed and published.

Our case is unique in that we could successfully perform this using our own easily reproducible technique in a two-stage total laparoscopic procedure and copper T multiload 375 without the copper coil used to preserve the uterus, thus restoring menstrual function and providing her a possibility to carry pregnancy in future.

CASE REPORT

A 28-year-old woman presented with lower abdominal pain occurring cyclically for seven to eight days each month since, 14 years of age. She was a diagnosed case of MRKH syndrome on ultrasound since then, when she presented for the first time with primary amenorrhea with normal secondary sexual characteristics, with age and sex appropriate height and weight. Genetic testing result showed female karyotype of 46, XX,22ps+ pattern. Physical examination revealed absent vagina. Abdominal ultrasound revealed hypoplastic uterus with bilateral mild bulky ovaries. Both kidneys were however, normal.

An MRI confirmed the presence of bilateral uterine structures with endometrial lining with no communication between them, adjacent to bilateral ovaries. Right structure measured 3.3×2.7 cm with 4.4 mm endometrium while left structure measured 3.3×2.5 cm with 4.1 mm endometrium. Rest of the uterus including cervix was absent. Bilateral ovaries showed polycystic changes (Figure 1).

A closed laparoscopy was performed. 4 ports were created: one supra-umbilical 10 mm port for the camera and two 5 mm ports one on each side, at the junction of medial two-thirds and lateral one-third of spin umbilical line-one each for the operating surgeon and the assistant surgeon. A third 5 mm port was created between these two in the infra umbilical region. A careful dissection of urinary bladder anteriorly and rectum posteriorly was performed using ultrasonic scalpel.

A peritoneal pull through neovagina was created. Bilateral uterine buds were mobilized medially, incised up to endometrium and anastomosed with each other with barbed sutures and ultimately purse string suturing with the created neovagina and lower part of formed uterus was done. A 14 Fr silicone catheter was passed through the neovagina and introduced into the endometrial cavity (inferior portion). Sutures were placed around the catheter to complete the uterine anastomosis.

The patient was discharged on third post operative day, and was advised daily use of vaginal mould and conjugated estrogen tablets (0.625 mg) twice daily for four months. However, the silicone catheter expelled out spontaneously two months after her OT. Repeat US showed bulky anteverted uterus with two separate thick and cystic endometrial strips on transverse scan.

Patient was put on tablet estradiol valerate (0.5 mg) for 18 months but still had amenorrhea. MRI pelvis was done both before and after distending the vaginal canal with sterile water-soluble gel. The reconstructed uterus showed adenomyotic changes. Both uterine components showed normal morphology and endometrial cavities. Well-distended vaginal canal was seen. The gel did not cross cervical canal. Bilateral ovaries and other abdomen structures were normal.

Two years after the first operation, patient was admitted for laparoscopic re-anastomosis. Anteriorly bladder and posteriorly rectum were dissected down. Uterine fundus was incised with ultrasonic scalpel and cavity was found to be small and blind. Cranial part of vagina incised with ultrasonic scalpel over a sponge on holder. Posterior uterine wall sutured with the vagina with Prolene 2-0. Silicone catheter and copper T multiload 375 were introduced through the vagina into the uterine cavity and fixed with PDS II suture; the copper coil was removed through the vaginal route (Figure 2).

Anterior walls of uterus and vagina were reconstructed with PDS II (interrupted sutures), keeping the catheter and Copper T arms in the cavity and stem in the anastomotic area (Figure 3). Surgical was placed over raw areas for better healing. Patient was discharged on the fifth post operative day on tablet Ethinylestradiol & tablet Norgestrel.

Patient had her first menstruation 20 days after her second laparoscopy followed by monthly cycles till now with medical assistance (tab Ethinylestradiol & Norgestrel). Catheter was removed 90 days after her operation, while Copper T stem was left inside to maintain the utero-vaginal continuity. Post-operative ultrasound was done after 3 months which showed normal sized uterus with 6.7 mm thick endometrium. Bilateral ovaries where normal in size. Patient is under regular follow-up and is doing well at present.

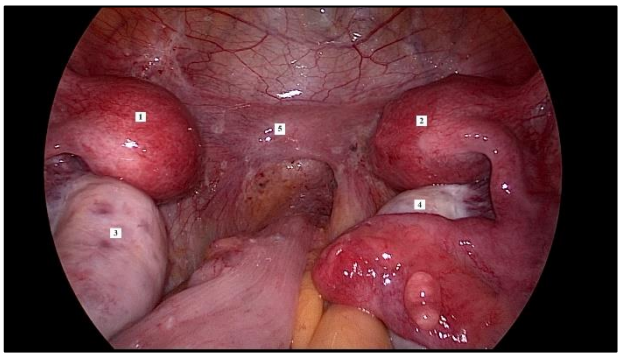

Figure 1: Global view of the pelvis showing (1) left uterine horn, (2) right uterine horn, (3) left ovary, (4) right ovary and (5) peritoneal plate.

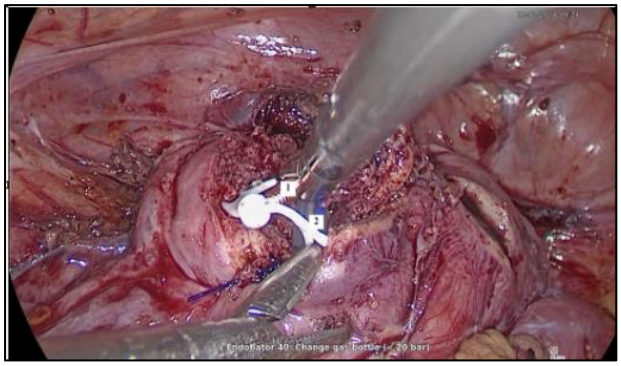

Figure 2: (1) Multiload copper T 375 and (2) silicone catheter being introduced into the endometrial cavity.

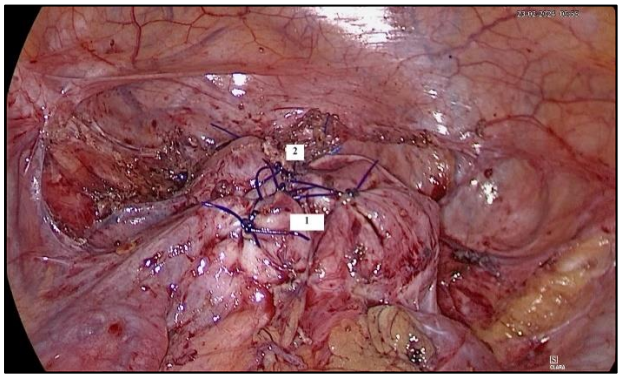

Figure 3: Final laparoscopic view showing (1) reconstructed uterus and (2) peritoneal pull-through neovagina anastomosed with PDS II suture.

DISCUSSION

MRKH syndrome should be managed in a holistic approach including psychological support. Laparoscopy has been used since long for diagnostic purposes, for the creation of neovaginas or for resection of rudimentary horns.[3] With the advancement of laparoscopic procedures, utero-vaginal reconstruction, although challenging, is gradually gaining momentum. Early diagnosis appears necessary to avoid the development of pelvic-associated lesions that may lead to aggressive surgeries such as adnexectomy or hysterectomy.[2] Re-establishing utero-vaginal continuity has a considerable complication rate.[4] The most common are secondary vaginal stenosis and cervical stenosis causing dysmenorrhea and thus requiring multiple canalization procedures.[2] This patient presented first at adolescence with primary amenorrhea and was advised for a vaginoplasty at a later date. However, MRI report prompted an attempt at utero-vaginal reconstruction which would restore the patient's menstrual as well as sexual function. She was counselled regarding the possible complications that may arise during the long-term follow-up and was aware that recurrent obstruction may necessitate hysterectomy as the last therapeutic option.[2] Although we could establish the anatomy of the uterovaginal continuity in the first sitting, the second sitting successfully restored the menstrual function of the patient. The rarity of the syndrome implies that most treating centers only acquire preference and thus expertise for a single procedure.[5] This may result in reporting and publication biases in the available literature concerning outcome and complications and it complicates the initiation of comparative studies including both surgical and non-surgical approaches.[5]

CONCLUSION

We have shown that a total laparoscopic procedure is invaluable in treating rare anomalies such as the MRKH syndrome particularly with a functioning endometrium. We developed this easily reproducible laparoscopic technique to preserve the uterus. Our unique technique of using the multiload copper T stem (without the copper to prevent inflammation) acts as a stent which prevents further stenosis and ensures regular menstruation. The addition of postoperative estrogen also helps in increasing the size of the uterus with functioning endometrium.

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