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High Transverse Vaginal Septum with Vaginal and Cervical Agenesis in a 13-Year-Old Female: A Case Report

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ABSTRACT

Background & aim: A vaginal septum is a condition in which the wall of the tissue dividing vagina does not resolve completely. This complication can be transverse and longitudinal. Although the occurrence of a transverse vaginal septum is 2 per 100,000 births, the exact etiology of this anomaly remains still unidentified. In this study, a case of high transverse vaginal septum with vaginal and cervical agenesis is reported.

Case report: A 13-year-old single and virgin girl presented with cyclic abdominal pain. On examination and magnetic resonance imaging, vaginal and cervical agenesis was diagnosed. Surgical therapy included an incision in the lower part of the uterus at the site of the bulging and the use of a hysterometer passing down the uterus. The cervix was not touched, cervical agenesis was diagnosed, and the patient underwent abdominal hysterectomy and bilateral tubal salpingectomy. The pathological results confirmed the cervical agenesis.

Conclusion: It is recommended that in patients with high transverse vaginal septum, hysterectomy be performed for the treatment of cervical and vaginal agenesis. However, it may not be a suitable treatment choice for some cases, and it is better to decide based on the patient's condition.

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Introduction

A vaginal septum is a condition in which the wall of the tissue dividing vagina does not resolve completely. This complication can be transverse and longitudinal. In some women, the vaginal septum is partial or does not run along the length and width of the vagina (1). The embryologic etiology of transverse vaginal septum is the unsuccessful absorption of the tissue between the caudal end of the fused Müllerian ducts and the vaginal plate. Although the occurrence of a transverse vaginal septum is 2 per 100,000 births, the exact etiology of this anomaly remains still unidentified (2). Numerous girls do not recognize that they have a vaginal septum until they reach puberty. The signs of this condition can be pain, uneasiness, or irregular menstruation. Sometimes, girls do not know this until they become sexually active and experience pain during intercourse. However, some women with a vaginal septum do not have any symptoms (1). Cervical agenesis is a complex female genital malformation including the failure of resorption and organogenesis. Some patients with cervical

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abnormalities will have simultaneous vaginal agenesis, mainly those with cervical agenesis, and one-third have a uterine abnormality (3). Patients with cervical agenesis characteristically present in early adolescence, around the time of menarche, with amenorrhea and cyclic pelvic pain produced by the obstruction of menstrual flow from the uterus. Cervical agenesis occurs during fetal development, and happens in 1 per 80,000 women that is related to the abnormality of the vagina (2).

In this study, a case of 13-year-old female with a high vaginal septum along with vaginal and cervical agenesis is reported.

Case Report

A 13-year-old female (Single/Virgin) was referred to the Female Pelvic Floor Medicine and Surgery Department of Tehran University of Medical Sciences, Tehran, Iran, for primary amenorrhea and abdominal pain that was cyclic for 3 to 4 days each time and started 6 month ago. Ultrasound image showed a distended uterus with a clot inside, clotted fluid with low-level internal echoes, hematometra (uterine hematometra is collection or retention of blood in the uterus most commonly due to an imperforate hymen or transverse vaginal septum) and hematocolpos (Hematocolpos is the vaginal retention of menstrual blood at puberty). In most cases, this condition causes painful amenorrhea, and rarely pelvic mass syndrome), and a brief free fluid in the cul-de-sac. The karyotype of the patient (46, xx), as well as hormonal tests of follicle-stimulating hormone, luteinizing hormone, thyroid-stimulating hormone, and prolactin were normal. However, anti-müllerian hormone was obtained 1.8 ng/ml.

Dimensions of the uterus included 52×53×127 cm with endometrial cavity enlargement in favor of a hematoma which spread to the lower part of the uterine trunk in magnetic resonance imaging (MRI). However, the cervix and vaginal lumen were not observed. On the left, the adnexa of the uterus showed endometriosis with dimensions of 29×23 cm (Figure 1).

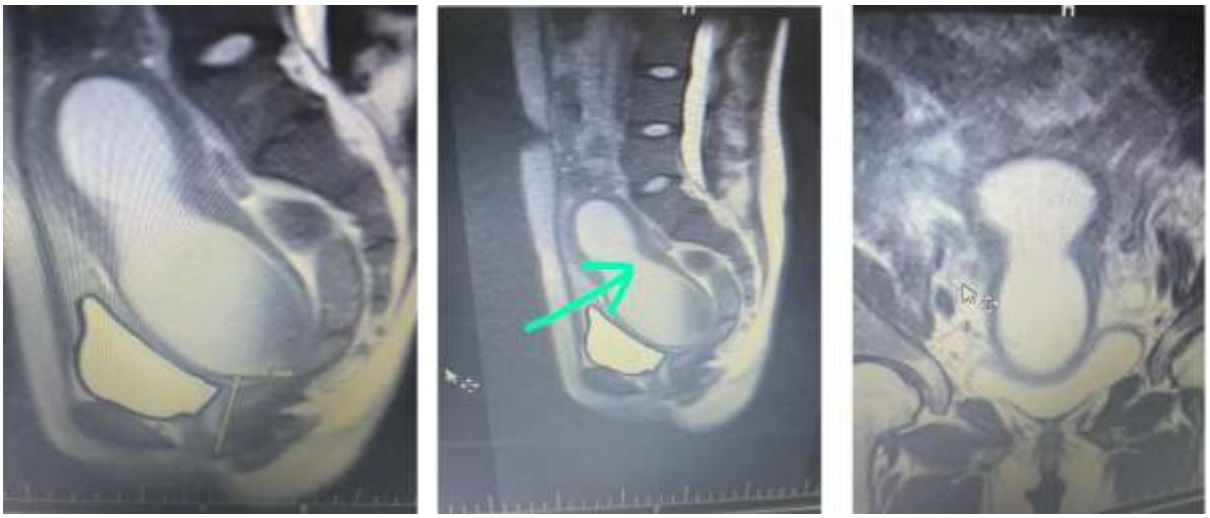

Figure 1. MRI image with endometrial cavity enlargement

Bulging, and the hysterometer was passed down the uterus. The cervix was not touched, and cervical agenesis was diagnosed. Because of the long-term complications and the importance of regular follow-up, the patient was examined under anesthesia. The vaginal entrance was touched by one cm. The rectal examination showed no mass in the first five cm, and the vaginal tissue was not touched. However, a large mass about 15 cm was touched below the umbilicus, the uterus was enlarged and was touched in the rectal examination;

However, the same cervical tissue was not touched.

The vaginal and cervical agenesis was diagnosed; therefore, hysterectomy was

suggested. After 10 days, the patient underwent laparotomy due to her parents' dissatisfaction with hysterectomy. After opening the abdomen, the appearance of the uterus was normal. The tubes were without hematosalpinx. A two-cm incision was made in the lower uterine segment. The blood retention in the uterus was removed, and the inside of the uterine cavity was examined with a finger, which apparently touched the cervix. Vaginoplasty was performed for the patient, and a Foley catheter was inserted into the uterine cavity from the bottom of the vagina. At the end of the operation, a tube was inserted into the vagina and the patient was discharged with a uterine catheter. However, the patient returned after one week, and she was examined under anesthesia. The uterine catheter was removed, and the vagina was washed with normal saline. During the examination, the balloon of the Foley catheter was touched above the high septum and a new catheter was inserted.

The patient returned after three months, during this time, she had three times spontaneous menstruation, and she presented with spontaneous removal of the uterine catheter. She was again examined under anesthesia. The septum was opened with bougie of number two, and the length of the uterus was about 4 cm. The insertion of the Foley and Metzer catheters failed. The patient was finally discharged and requested to return when there was no menstruation. The patient returned after 11 months due to abdominal pain and no menstrual bleeding for two months. The ultrasound was performed and she was referred with hematocolpos again. During examination at the operating room, the vagina was opened about 3 cm, the abdomen was opened, and the vaginal atresia was observed.

Surgical therapy included an incision in the lower part of the uterus at the site of the bulging and the use of a hysterometer passing down the uterus. The cervix was not touched, cervical agenesis was diagnosed and subject to obtaining informed consent of patient's parents the patient underwent abdominal hysterectomy and bilateral tubal salpingectomy (Figure 2). The pathological results confirmed the cervical agenesis.

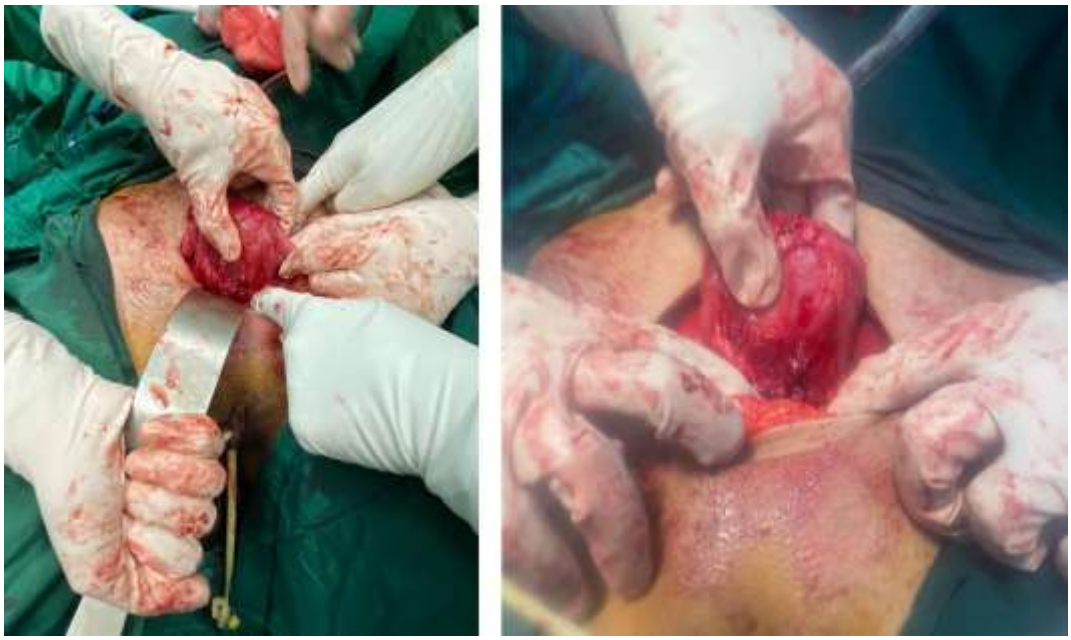

Figure 2. Surgical view of the abdominal hysterectomy

Discussion

In this study, a case of vaginal and cervical agenesis was reported. The differential identification of the vaginal septum includes an imperforate hymen which may be manifested with primary amenorrhea and pelvic pain. Patients with this condition usually have no vagina; however, about 25% of them have blind vaginal pouches (2). The frequency of congenital uterine abnormalities ranges from 0.3% to 10%. Regarding the embryologic etiology, a complex range of genital malformations is controversial due to inadequate progression in the embryonic developmental stage. The combination and canalization of Müllerian ducts progress in a caudad-to-cranial direction based on the classic unidirectional theory, which makes it incredible to clarify how the whole septate uterus developed in the absence of the cervix (3).

Vaginal septum differs in thickness, with thicker septum sited nearer to the cervix. The maximum common position of the septum is in the upper part of the vagina at the embryologic connection between the vaginal plate and the caudal end of the attached Müllerian ducts. According to Lodi's series, vaginal septum was situated in the upper third of the vagina (about 46%), the middle third (40%), and the lower third (14%) (4). In the present case, the septum was placed near the cervix at the upper end of the vagina, and it was very near to both bladder and rectum. At first, an identification of cervical atresia was supposed; however, after cutting the edges of the septum, it was established to be the high transverse vaginal septum.

An imperforate hymen is usually the greatest manifestation at the maturity of the female genital tract with a frequency between 0.01% and 0.05% in neonates, when no discharge tract is accessible for menstrual flow. The result of maternal estrogen secretion in both prenatal and postnatal periods might be the reason for mucus secretion by the cervical glands. The usual reasons for secretory hydrometrocolpos include an imperforate hymen, transverse vaginal atresia, and vaginal septum with or without the persistence of a urogenital sinus of cloaca (5). Hematometra with or without hematocolpos is common, which leads to recurrent cyclic lower abdominal pain without observable menstrual release and a mass was created in the central lower abdomen and pelvis. The current case referred with lower abdominal pain. On investigation, partial vaginal agenesis with a 4-cm vagina was observed. The pelvic ultrasonography showed hematometra without a cervix. The surgical therapy involved the creation of an ostium using a 1.5-cm midline vertical hysterotomy incision, trimming away the fibrous tissue at the distal of the uterus, and the proximal end of the vagina was attached to the uterus by the abdominal route.

There are three resection methods for transverse vaginal septum: laparoscopic resection, laparoscopic abdomino-perineal vaginoscopy, and simple vaginal excision (1). The maximum surgical approaches for cervical agenesis involve a transvaginal or transabdominal method to make an ostium over the thick fibrous cavity with uterus and vagina by the request of stents, with or without a surrounding complete or split-thickness skin graft (7). The most common complication of resection is vaginal stenosis. In some cases, postoperative vaginal dilation may be helpful in reducing scarring and stenosis (8). As reported by Sardesai and colleagues, this technique may result in subsequent closure of the haematocolpos hole, restenosis changes, and recurrence (9). Han et al. reported a 23-year-old woman with vaginal atresia and cervical agenesis. They suggested the probable completion of the upper fragment fusion of the Müllerian ducts. They reported that the resorption of the median septum didn't occur; however, a complete septum is formed subsequent to this condition. At the same time, at the lower fragment of the Müllerian duct, the fusion development did not happen or abnormal growth results in cervical agenesis and upper vaginal atresia (10). Dennie et al. reported that hysterectomy is the best treatment for a girl who begins menstruation with haematocolpos (11). Also fibrous tissues with stenosis can lead to postoperative intense infection or septicemia in some patients leading to recommend hysterectomy for this group (10). In the present case, the vaginoplasty was unsuccessful, in the next stage the pathological results confirmed cervical agenesis. According to the measures taken for the patient, it seemed that the best treatment was hysterectomy.

Conclusion

Based on the present case, the recent recommendation for treatment of cervical and vaginal agenesis is hysterectomy. However, it may not be a suitable treatment choice for some cases, and it is preferred to decide based on the patient's condition.

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conflicts of interest

Authors declare no conflicts of interest.

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