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LAPAROSCOPIC VAGINOPLASTY USING MODIFIED PERITONEAL PULL DOWN TECHNIQUE WITH EXCISION OF RIGHT FUNCTIONING UTERINE HORN IN A CASE OF MRKH SYNDROME

Obstetrics & Gynaecology

Dr Shaikh Madiha Tariq* Third Year Post Graduate Resident, Department Of Obstetrics And Gynaecology, Seth GS Medical College And KEM hospital, Mumbai. *Corresponding Author

Dr Hemangi Kansaria Professor And Head Of Unit, Department Of Obstetrics And Gynaecology, Seth GS Medical College And KEM hospital, Mumbai.

Dr Karishma Gogada Senior Resident ,department Of Obstetrics And Gynaecology, Seth GS Medical College And KEM hospital, Mumbai.

Dr Sameer Rege Professor And Head Of Unit, Department Of Surgery, Seth GS Medical College And KEM Hospital, Mumbai.

ABSTRACT

Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome is a congenital disorder characterized by uterovaginal agenesis in females with normal secondary sexual characteristics and a 46,XX karyotype. We present the case of an 18-year-old female with primary amenorrhea and cyclic abdominal pain. Imaging revealed Type I MRKH with bilateral rudimentary uterine horns, one containing functional endometrium. She underwent laparoscopic vaginoplasty using a modified Davydov technique with excision of the symptomatic uterine horn. The postoperative course was uneventful, and a 10 cm neovagina was successfully created. This case highlights the efficacy of a minimally invasive approach in managing MRKH with a functioning uterine remnant.

KEYWORDS

MRKH syndrome, laparoscopic vaginoplasty, primary amenorrhea, rudimentary uterine horn

INTRODUCTION

MRKHS (Mayer-Rokitansky-Küster-Hauser syndrome), also called as Müllerian agenesis or Müllerian aplasia, is the term used to describe congenital absence of both uterus and vagina. Uterus, cervix, and upper portion of the vagina are also missing. Given their distinct embryonic origin, normal ovaries usually survive, and part of the distal fallopian tubes are visible. Only a tiny, primitive Müllerian horn devoid of endometrial activity is present in the majority of patients with Müllerian agenesis. However, active endometrium develops in 2–7% of women with this disease, and patients usually suffer with cyclic stomach pain [1]. The symptomatic rudimentary horn must be surgically removed. Traditional conception is impossible with Müllerian agenesis; however, gestational surrogacy, oocyte retrieval, and fertilisation can all help produce pregnancy. [2]

2. Case Report

18-year-old, unmarried , nulligravida, presented to tertiary care centre with chief complaints of primary amenorrhea with intermittent mild to moderate intensity dull aching pain in lower abdomen every month lasting for one day. Thelarche began at age 12 and pubarche at age 13, according to the history of pubertal development. The medical or surgical history was insignificant. None of the family members had a history of congenital abnormalities. There was no history of hormone or radiation therapy exposure during pregnancy, and the mother's obstetric history was unremarkable. At 16 years of age, patient was evaluated for primary amenorrhea, MRI revealed aplastic uterus with rudimentary horn and absent vagina (V4).

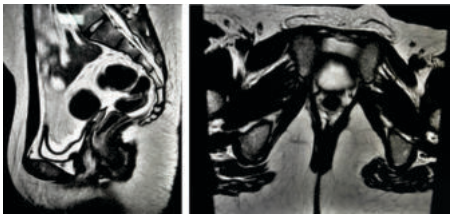

Figure 1. A) Saggital section of MRI pelvis showing absence of vagina B) Coronal section of MRI pelvis showing rudimentary horns interconnected by fibrous band

Her weight was 55kg, her height was 152cm, as well as her arm span has been normal based on a general medical examination. The external female genitalia were grossly normal, the breasts were well-developed, and the pubic and axillary hair distribution was as per age (Tanner stage V). Only the urethral aperture and a vaginal indentation were visible upon local examination. The RFT (renal function test), LFT (liver function test), as well as complete blood counts, were all within normal limits. Hormonal profile was within reference range (serum luteinizing hormone, follicle-stimulating hormone,anti-müllerian hormone, thyroid-stimulating hormone, prolactin), confirming normal female reproductive hormones. Karyotype was found to be 46XX.

Review MRI at our institute revealed mullerian agenesis with uterine remnant on both sides, right uterine remnant having functional endometrial cavity. No such functional uterine endometrial cavity in left uterine remnant. Cervical and vaginal agenesis (U5aC4V4) was also noted . Additionally, both ovaries were normal in appearance, and no urinary tract anomalies were noted. Hence, confirming the diagnosis of Type 1 MRKH syndrome. Post psychological counselling regarding fertility and childbearing, patient and relatives agreed to vaginoplasty. Her parents revealed that she will get married within 3 to 4 months. The patient underwent Laparoscopic vaginoplasty using modified peritoneal pulldown techniques with uterine strand incision with excision of right functioning uterine horn after informed consent. Laparoscopic examination revealed bilateral uterine horns interconnected by a fibrous band, normal bilateral fallopian tubes and ovaries.

A 5 cm transverse incision was made in front of the band connecting the uterine horns, and a anterior and posterior peritoneal fold was mobilized. Right rudimentary uterine horn excised as it showed functioning endometrium on MRI. An incision was given from vaginal end. The rectovesical space was dissected. Laparoscopically uterine strand was then divided, and posterior and anterior flaps pulled down and sutured to neovaginal introitus. Neovaginal apex was created using absorbable sutures while ensuring an optimal length. A resin mould was placed to preserve vaginal patency, and the uterine strands have been fastened to lateral sides of neovaginal apex to avoid prolapse.

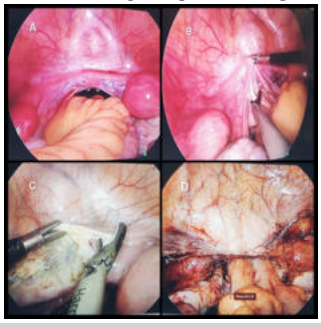

Figure 2. A) laparoscopic view showing two rudimentary uterine horn interconnected by fibrous septa. B) dissection of posterior peritoneal fold. C) dissection of rectovesical space. D) Posterior peritoneal pull through

On the seventh postoperative day, the mould was removed, and a neovaginal examination and irrigation were carried out while under moderate sedation. Urinary catheter remained in place for seven days. The patient was advised self dilation using silicon vaginal dilators, to be kept in situ for 12-24 hours. The postoperative course was uneventful, and she was discharged with a neovaginal length of 10 cm. Follow-up at 1 month scheduled.

Figure 3.- Per speculum examination on day 30 post procedure, neovaginal length of 10 cm

DISCUSSION:

Müllerian agenesis, or MRKH syndrome, affects 1 in 4,500–5,000 females. There is vaginal and uterine absence or hypoplasia due to incomplete müllerian duct development. The vaginal canal, which frequently manifests as a dimple beneath the urethra, is either nonexistent or reduced. There may be horns or a remnant of the uterus, with or without an endometrial cavity. Ovaries, derived from a different embryologic source, are typically normal but may be in atypical locations[1]

This condition is classified into two types: Type A, which involves isolated Müllerian duct anomalies, and Type B, which is associated with renal and/or skeletal abnormalities. When a patient presents with primary amenorrhoea during adolescence, the diagnosis is usually made. Early detection of this condition is greatly aided by radiological imaging.[3]

Most cases are sporadic, though familial clustering suggests a genetic basis with autosomal-dominant inheritance and reduced penetrance. Recurrent chromosomal abnormalities in 1q21.1, 16p11.2, 17q12, 22q11.21 were detected in MRKH patients using high-resolution array-CGH and MLPA. LHX1, TBX6, RBM8A mutations were detected in these regions. In patients with hyperandrogenemia, WNT4 mutations were found, while WNT9B mutations were also linked to MRKH. [4]

MRKH syndrome presents as primary amenorrhea with normal puberty, indicating intact ovarian function. Diagnosis involves ultrasound for uterine and vaginal abnormalities, with MRI confirming details and detecting anomalies in Type B cases. Genetic testing confirms a 46, XX karyotype. [1]

Treatment and psychological counselling are part of the management of MRKH syndrome. Non-surgical methods, like Frank's and Ingram's techniques, use vaginal dilators and are first-line treatments due to high success rates (7892%). Numerous surgical methods have been explained. There are two categories of these methods: a. Vechietti procedure (open/laparoscopic) based on traction b. Graft-based: Intestinal vaginoplasty (mixed vaginal and open/laparoscopic approach), Abbe-McIndoe technique (vaginal approach, skin grafting), and Davydov procedure (combined vaginal and laparoscopic approach, peritoneal graft). Research indicates that the sexual results of surgery and non-surgical procedures are comparable.[5]

In the Davydov colpopoiesis technique, an epithelialised neovagina is created from the patient's peritoneum. The first step in this three-step process is to dissect the area between the rectum, bladder, and urethra. Next, the peritoneum is mobilised and sutured to the vaginal vestibulum's margins. Because postoperative dilatation is necessary, this technique is especially appropriate for women who have had significant pelvic procedures. It is regarded as one of the most straightforward, secure, and quick methods available, with several benefits, like enough lubrication that results in satisfying intercourse, the lack of granulation tissue, and the avoidance of scarring. Although certain incidences of ascending infections have been documented, patients also benefit from minimum bleeding, shorter hospital stays, less postoperative pain, speedy recovery, or a positive cosmetic outcome.[6]

The presence of the uterine strand during surgery frequently causes granulation tissue to form at the neovaginal apex, which also affects the final neovaginal length.

In this instance, the alteration improves on a laparoscopic vaginoplasty procedure that has already been documented. The uterine strand was cut longitudinally and transversely, the anterior and posterior peritoneum were extensively mobilised, and supravesical and suprarectal peritoneum were sutured to construct neovaginal apex at appropriate length. Furthermore, further structural support is provided by bilaterally fixing incised uterine strand to neovaginal apex. This method reduces granulation at the apex, permits a neovagina of any necessary size, guarantees sufficient peritoneal extension to the vaginal introitus, and strengthens the structure to avoid prolapse and perforation during sexual activity[6]

Due to non-compliance and unwillingness to use Frank dilation therapy, our patient underwent the Davydov procedure with modified peritoneal pull through chosen for its simplicity, safety, and effectiveness in MRKH syndrome. [7]The surgery was complication free with minimal blood loss, achieving a 10 cm vaginal length. Postoperatively, she was advised daily self-dilation (10 × 3 cm). At one month, a 10 cm length was maintained. Follow-ups have been scheduled at 3,6, 12, 24 months to assess long-term outcomes.

One of the main causes of irreversible uterine infertility is MRKH syndrome. Alternatives for fertility involve adoption, surrogate pregnancy, as well as successful births via uterine transplant. Tissue Engineering Technology, a major development in regenerative medicine, includes growing smooth muscle and vaginal epithelial cells on biodegradable scaffolds to create vaginal tissue for surgical repair in patients with MRKH. [8]

CONCLUSION

Müllerian agenesis is rare congenital condition that requires both significant psychosocial assistance and a teambased medical treatment. Clear communication and emotional strength are crucial for effective intervention. Noninvasive vaginal dilation remains the primary treatment, while minimally invasive neovaginal reconstruction methods, particularly the laparoscopic Davydov procedure, are regarded as reliable and efficient. The success of treatment depends on the effectiveness of the initial surgery, proper postoperative care, and patient compliance. Fertility alternatives include adoption and gestational surrogacy, while uterine transplantation is under investigation, though its practicality remains uncertain. In the future, bioengineered autologous vaginal reconstruction may emerge as a promising solution.

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