Acta Scientific Clinical Case Reports

Volume 2 Issue 6 June 2021

Case Report

Laparoscopic Vaginoplasty after a Failed Vaginoplasty in a Case of MRKH Syndrome: A Case Report

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Received: May 05, 2021

Published: May 24, 2021

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Abstract

Background: Mayer-Rokitansky-Kuster-Hauser syndrome is a rare disorder characterized by congenital aplasia of vagina with possible concurrent absence of uterus and/or fallopian tubes in the setting of a normal female karyotype. Patients usually present with primary amenorrhoea in presence of normal secondary sexual characteristics and external genitalia. There are various methods for treating the associated vaginal agenesis, both non-surgical and surgical. Here we present a case of laparoscopic vaginoplasty using amnion graft in a patient with previous failed vaginoplasty.

Case Presentation: A 22 year old female diagnosed as a case of MRKH syndrome on basis of clinical and Magnetic Resonance Imaging findings with history of failed vaginoplasty 6 months earlier, came to outpatient department desirous of restoration of patency of vaginal canal. Laparoscopic assisted vaginoplasty with insertion of amnion graft was used to create a neovagina. Patient discharged after counselling for serial dilator therapy.

Conclusion: Vaginoplasty using amnion graft seems like a feasible option owing to its easy availability, low cost, safety, physiologic epithelialization of vaginal tract.

Keywords: MRKH Syndrome; Case Report; Primary Amenorrhoea; Vaginal Agenesis; Vaginoplasty; Amnion Graft

Abbreviations

MRKH: Mayer Rokitansky Kuster Hauser; MRI: Magnetic Resonance Imaging; MURCS: Mullerian agenesis, Renal Agenesis, Cervicothoracic Somite abnormalities; GRES: Genital Renal Ear Syndrome; OPD: Outpatient department

Background

Mayer-Rokitansky-Kuster-Hauser syndrome is a very rare disorder characterized by congenital aplasia of uterus, fallopian tubes and upper 2/3rd's of vagina with the presence of normal functioning ovaries. It is the second most common cause of primary amenorrhoea after gonadal dysgenesis [1]. A shallow vaginal pouch is present, and the fallopian tubes, ovaries, and secondary sex characteristics are normal [2]. This syndrome was first described by Mayer in 1829 and Rokitansky in 1838. Mullerian or paramesonephric ducts differentiate into fallopian tubes, uterus, cervix and upper part of vagina and any abnormalities during the development results in various malformations [1]. The incidence of MRKH syndrome has been estimated as 1 in 4500 female births. Isolated utero-vaginal aplasia is referred to as Rokitansky sequence or to type I (isolated) MRKH syndrome. Incomplete aplasia and/or associated with other malformations, is generally referred to as

MURCS association (or type II MRKH syndrome). In this case, the term GRES (Genital Renal Ear Syndrome) can also be used [3]. The typical initial presentation in MRKH is primary amenorrhea in an otherwise normally developed adolescent female. When physical examination findings are consistent with absent or hypoplastic vagina, the immediate differential diagnosis includes MRKH and complete androgen insensitivity syndrome, which is due to an inactivating mutation in the androgen receptor. Once the diagnosis of MRKH is suspected, imaging studies have a central role in unveiling the degree and extension of gynaecologic and extra-gynaecologic abnormalities [4]. Timing of neovagina creation should be deferred until late adolescence to allow consent and compliance. There are different approaches to treatment with respect to the type - whether conservative or surgical, access route - vaginal, transabdominal or laparoscopic, material used to cover neovagina - skin graft, peritoneum, intestinal tissue, amniotic membranes.

Here we present a case of a 22 year old female with MRKH Syndrome Type I, with a previous history of failed vaginoplasty in the past, for whom laparoscopy assisted vaginoplasty with amnion graft insertion was performed with success.

Case Presentation

A 22year old Asian female, came to OPD with complaint of primary amenorrhoea. On clinical examination, she had well developed secondary sexual characters, on local examination revealed obliteration vaginal passage with effective length less than 1 cm (Figure 1). MRI was done which revealed shallow vagina for a length of 2 cm from introitus, rest of the vagina was not distensible. Uterus was hypoplastic, 19 x 9 mm in size, bilateral ovaries were found normal. Imaging did not reveal any other malformations. Buccal mucosa smear revealed normal XX karyotype. Patient was diagnosed as a case of MRKH on the basis of clinical examination and above investigations. Patient had a history of vaginoplasty performed 6 months earlier, with vaginal dilators given post operatively for maintaining patency of vaginal passage. On her one month follow up, patient complained of narrowing of vaginal canal and passage of faecal matter through vagina. On clinical examination, a small rent in the rectum was palpable, probably due to dilator trauma. Subsequently a fistulogram was done which reported no fistulous communication between neo vagina and rectum with smooth flow of contrast which indicated that the rent had healed.

Figure 1: Clinical examination showing obliterated neo vagina after previous failed vaginoplasty.

Since the patient was planning marriage in the near future, patient was posted for Laparoscopy assisted vaginoplasty with amnion graft placement after all investigations for fitness for surgery. On laparoscopy, utricle was noted, bilateral fallopian tubes and ovaries were healthy, urinary bladder and rectum were found adherent (Figure 2). Dissection was performed in order to create a space between bladder and rectum by excising the fibrous tract after prior infiltration with vasopressin. Vaginal passage demarcation was done guided by a dilator passed through vagina. Vaginal opening was created and extended achieving an effective length of about 8 cm. After checking rectal integrity with methylene blue dye, peritoneal closure was done using Vicryl no 1 (Figure 3). Vaginal mould was created using a 20 ml syringe wrapped with gauze on which a condom was placed, on which amnion graft was wrapped. This was placed in the neovagina. Vulval stay sutures were taken to keep in place (Figure 4). Continuous draining bladder catheter was left in situ for 7 days. On post-operative day 8, the mould was replaced with a fresh one, which was kept for another week. Removal of the mould subsequently revealed that the amnion graft had taken up and was in healing phase with demonstrable vaginal depth up to 8 cm (Figure 5). The post-operative period was uneventful and the patient was discharged after counselling for serial dilator therapy. Subsequent one month and 6 months follow up visits demonstrated adequate epithelialization of the neovagina with no constriction. The patient was satisfied with using vaginal

dilators for maintaining patency of the neo-vagina. The patient and family were counselled about the benefits of early marriage following surgery.

Figure 2: Laparoscopic view of pelvis with arrow indicating the utricle adherent to the bladder and rectum. Fallopian tubes and ovaries appear normal.

Figure 3: Laparoscopic view of pelvis showing normal fallopian tubes and ovaries with peritoneal approximation after creation of neo vagina.

Figure 4: shows vulval approximation sutures taken after insertion of mould covered with amnion graft in the neo vagina.

Figure 5: Healing granulation tissue of neo vagina on per speculum examination after removal of mould with demonstrable vaginal length of 8 cm.

Discussion

Treatment consisting of creating a neovagina must be offered to patients only when they are ready to start sexual activity. Of the two main types of procedures, the first one consists of the creation of a new cavity and can be nonsurgical or surgical. The second is vaginal replacement with a pre-existing canal lined with a mucous membrane like bowel [5].

ACOG committee guidelines, 2018 recommend that primary vaginal elongation by dilatation is the appropriate first line approach in most patients because it is safer, patient controlled and more cost effective than surgery. Surgery should be reserved for patients who are unsuccessful with primary dilator therapy or who prefer surgery after a thorough informed consent discussion with her gynaecologic care provider and her guardians [6].

Since centuries, surgical creation of an artificial vagina is being attempted with varying success. Columbus was perhaps the first to describe the congenital absence of vagina in 1559. Dupuytrens was the first to report vaginoplasty using tampons to maintain patency. Abbes were the first to cover the mould with split thickness skin graft. However, it was McIndoe and Bannister who emphasized the need for dissecting adequate space, inlaying split thickness skin grafts over the moulds and most importantly continuous and prolonged dilatation during the contractile phase of healing [7].

The problem with skin graft is the additional scar to the patient, need for a plastic surgeon, special instruments for graft harvesting, and its non- physiologic nature because of hair growth. Various modifications of the original Abbe-McIndoe technique use differ-

ent grafts like amnion, oxidized cellulose, peritoneum, autologous buccal mucosa, and cultured autologous vaginal mucosal cells [8].

In the present case, amnion graft was used to create the neovagina because of some characteristics of amnion membrane which make it one of the ideal substances for this procedure. Amnion is readily available, and there is no need for additional incisions as at the donor site in the split skin graft technique. Furthermore, there have been no problems with immune rejection, because human amniotic epithelial cells do not express on their surface histocompatibility antigens such as HLA-A, -B, -C and -DR or _2- macroglobulin [2]. It has antimicrobial properties, has anti-fibroblastic activity and thus prevents fibrosis. It also has cell migration and growth promoting activity and it promotes epithelialization [8].

Sarwar., *et al*. 2010 conducted a study of vaginoplasty using amnion graft in 28 patients of MRKH. All patients except one (96.43%) had uneventful surgical procedures and successful outcomes. Follow up at 3 months was satisfactory in 89% patients, while 11% required a minor second procedure in the form of digital dilatation due to vaginal constriction secondary to poor compliance. Follow up at 6 months was satisfactory in 100% of patients in terms of anatomical and functional results [9].

Vatsa., *et al*. 2017 conducted a retrospective study on 50 women with MRKH who underwent neo-vaginoplasty. Modified McIndoe's vaginoplasty was done in all patients using human amnion graft. Mean (+/- SD) vaginal length after surgery was 8.2 +/- 1 cm. Mean vaginal length at 6month follow-up in sexually active patients was significantly longer as compared with patients who were not sexually active after surgery. Vaginal biopsy done in 4 patients after 6 months showed complete epithelialization of vaginal mucosa [8].

In our case, the use of laparoscopic dissection of vesico-rectal space was beneficial as it was performed under vision and allowed excision of fibrous tract formed as a result of previous vaginoplasty.

At present there are many techniques for creation of neovagina but there is no consensus yet regarding the ideal method and the ideal graft. The surgical technique should be based on the experience of the surgeon and the procedure undertaken only after appropriate counselling in motivated females who are either sexually active or planning marriage in near future.

Conclusion

MRKH syndrome is a rare disorder characterized by primary amenorrhoea and treatment needs to be directed towards creation of neovagina which can facilitate sexual function for the female. Though there is a whole armamentarium of choices available, the ideal method is not yet defined. Patients who are planning marriage are ideal candidates for intervention. They should be counselled and informed about all available treatment options. Vaginoplasty using amnion graft is a safe, easily available, cost effective, and offers physiological epithelialization of vaginal canal. But regular follow up and dilator therapy or sexual intercourse is essential for maintaining patency of vaginal passage.

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Volume 2 Issue 6 June 2021
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