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Vaginoplasty with Amnion Graft: Management of Mayer-Rokitansky-Kuster-Hauser Syndrome

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ABSTRACT

Mayer-Rokitansky-Kuster-Hauser syndrome also known as mullerian agenesis is a rare congenital condition in which there is absence of uterus along with upper vagina. Patient usually presents with primary amenorrhea with or without cyclical lower abdominal pain but have normal secondary sexual characters. Modified McIndoe Vaginoplasty with amnion graft is the commonest surgery performed worldwide. A 23 year old girl with normal secondary sexual characters presented with primary amenorrhea with cyclical lower abdominal pain; on examination blind vagina was present. Vaginoplasty with amnion graft was done and vaginal mould was placed. Vaginal dilatation with Hegar's dilator was done weekly until 6 weeks. She is under regular follow-up at present and advised for regular manual dilation at home. McIndoe Vaginoplasty with amnion graft is a simple yet rewarding procedure especially in low resource countries like ours, with good success rate and with minimal postoperative complications.

Keywords: Amnion graft; Mayer-Rokitansky-Kuster-Hauser Syndrome; Modified McIndoe Vaginoplasty; Primary amenorrhea; Secondary sexual characters.

INTRODUCTION

Mayer-Rokitansky-Kuster-Hauser (MRKH), also referred to as müllerian agenesis, müllerian aplasia or vaginal agenesis is a rare condition with an incidence of 1 per 4,500-5,000 females.[1] There is embryonic underdevelopment of mullerian duct leading to agenesis of uterus and upper vagina with normally functioning ovaries. It is associated with anatomical as well as psychological and social problems like painful sexual intercourse, primary amenorrhea and infertility and surgery aiming at reconstruction of vagina of adequate length and width to improve the sexual life is the main method of treatment.[2] Among the various procedures, Modified McIndoe Vaginoplasty with amnion graft is simple and rewarding procedure with minimal complications.[3]

CASE REPORT

A 23 years unmarried female, referred case from Bardiya, Nepal, came to our OPD with the complaint of cyclical lower abdominal pain for 6 years, pain was crampy in nature lasting for 2 days with no aggravating factors and relieved with analgesics. However, she had not attained menarche till date. Her past medical and surgical history were unremarkable. Her mother's obstetric history was also unremarkable.

On examination, her general condition was fair with stable vitals. Systemic examinations were normal. Her breasts were well developed at Tanner stage V. Axillary hair was normal. Gynecological examination revealed pubic hair at Tanner stage V and normal labia majora and labia minora. However, vagina was blind ended

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with length of around 2 cm. Uterus size could not be appreciated. Her renal and skeletal examinations were normal.

All her blood investigations were within normal limits. Her ultrasonography of abdomen and pelvis showed small sized anteverted uterus of about 3.1x2x1.2 cm with regular outline with rest of the scan normal. Her MRI scan of pelvis also showed hypoplastic uterus of size 3.7x2.2x1.1 cm with loss of endometrium and myometrium differentiation with rest of the visualized pelvic structures normal.

With all these findings the diagnosis of Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome was made and she was planned for Vaginoplasty with amnion graft.

On the day of surgery, following elective caesarean section, amnion was obtained after separating it from placenta; after getting consent from the donor patient. The separated amnion was washed thoroughly with sterile normal saline and kept in saline solution till further use. A 9 cm long mould was made from plaster of Paris rolled over a plastic rod which was then covered with two sterile condoms. The patient was kept in lithotomy position. After painting and draping, Foley's catheterization was done. A transverse incision was made over hymen and a potential avascular space of 7 to 8 cm was created in between urinary bladder and urethra above and rectum below with gentle digital manipulation. The amnion was mounted over the mould and its ends sutured so that amnion covered all parts of the mould. The amnion mounted mould was then inserted into the newly created vaginal canal gently so that amnion did not get damaged. Bilateral labia majora then sutured to hold the mould in place and to prevent expulsion. Foley's catheter was kept in-situ.

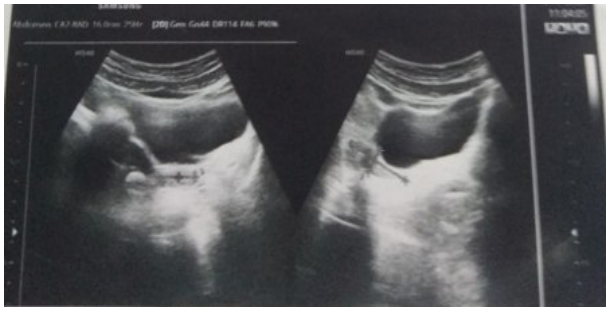

Figure 1. Ultrasonography showing hypoplastic uterus.

Figure 2. MRI showing hypoplastic uterus with normal adnexa.

Post operatively patient was kept on antibiotics. Mould and the Foley's catheter were removed on day 11 of surgery. There was 7 cm of vaginal canal created with no signs of infection. The amnion lining had been well taken up. The cavity was gently cleaned. Patient was discharged with the advice to follow up after one week. Patient was also counseled about manual dilatation of vagina regularly at home. After one week of discharge, she was called for evaluation under anesthesia and dilatation done with Hegar's dilator. Vaginal length of 7 cm was noted. She was then advised for regular manual dilatation at home and regular visit to hospital for evaluation.

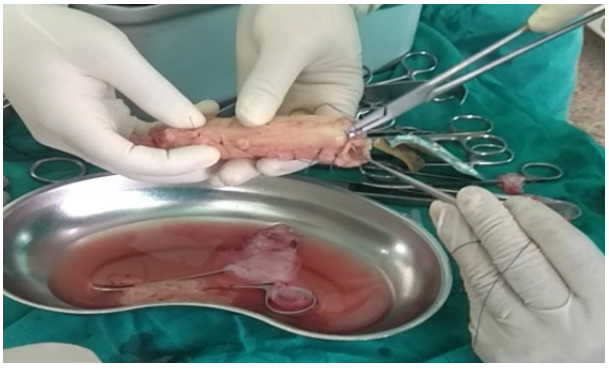

Figure 3. Preparing amnion graft mould.

DISCUSSION

MRKH is the second most common cause of primary amenorrhea but most common cause of vaginal agenesis. Most of the patient have absent upper vagina, cervix and rudimentary uterus which are nonfunctional while 7-10% may have functional endomentrium.[3] MRKH syndrome is generally divided into two subtypes: MRKH type 1, in which only the upper vagina, cervix and the uterus are affected, and MRKH type 2, which is associated with additional malformations generally affecting the renal and skeletal systems, and also includes MURCS (Mullerian Renal Cervical Somite) characterized by cervico-thoracic defects. MRKH syndrome is mainly sporadic; however, familial cases have been described indicating that, at least in a subset of patients, MRKH may be an inherited disorder.[1]

Our case fits in Type I MRKH syndrome with sporadic onset. Clinical picture showed a girl with primary amenorrhea, normal secondary sexual characteristics with no androgen excess and with hypoplastic uterus and blind vagina. Diagnostic imaging includes USG and MRI, where MRI is gold standard. The treatment consist of creating a neovagina to improve sexual function and it must be offered to patients only when they are ready to start sexual activity and can do manual dilatation as counseled. Our patient, on the threshold of marriage was an ideal candidate for this surgery.

The McIndoe technique was first described in 1938 by Bainster and McIndoe. Despite the existence of several alternative methods, there is still no consensus regarding the best option for surgical correction.[4] The original surgery used split skin or full thickness skin grafts to line the newly created vagina. Modified McIndoe vaginoplasty procedure uses various alternatives like amnion, peritoneum, labia minora grafting, or synthetic materials; among which amnion graft is most commonly used since it is very cheap, easily available, its anti-inflammatory and anti-scarring properties helps in regenerative and healing processes[5] and it lacks any antigenicity like HLA-A, B, C, and DR antigens on their surfaces hence acute immune rejection does not occur after the transplantation of human amniotic epithelial cells.[6]

A study showed more than 96% success rate of Modified McIndoe Vaginoplasty using amnion graft done among 28 patients.[7] In a case series of Modified McIndoe Vaginoplasty using amnion graft, some observed more than 92.8% structural correction and 85% functional correction at the end of 1 year follow up.[8] Another study showed satisfactory postoperative results including mean vaginal length of 8.4 cm in 82% and satisfactory sexual function in 56% patients.[9]

Postoperative dilation is essential to prevent significant neovaginal stenosis and contracture; therefore, these techniques are not recommended if the patient objects to dilation. Dilators must intermittently be used until the patient engages in regular and frequent sexual intercourse.

The other issue of concern is 'Fertility and Childbearing'. Fertility using surrogacy and assisted reproductive techniques may be an option for these patients. Female offspring of women conceived by assisted reproductive techniques have normal reproductive tracts.[10]

CONCLUSIONS

Although new techniques of vaginoplasty have evolved over the years, using various approach such as laparoscopy, laparotomy and by using various materials as graft such as skin, amnion, peritoneum, labia minora; Modified McIndoe vaginoplasty with amnion graft is still a safe and effective procedure with minimal post-operative complications and chance of rejection. The technique is simple and safe and provides functional vagina for satisfactory sexual function in majority of the patients. However, proper mould use after surgery and regular dilatation of neovagina remain the cornerstone of the treatment.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

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