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Case Series

Sigmoid Colonovaginoplasty – Case Series at the National Obstetric Fistula Center Katsina

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ABSTRACT

Mullerian duct abnormalities are rare and the exact incidence is unknown, patients may present with absence of the uterus, fallopian tubes, cervix and upper two third of the vagina, ovaries are usually present and functional. We treated 3 patients from 2020 to 2023, the average age was 23 years both presented with primary Amenorrhea, with well-developed secondary sexual characteristics, no cyclical abdominal pain or features of hyperandrogenism, and on examination they had normal external genitalia with short vagina or only a dimple. Radiological findings showed features of MRKH (present ovaries, absent or hypoplastic uterus, fallopian tubes and cervix). No renal abnormalities. A definitive repair with creation of a neovagina using the sigmoid colon segment was performed in a single stage which offers a promising and safe alternative for patients with MRKH.

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INTRODUCTION

The absence of an adequate capacity or vaginal agenesis/aplasia is of great concern to a woman especially those of reproductive age group, commonly due to absent menses, failed or difficult coitarche and infertility.

Mullerian duct anomalies are rare and can present with abnormalities in the upper vagina, uterus and fallopian tubes.[1, 2] The exact incidence of vaginal agenesis is unknown.[1] some literatures put the incidence at 1 in 4000 to 1 in 10,000 female life births.[3,4]

The vagina is derived from interaction between the uterovaginal primodium (mesoderm) and the pelvic part of the urogenital sinus (endoderm). Mullerian abnormalities are often associated with abnormalities of the renal and axial skeletal systems and they are often characterized with functioning ovaries and age-appropriate external genital.[5] Mayer Rokitansky Kuster Hauser

Syndrome (MRKH) is characterized by congenital aplasia of the uterus and upper two thirds of the vagina, with a normal female secondary sexual characteristic and a normal karyotype (46XX).[6]

There are two types of this syndrome, Type 1 describes the agenesis of only the Mullerian derivatives (uterovaginal agenesis) while Type 2 are malformations associated with upper urinary tract, the skeleton and the ontological and cardiac spheres.[7]

There are several methods of treatment ranging from non-surgical (conservative) to surgical reconstruction techniques for vaginal aplasia, surgical methods are either conventional or laparoscopically. None of the techniques has proven to be superior to the others, and no consensus on an ideal intervention.[8]

CASE REPORTS

CASE 1: A 20-year-old student, Para $0+^0$ who presented with primary amenorrhea and a six month history of inability to perform coital activity with her husband having being married for 6 months.

She noticed her peers had attained menarche about 6 years ago while she was still yet to menstruate though she has developed breast, pubic and axillary hair, no associated cyclical pain, nipple discharge, excessive heat or cold intolerance, no abnormal hair growth.

On examination, she was healthy looking with no pallor, jaundice no hirsutism or acne, her vital signs were normal. She had normal axillary and pubic hair with well developed breast & nipples (Tanner Stage V). Her abdomen was flat with no abdominal mass felt while pelvic examination shows normal external genitalia (labia majora, labia minora, clitoris) external urethral orifice was normal in position, a vaginal dimple seen about 2cm deep. On per rectal examination the vaginal tube and the uterus were not felt while the anal opening was normal.

She was diagnosed as a case of primary amenorrhea due to MRKH. She had abdomino-pelvic ultrasound scan which visualized both kidneys. Right ovary was normal in size, shape and position while the uterus and vagina were not visualized, her E/U/Cr were all within normal limits, PCV was 37%, FBS - 4.6mmol/L. Serology's were all negative.

Karyotyping, IVU, diagnostic laparoscopy was not done due to non-availability of the tests in our facility and financial constraints.

She was counselled and prepared for sigmoid–colonovaginoplasty, she gave consent had bowel preparation for three days before the procedure.

Procedure

Patient was cleaned and draped under general anesthesia initially in a supine position, a Pfannestiel incision was made and deepened through the rectus abdominis muscle to reach the peritoneal cavity. Examination reveals both ovaries present, rudimentary uterus, small dimple around the anatomical site for vaginal orifice.

The sigmoid loop was identified and mobilized along its length and vascularity with its mesentery (inferior sigmoid artery) and about 12cm of the sigmoid colon was isolated by sectioning the upper and lower ends and cleaned with saline, colo-colic (end to end) anastomosis was done to restore the sigmoid colon to the rectum.

The patient was positioned in exaggerated lithotomy position, with the help of an assistant. The Hagar dilator was pushed through the vaginal remnant to open up into the abdominal cavity and the lower part of the resected segment anastomosed with the vaginal remnant and the other end closed to make it a blind ended tube thereby forming a new vagina. The abdominal incision closed in layers and pressure dressing applied, urethral catheter inserted and Dildo dilator (Vaginal mold) left in situ.

She was on nasogastric tube, IVF and antibiotics, NPO for 5 days following surgery, her only complain was excessive vaginal discharge, and she was counselled on antibiotics and perineal hygiene, to change Dildo with lubricant regularly. She did well and was discharged after 18 days, she commenced coitus at 6 weeks post-surgery, vaginal depth was about 12cm, and capacity was 4cm. she was followed up for 3/12 and had done well.

CASE 2: A 25-year-old housewife, Para $0+^0$, who presented with primary amenorrhoea, inability to achieve penetrative intercourse having being married for 2 years, no associated cyclical abdominal pain, no urinary symptoms, while secondary sexual characteristics were said to be developed normally.

On examination, she was not pale, afebrile, anicteric, a cyanosed, her breast and pubic hair were Tanner Stage IV, abdominal examination reveals no mass palpable, pelvic examination revealed normal pelvic hair pattern, normal external genitalia with a blind ending vagina (2cm). An assessment of primary amenorrhoea secondary to MRKH Rule out Androgen insensitivity syndrome, her FBS was 4.1mmol/L, E/U/Cr were within normal limits. Serology were negative, urinalysis was negative of protein and glucose, abdomino pelvic ultra sound scan showed hypoplastic uterus, both ovaries and kidneys were normal in size & position.

She was asked to group and cross match 2 units of blood and commenced on bowel preparation following counselling on her condition and consent for surgery obtained. She eventually had sigmoid colonovaginoplasty (Procedure described above) with the following intraoperative findings – absent uterus, upper 2/3 of the vagina, both ovaries and fallopian tubes were present.

She had IVF, NGT, antibiotics, analgesics, subcutaneous enoxaprin for 3 days, 2pints of blood were transfused, she commenced graded oral sips a week later bowel sounds were present and normoactive, she was however discharged 2 weeks post-surgery, she was seen in the clinic 6 weeks post-operative, the site had healed and was counselled to continue the use of Dildo, at 8^{th} week the vulva and vagina were normal. Vaginal capacity

was 5cm, and vaginal depth–was16cm.–She resumed coitus and was doing well.

CASE 3: A 25-year-old, single NCE graduate who presented with complaints of amenorrhea, no cyclical abdominal pain, no excessive hair growth, discharge from the nipples.

On examination, she was not pale, afebrile, anicteric, no hirsutism or acne. Breast development was at Tanner stage V. Abdominal examination reveals no pelvic mass, vaginal examination reveals normal labia, vaginal dimple, no orifice, on digital rectal examination reveals normal anal orifice and uterus could not be palpable.

Her investigation results were as follows, PCV 34%, E/U/Cr all parameters were with in normal limits and serology were all non-reactive, a karyotype which was 46XX. Abdominopelvic ultrasound scan reveals a hypoplastic uterus, she was counselled and consented to surgery which she had-sigmoid colonovaginoplasty (procedure described above).

She had wound breakdown and managed with antibiotics and daily wound dressing, she eventually had secondary wound closure and was discharged one week later. She presented for follow-up 2 weeks later and was doing well, vaginal capacity was 4cm and depth was 10cm long and complaint was that of vaginal discharge. She was counselled on personal hygiene and was to continue the use of Dildo.

She presented 8 weeks after her last visit she was doing better. Vaginal discharge has reduced significantly, she was asked to continue the use of Dildo as she was still unmarried.

DISCUSSION

Management of vaginal agenesis due to MRKH is somewhat tasking especially in a low resource setting like ours. In spite all the challenges is pertinent to institute the appropriate treatment which takes into account both the associated abnormalities and the psychological components.[6-9]

The definitive management of this group of patients will depend on the age, emotional and sexual needs, [10] for effective treatment, the approach must be multi-disciplinary involving the surgeon, the gynecologists, urologist, plastic surgeon, psychologist depending on the type of neovaginoplasty done in the center.

Both patients treated in our facility presented with similar complains of primary amenorrhoea having developed secondary sexual characteristics, 2 of them were married and the third was still single, the married started having problems with penetrative sexual intercourse, this was because of absent or poorly developed vagina. However, other associated abnormalities like urinary symptoms and musculoskeletal problems were absent though we are usually limited due to financial constraints and investigative modalities in our center. This also explains why karyotype or buccal smears were not done in the first 2 patients only the third could afford it.

There were no classical features of undescended testes and hypospadias.[11] On examination all had normal stature with well developed breast, pubic, axillary hair, no features of hyperandrogenism, abdominopelvic ultrasound revealed absent and hypoplastic uterus respectively. Hence, the diagnosis of vaginal mullerian agenesis.

All of them were counselled on the options of treatment and the possible side effects of both options of non-surgical methods and surgical methods and all patients settled for surgical method of neovaginoplasty.

Sigmoid colonovaginoplasty was offered to them being the method feasible in the center and the competence, the first and third patients had episodes of vaginal discharge but with reassurance and good perineal hygiene it reduced significantly. Other surgical methods are McIndoe procedure, Williams Vulvo vaginoplasty, Vecchetti procedure and Davydov procedures.[12]

Sigmoid vaginoplasty has the following potential disadvantages, chronic vaginal discharge, foul odor, stenosis at the anastomotic site and the risk of developing adenocarcinoma in the graft.[12]

Following surgery, they all had Dildo to keep the reconstructed vagina patent and regularly changed for about 6 weeks until healing is completed and the 2 first patients were advised to commence sexual intercourse as soon as healing was achieved and neovagina allows it without discomfort.[13]

Fertility issues will still remain a big challenge because above surgical approach only correct the vaginal defect, though ovaries are functionally normal, invitro fertilization and surrogacy still are the available options using the woman own eggs, there are trials on uterine implantation currently that may hold hope for these women in the future, this has been successful in animal models.[14]

CONCLUSION

Sigmoid colonovaginoplasty has been a treatment of choice in our center for management of patient with vaginal hypoplasia or agenesis in our facility with satisfactory outcome and quick return to penetrative sexual relationship despite the complications, patients still do well and improved significantly. Other complications in some studies of possibility of adenocarcinoma will necessitate following up the patients for life.

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