Case Report

Congenital Atresia of Uterine Isthmus: Successful Diagnosis and End-To-End Anastomosis

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A B S T R A C T

Background: Müllerian duct anomalies are rare and occasionally diagnosed in adolescents with primary amenorrhea, abdominal pain, and sexual difficulties. They are present in a variety of forms and sometimes difficult to appropriately classify. The management of malformations remains controversial.

Case: A 15-year-old girl with primary amenorrhea and cyclic lower abdominal pain was found on laparoscopic examination to have an asymmetric ball-shaped uterus with isthmus stenosis suspended in the pelvis. The junction between the lower segment of uterus and the cervix was very thin and stenotic with scar-like tissue changes. Combined with pathologic evaluation, it was finally diagnosed as congenital atresia of uterine isthmus. Thus, an end-to-end anastomosis was performed instead of surgical resection.

Summary and Conclusion: Müllerian duct anomalies in a variety of forms can be difficult to diagnosis correctly and treat appropriately. Preservation of reproductive ability is the first objective of all treatments.

Key Words: Müllerian anomaly, Atresia, Uterine isthmus, Anastomosis

Introduction

The incidence of congenital Müllerian duct anomalies is approximately 1%-3%.[1] They are occasionally diagnosed in adolescents with primary amenorrhea, abdominal pain, and sexual difficulties. According to the widely accepted American Fertility Society (AFS) classification revised in 1988, Müllerian anomalies include 7 distinct categories.[2] However, due to being present in a variety of forms, Müllerian anomalies are sometimes difficult to appropriately classify and the management of some malformations remains controversial. Regardless, preservation of reproductive ability is the first objective of all surgical treatments.

Case

A 15-year-old girl with primary amenorrhea had a history of 6 months of lower abdominal pain. She had been diagnosed with acute appendicitis at a hospital and thus underwent an appendectomy 2 months earlier. However, the symptom of her abdominal pain was not relieved after the surgery. Thus, she presented to our outpatient gynecology clinic at Jinling Hospital. Our examination showed that her secondary sex characteristics were normal (Tanner staging 4 and 5 for breast) and her hymen was intact. Considering that the patient was a young adolescent who may not tolerate a gynecologic examination, a bimanual rectal examination was performed initially, not helping with the diagnosis. The rectal ultrasonography showed that although her uteri corpus (3.5 × 2.6 × 3.5 cm) and ovaries (right 2.6 × 2.6 cm, left 1.9 × 1.1 cm) were normal, there was an echo (2.5 × 2.0 × 2.5 cm) in front of the uteri corpus and the endometrial line and cervix could not be visualized.

For economic reasons, the patient did not undergo pelvic magnetic resonance imaging (MRI) to further assess the development of her cervix. Instead, hysteroscopy was used to acquire the imaging features of her cervix. It revealed that the external orifice of the cervix had a normal appearance in both size and morphology and the vagina was 6 cm long. In addition, the blood tests and the abdominal ultrasound examination showed that her serum hormonal levels, kidneys, and ureter were all normal. She had no history of pelvic trauma or exposure in utero to diethylstilbestrol.

Based on this information, we speculated that the patient most likely had congenital cervical atresia. To confirm our speculation, a combined laparoscopic and hysteroscopic approach was taken after informed consent was obtained from the patient and her parents. The patient was placed in a dorsal lithotomy position under general anesthesia with a small speculum placed into her vagina. A metal probe was inserted through the external cervix but could only be inserted to 4 cm. Therefore, a hysteroscopy was performed to observe the cervical canal. It was found that although it had normal morphology, the cervical canal ended abruptly at the neocervix (Figs. 1B). Thus, laparoscopy was performed and revealed an asymmetric ball-shaped uterus that was suspended in the patient's pelvis, with the left ovary and round ligament smaller than the right, which seemed to lose support from the upper cervix (Fig. 2A). Moreover, although the fallopian adhesion between the broad ligament and the fimbria end was normal,

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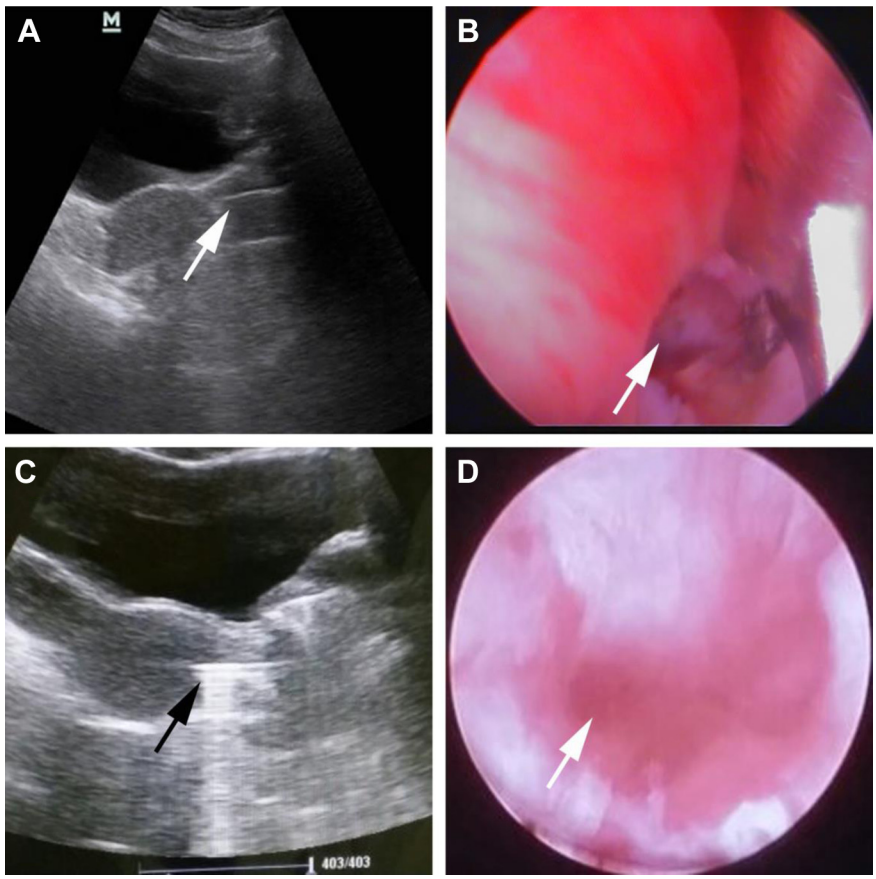

Fig. 1. (A) Preoperative uterus and probe located in the cervical canal. (B) Preoperative distal cervix like a blind end of cervix on hysteroscopy. (C) Postoperative uterus with the arrow located in the uterine cavity. (D) Postoperative distal cervix resembling a slit.

endometriosis was found in the right ovarian surface and the pelvic fossa (Fig. 2B, C). Thus, cauterization of pelvic endometriosis and lysis of adhesions were performed. With careful dissection of the vesicouterine space, a very thin stenotic junction was found between the lower segment of uterus and the cervix. Even after resection of the stenosis, there remained sufficient tissue to anastomose (Fig. 2D).

Via hysterotomy an end-to-end anastomosis of the lower part from the uterus to the cervix was performed (Fig. 3A). The stenosis tissues were found to be fibrous, but there was no hematometra drained from the cavity of uterus. Therefore, a catheter was inserted into the uterine cavity and anchored by its balloon (Fig. 3B). Antibiotics were administered for 24 hours postoperatively. In addition, because the patient had experienced regular periodic abdominal pain, the menstruation after surgery was predicted to probably cause cramps.

Thus, starting on postoperative day 2, a plan with estradiol tablets and dydrogesterone/estradiol tablets (estradiol 1 mg/d for the first 14 days, estradiol 1 mg and dydrogesterone 10 mg/d for the last 14 days of each month) for 3 months was carried out to allow restoration of the normal shape and size of uterus and prevent reformation of adhesions potentially caused by retrograde menstruation.[3,4] However, due to abdominal pain occurring within the first 11 days of the third month, the last administration was replaced by 3 days of estradiol 1 mg and 14 days of dydrogesterone 10 mg/d. The uterus and the cervix were found on ultrasonographic transabdominal imaging to be in their anatomically normal positions at postoperative day 5. In our patient, no contraceptive was used to avoid pregnancy, because our patient was a high school student and a virgin without sexual experience or risk of pregnancy. Combined with the pathology of the resected tissues, including uterine smooth muscle tissue and endometrial glands (Fig. 4), it was determined that the patient should be diagnosed with congenital atresia of uterine isthmus.

Finally, the patient left the hospital without pain or postoperative complications. A follow-up visit at postoperative months 1, 3, 6, and 12 was planned. For each visit, details about menstrual cycle and pain were sought, and the uterus, cervix, and ovaries were examined on ultrasonography. Because, 2 weeks later, the patient had normal menses, the catheter was removed from her body. Regrettably, during the hysteroscopy to observe the anastomosis site at 3 months, the probe could not pass through at the location suspected in the uterine cavity determined during abdominal ultrasound examination (Fig. 1C). The anastomosis site looked like a slit (Fig. 1D). However, the patient had regular menstrual periods, stable menstrual flow and no abdominal pain during the 6-month follow-up.