Uterine Conserving Surgery in Patient With Cervicovaginal Agenesis: A Case Report

Maryam Deldar Pesikhani1, Zinat Ghanbari2, Seddigheh Borna1, Leila Pourali2, Hossein Chegini3, Zahra Lotfi1

1 Department of Obstetrics and Gynecology, School of Medicine, Tehran University of Medical Sciences, Tehran, Iran
2 Department of Obstetrics and Gynecology, School of Medicine, Mashhad University of Medical Sciences, Mashhad, Iran
3 School of Medicine, Tehran University of Medical Sciences, Tehran, Iran

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Abstract- Congenital absence of the vagina with variable uterine development known as Mullerian agenesis or Mayer-Rokitansky-Kuster-Hauser syndrome. Cervicovaginal agenesis in the presence of normal uterus is very rare. Conservative surgery has recently been suggested in patients with congenital cervicovaginal atresia in order to relieve the symptoms and maintain fertility. A 13-year-old female referred to a pelvic floor clinic, because of primary amenorrhea and severe cyclic pelvic pain. Ultrasonography revealed a large amount of blood accumulation in the uterine cavity, and also, the cervicovaginal agenesis was reported. Both ovaries were normal. A neovagina was created by dissection of the space between the bladder and rectum. Under ultrasonography guidance, two Pezzer catheters were inserted between uterine ending and neovagina, so the catheters kept the canal patent, a soft mould was applied to prevent the vaginal stricture. Cervicovaginal agenesis, accompanied by normal functional endometrium, is a rare but challenging Mullerian anomaly in the case of surgical treatment. One of the successful conservative treatment in a fully educated patient is the vaginal reconstruction and uterovaginal anastomosis by stenting and continues the application of vaginal mould.

Keywords: Vaginal agenesis; Conservative treatment; Pelvic pain; Amenorrhea

Introduction

Vaginal agenesis, also known as Mullerian agenesis or Mayer-Rokitansky-Kuster-Hauser syndrome (MRKH), refers to the congenital absence of the vagina with variable uterine development (1). The prevalence of vaginal agenesis is 1 in 5000 live female births (2). It is usually accompanied by cervical and uterine agenesis; however, 2-7 percent of these women have functional endometrium (3). Cervicovaginal agenesis in the presence of normal uterus is very rare and may present with cyclic abdominopelvic pain secondary to hematocolpus, hematometra, hematosalpinx, or endometriosis (4). Because of these complications, the reproductive capacity is low, and total abdominal hysterectomy is proposed as a therapeutic choice for many years (5). Despite this approach, conservative surgery has recently been suggested in patients with congenital cervicovaginal atresia in order to relieve the symptoms of retrograde menstruation and maintain the fertility capacity (6) although the conservative strategy maybe leads to severe complications (7).

The aim of this study was to report the successful conservative surgery in a rare case of cervicovaginal agenesis with normal uterus.

Case Report

A 13-year-old female adolescent referred to the pelvic floor clinic of an academic hospital, Tehran University of Medical Sciences, Tehran, Iran, because of primary amenorrhea and cyclic severe pelvic pain since four months before the visit. Initial physical examination showed normal external genitalia and normal secondary sexual trait. Ultrasonography revealed a large amount of blood accumulation in the uterine cavity about 16x10 cm in size, the cervicovaginal agenesis was reported. Both ovaries were normal. The left kidney was normal, but the right one was not seen (right kidney agenesis). Magnetic resonance imaging (MRI) showed blood
products distending the endometrial cavity and cervical canal. Although the lower vaginal canal seemed atretic (Figure 1,2). Rectoabdominal examination under general anesthesia and concurrent transabdominal sonography revealed complete vaginal agenesis and a likely atretic cervix with a huge abdominopelvic mass of about 20 cm in size. A neovagina was created by dissection of the space between the bladder and rectum. A large amount of dark sticky blood was drained at this time (about 1000 ml). After irrigation of the new vaginal cavity with warm saline, the normal cervical structure couldn’t be identified during operation, so under ultrasonography guidance, after the uterus regains normal size and shape, we inserted two Pezzer catheters (size 22) from the opening pathway between uterine ending and neovagina, so the catheters kept the canal patent. No graft was used, but a soft mold was applied to prevent the vaginal stricture. A prophylactic antibiotic was administered for seven days. One week after surgery, the patient returned to the operating room to evaluate the vaginal cavity, so the mould removed and after vaginal irrigation with warm saline, the new mould inserted and both Pezzer catheters which kept the uterine end, open; retained in place for several months, ultrasonography confirmed the normal location of catheters (Figure 3). Before hospital discharge, the patient was trained to use the mould regularly until to have regular intercourse. After a nine-month regular follow up, she experienced normal mensural cycles without any complication. Informed consent was obtained from the patient to publish information and related images.

**Figure 1.** MRI (magnetic resonance imaging) in coronal view showed blood products distending endometrial cavity and cervical canal

**Figure 2.** MRI in the sagittal view showed a large amount of blood in the uterus, and also lower vaginal agenesis was noted

**Figure 3.** Ultrasonography showed catheters in normal position between the uterus and vagina

**Discussion**

Vaginal agenesis is most commonly associated with MRKH syndrome and characterized by normal female karyotype, phenotype, and normal endocrine status, the ovaries and secondary sexual characteristics develop normally (1). The accompanied anomalies, which usually consist of renal and skeletal malformations, are reported 40% and 20%, respectively (1). The current case had a normal female genotype and phenotype. The ultrasound also showed unilateral agenesis of the kidney, but the MRI revealed no skeletal abnormalities.

Previously, the recommended treatment of cervicovaginal agenesis with a normal uterus was hysterectomy because of the high failure rate of uterovaginal anastomosis and also the risk of severe pelvic infection and sepsis (7).

Rebecca et al., reported two cases of cervicovaginal agenesis with the normal uterus, which managed them with a conservative surgical approach. They created neovagina and connected it to the uterine cavity, but after six months, there was no passage between the uterus and neovagina. Despite recurrent reconstructive surgeries, the patient presented with pelvic infection and sepsis, which the hysterectomy was mandatory (7). They also reported the second similar case with the same approach; two months after conservative surgery, the patient showed vaginal canal stenosis, and hematometra was noted, so definite surgery planned, but because of septic shock, she developed renal failure, respiratory failure, and coagulopathy (7).

Considering these harmful consequences of uterine conserving surgery, in the case of cervicovaginal agenesis, there are some important issues for both the patient and surgeon before choosing the best surgical decision: first, the patient and her parents should be completely informed about regular follow-up visits, appropriate hygiene, regular use of vaginal dilators after surgery. Second, in conservative surgery after the creation of the vagina in the case of cervical agenesis or cervical malformation, insertion of the stent between the lower uterine segment and neovagina may prevent
cervical stenosis. The exact duration of remaining the stent in place is not obvious, but some literature recommends at least six months to allow complete epithelialization around the catheter (8).

Cervicovaginal agenesis, accompanied by normal functional endometrium, is a rare but challenging Mullerian anomaly in the case of surgical treatment. One of the successful conservative treatment in a fully educated patient is the vaginal reconstruction and uterovaginal anastomosis by stenting and continues application of vaginal mould.

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