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Case Report

Laparoscopic vaginoplasty with cervical neo vaginal anastomosis in a case of complete vaginal agenesis in a young girl

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ABSTRACT

Complete vaginal agenesis is an extremely uncommon mullerian abnormality. In this case report, we described a case of a female patient, age 14, a known case of complete vaginal agenesis presented with severe cyclical dysmenorrhea. MRI suggested no vagina with a normal uterus and cervix with hematometra. She underwent laparoscopic vaginoplasty with cervico-neovaginal anastomosis. After surgery, the patient experienced regular menstrual cycles without dysmenorrhea. On follow-up, hysteroscopy showed a patent anastomosis, and she continued to have regular menstrual cycles.

Keywords: Complete vaginal agenesis, Laparoscopy, Menstrual cycle, Cervico-neovaginal anastomosis

INTRODUCTION

In adults, the Mullerian duct forms the uterus, cervix, fallopian tube, and upper 1/3rd part of the vagina. Anomaly in the Mullerian duct is found in about 4.3% of the general population.¹ In 2013, the European Society of Human Reproduction and Embryology (ESHRE) proposed a newer classification system for Mullerian anomalies based on uterine anatomy. In addition, anomalies of the cervix and vagina are classified separately. In the ESHRE classification, cervical and vaginal agenesis are classified as C4 and V4, respectively.^{2,3}

Complete vaginal agenesis is a rare congenital Mullerian Anomaly, reported in 29% of cases. Its association with cervical agenesis is also rare, reported in 39% of cases of cervical agenesis. Primary amenorrhea and/or cyclical abdominal discomfort are typical symptoms of cervicovaginal agenesis that first appear around menarche.⁴ In these situations, the endometrium continues to function normally, but there is no way for the menstrual blood to leave the vagina, which causes hematocolpos and

then hematometra to form. If left untreated, the illness may worsen and develop hematosalpinx and endometriosis. Due of its excellent accuracy and thorough delineation of uterovaginal anatomy, MRI is the preferred test for evaluating Mullerian duct anomalies.³ Extensive endometriosis, which in severe cases damages reproductive capacity irreparably and demands salpingectomy and hysterectomy, may develop as a result of delayed diagnosis and treatment. Modern assisted-reproduction facilities and improvements in laparoscopic methods make conservative management both possible and advised.⁵ These methods include the creation of a neovagina and reconstruction of the cervix around various stents. Herein is presented a case of complete vaginal agenesis managed by laparoscopic vaginoplasty along with cervico-neovaginal anastomosis with a copper T placement afterwards.

CASE REPORT

A 14-year-old girl presented at the OPD with a history of abdominal pain worsening day by day and did not improve with analgesics. Five years ago, she had history of cyclical

abdominal pain and was diagnosed with complete vaginal agenesis. She underwent one surgery for vaginal correction to drain haemometra, which later did not relieve her symptoms.



Figure 1: MRI showing normal uterus and cervix with absent vagina.



Figure 2: Creation of neovagina by using Hegar's dilators of different sizes.



Figure 3: Drainage of the collected blood/hematometra.

Important findings of the complete-vaginal agenesis case are as follows. The age of the patient was 14 years. A history of one surgery for vaginal correction was present. MRI findings showed complete vaginal agenesis (Figure 1). The physical examination showed a normal secondary sexual profile, a soft abdomen without any organomegaly. The surgery was laparoscopic vaginoplasty, cervical-

neovaginal anastomosis from the anterior and posterior ends of the cervix at the 3 o'clock, 6 o'clock, 9 o'clock, and 12 o'clock positions.



Figure 4: Placement of the silicon catheter through the introitus to the uterine fundus.



Figure 5: Cervical-neovaginal anastomosis.

Follow-up appointments at 1, 3, and 6 months after surgery. The patient's menstrual cycle was regular, and 10 months later, she had no symptoms.

The first surgery was performed 2 years ago, where the vaginal incision and catheter placement for hematometra drainage was done but later turned out to be unsuccessful and lead to urethral injury. Since the patient did not have menses at the age of 12 years unlike her other friends and increasing cyclical abdominal pain, made her parents more worried about her condition. She doesn't have any significant personal, past, or family medical history, and there was no diethylstilbesterol exposure during pregnancy. A general physical examination revealed normal secondary sexual profile (breast-Tanner stage 4, pubic hair-Tanner stage 4) as well as an average build. Breast and thyroid examinations were normal. An examination of the abdomen revealed a soft abdomen without any organomegaly, although deep palpation revealed discomfort in the lower abdomen. Pelvic magnetic resonance imaging (MRI) revealed a normal-sized uterus measuring 7 cm by 6 cm by 5 cm. A normal cervix was visible with haematometra but the vagina was completely absent. The size, position, and appearance of

both ovaries were all normal. Complete vaginal agenesis was tentatively diagnosed based on these imaging results.



Figure 6: Hysteroscopic placement of copper T after the surgery.

After receiving informed consent, the patient was taken up for the surgery. Hegar's dilators of different sizes 1 to 14 with the help of lignocaine jelly, along the axis of the pelvis were used to create the neovagina. Frequent per rectal examinations were done (Figure 2). Plane between the bladder and rectum was created such that after gradual dilatation, we were left with two barrel-shaped cylinder-like structure with a septum in between, which we cut with diathermy pencil/scissors. Then fingers are introduced to enlarge the space and speculums pushing the bladder up and rectum down. Finally, a 5 cm vagina was created. Laparoscopically we entered the abdomen and dissected the bladder and rectum as much as possible. A portion of the cervix was found swollen because of the collected hematometra. Fascia of Denonviller was opened. Diluted vasopressin was injected into the isthmus region of the uterus the bulge over the cervix was incised and collected blood was seen coming out (Figure 3). The atretic portion of the vagina was cut using the help of the assistant surgeon sitting at the vaginal end. A silicone catheter was placed through the introitus and taken out through the fundus, later stitched with vicryl 1-0 sutures (Figure 4). Cervico-neovaginal anastomosis was done with 1-0 PDS (Figure 5). Peritoneal closure was done later, to form a protective layer over the anastomotic site. The catheter to be kept from 6 weeks to 3 months. Copper T placement was done afterwards at the end of the surgery (Figure 6).

The patient was kept nil orally for 24 hrs and then given a liquid diet for 1 day. She was allowed solid food on the

third day following the procedure. The patient was told to utilize a soft mould for the following 15 days before switching to a hard mould. Intraoperative and postoperative complications were not observed. She has now undergone three follow-up appointments at 1, 3, and 6 months after surgery. After one month, hysteroscopy revealed bilateral ostia and a normal, patent uterine cavity. After surgery, the patient's menstrual cycle was regular, and 10 months later, she had no symptoms.

DISCUSSION

The female genital tract can develop complete vaginal agenesis with a functioning uterus, which is a rare disease. Only a few cases were reported in the literature. Mullerian duct and urogenital sinus union results in normal vaginal development. The Mullerian tubercle makes up the upper two-thirds of the vagina and the urogenital sinus the lower third.^{6,7} Partial or full Mullerian agenesis are both possible. While complete mullerian agenesis (MRKH syndrome) is the most frequent type seen, partial mullerian agenesis is unusual and is characterized by a normal uterus and a small vaginal pouch distal to the cervix. MRKH syndrome is most frequently linked to vaginal agenesis.⁸

Primary amenorrhea, cyclical abdominal pain, and hematometra are the most typical symptoms. In certain situations, retrograde menstruation may cause the development of endometriosis. Chronic pelvic pain, palpable mass per abdomen, and urinary complaints are also encountered. Differential diagnosis includes imperforate hymen, vaginal agenesis, or transverse vaginal septum, which should be excluded before definitive management.^{9,10} Although clinical history and examination are crucial in such cases, pelvic imaging should be considered to know the exact anatomical anomaly. MRI is considered the gold standard for diagnosis as well as to look for associated renal tract anomalies. In our case, imaging was really helpful as it suggested a diagnosis of complete vaginal agenesis and hematometra. With improvements in surgical techniques and the development of laparoscopy, a minimally invasive method might be the best course of action.¹¹

Deffarges et al analysis of 18 women showed reduced dysmenorrhea, normal menstruation, and no postoperative endocervical canal occlusion. Only one patient out of 18 required additional canalization surgeries due to recurrent uterovaginal canal stenosis, resulting to salpingo-oophorectomy for pyosalpinx. Ten of the 18 patients (56%) experienced spontaneous pregnancies and delivered at term (36-38 weeks) via elective cesarean section. With this method, the likelihood of bladder and rectum injury is decreased, and the uterovaginal canal is reliably canalised.¹²

To prevent vascular or nerve damage, cervico-neovaginal anastomosis requires a detailed anatomic understanding of the retroperitoneum. There have been reports that the stenting lasts anywhere from three weeks and three

months. In our case, the patient is currently free of the symptoms of her menstrual cycle. After 6 weeks, her stent (a Foleys catheter) spontaneously discharged itself, and 9 months after the treatment, she is symptom-free. The possibility of a spontaneous pregnancy as a result of canalization seems very low. Associated endometriosis, pelvic adhesions, and a lack of normal endocervical canal glandular function may all contribute to this low fertility rate.^{13,14} For cases of infertility, assisted reproductive technologies may be considered. It is advised to deliver through cesarean section in all pregnancies.¹⁵

CONCLUSION

When a young girl has primary amenorrhea and cyclical abdominal pain, it's important to consider complete vaginal agenesis. Early diagnosis and treatment are crucial for a successful outcome. Accurate imaging of the pelvic anatomy is required to avoid complications. Cervico-neovaginal anastomosis is preferred with silicone catheter placement to reduce the likelihood of secondary vaginal stenosis. Early surgery is recommended to prevent complications like endometriosis. Regular menses have a positive psychological impact on these young women.

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