Case Report

Nearly Normal Congenital Cervical Fragmentation: A Hard-To-Diagnose and Successful End-To-End Anastomosis

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A B S T R A C T

Background: Congenital cervical fragmentation is a very rare genital tract malformation that usually presents in adolescence with primary amenorrhea and cyclic, monthly, lower abdominal pain. We report a nearly normal case of congenital cervical fragmentation and successful end-to-end anastomosis.
Case: A 15-year-old girl presented with primary amenorrhea with cyclic, monthly lower abdominal pain lasting for 15 months without any abnormal imaging findings (pelvic CT scan, ultrasonography, and hysteroscopy). Misdiagnosis and appendectomy was performed at the time of the initial lower abdominal pain. Diagnostic combined hysteroscopy and laparoscopy were performed in our hospital, and cervical fragmentation was diagnosed. A converted laparotomy end-to-end anastomosis was performed successfully, and regular menstruation was restored after the operation.
Summary and Conclusion: The malformation of nearly normal congenital cervical fragmentation is existent and hard to diagnose. As long as the patient has persistent primary amenorrhea with cyclic, monthly lower abdominal pain, even if no abnormal findings on imaging, obstructive malformation of the reproductive duct should be the primary suspicion. Diagnosis and treatment should occur as early as possible to preserve the patient's fertility. End-to-end anastomosis is the best method for this type of patient.
Key Words: Congenital cervical fragmentation, Anastomosis, Mullerian duct anomalies

Introduction

Congenital cervical fragmentation is a very rare genital tract malformation and is a type of congenital cervical dysgenesis. In 1995, Rock et al[1] organized congenital cervical agenesis and dysgenesis into the following 4 categories: cervical agenesis, cervical fragmentation, cervical fibrous cord and cervical obstruction. Grimbizis et al[2] have reviewed 116 cases with congenital cervical agenesis or dysgenesis from 1900 to 2004, in which congenital cervical fragmentation only accounted for 5.2 % (6/116) of cases, and only 1 successful cervical-to-cervical anastomosis procedure was performed. These patients were usually diagnosed in adolescence because of primary amenorrhea with cyclic, monthly lower abdominal pain and abnormal signs such as vaginal aplasia or abnormal imaging findings. In this report we describe the case of a 15-year-old girl with primary amenorrhea and cyclic, monthly, lower abdominal pain lasting for 15 months without any abnormal signs under gynecological examination, blood tests or imaging findings. To our knowledge, this is the second case of congenital cervical fragmentation without any abnormal imaging findings. Here, we report the case and review the relevant literature.

Case

A 15-year-old girl was admitted to our gynecology department for primary amenorrhea with cyclic, monthly, lower abdominal pain lasting for 15 months without any abnormal signs from gynecologic examination, imaging or blood tests. She underwent an appendectomy 15 months previously without abnormal findings of her ovaries at that time. Since then, cyclic lower abdominal pain occurred 5-6 days per month. She went to many gynecologic outpatient clinics and underwent many gynecologic examinations, pelvic CT, pelvic ultrasonography, chromosome and female hormone test, and even hysteroscopy, but all of them were normal. She also received cyclic estrogen-progestin therapy for 3 months, but still experienced amenorrhea. In July 2013 she came to our hospital. Physical examination revealed the development of secondary sex characteristics, and the female external genital phenotype was normal. Bimanual rectal examination was normal. No abnormality of the urogenital tract, such as uteri corpus and the cervix (Fig. 1, A), of either ovary, or of kidneys was found by ultrasonography. A combined diagnostic hysteroscopy and laparoscopy was performed under general anesthesia. After anesthesia, first, the patient was placed in a dorsal lithotomy position and then a small speculum was placed into her vagina. Normal vaginal and cervical morphology (Fig. 1, B) was observed. Second, a cannula was used to test the vaginal, cervical canal and endometrial cavity. The vaginal length was 7 cm, but the cervical canal length only allowed the cannula to be inserted 3 cm from the external cervical os

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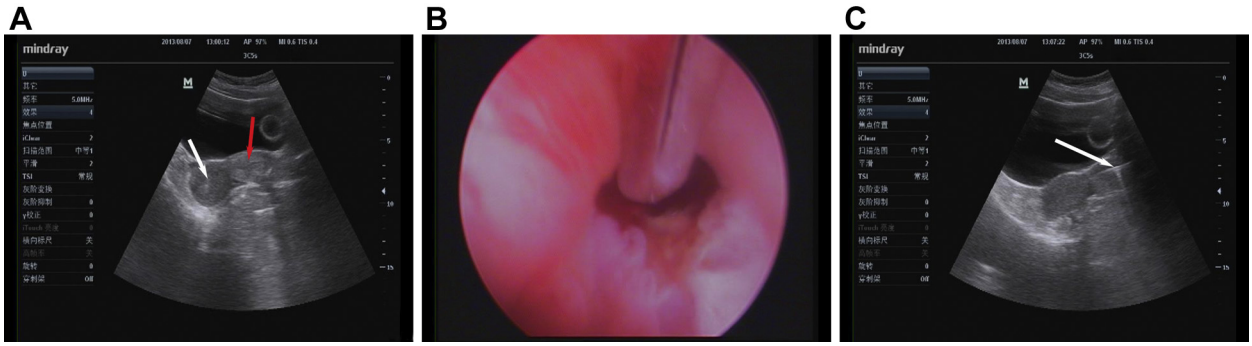

Fig. 1. (A) Nearly normal uteri corpus (*white arrow*) and cervix (*red arrow*) by ultrasonography; (B) normal morphology of cervical os by speculum; (C) cannula (*arrow*) inserted into the cervical canal only 3 cm from the external cervical os by ultrasonography.

(Fig. 1, C). Third, a hysteroscopy was performed. During the hysteroscopy, a transverse cervical diaphragm 3 cm from the external cervical os was observed; this feature prevented the entry of the cannula into the endometrial cavity. A hysteroscopic attempt to pass through the occluded cervical canal failed. Laparoscopy was performed. During laparoscopy, we found that the uterus was over-retroflexed, normally sized and had obviously high bladder-peritoneal reflection (Fig. 2, A). There was an abnormal feeling, as if there was no connection between the uterus and cervix, when the cervix was shaken. The left fallopian and ovary was adhered to the peritoneum. The left ovary was normal in size and morphology, and the left fallopian fimbrial end was blunt after adhesion lysis. The right fallopian tube was normal; 2 chocolate cysts with dimensions of approximately $2 \times 1 \times 1$ cm and $1.5 \times 1 \times 1$ cm were observed in the right ovary (Fig. 2, B), and a cystectomy was performed. After dissection of the bladder-peritoneal reflection space, the cervix was disconnected from the uteri corpus (Fig. 2, C) except for the peritoneum and a very small fibrous cord. We decided to convert the laparoscopy to laparotomy. In laparotomy, we found that both ends between the lower part of the uterus and the upper part of the cervix were occluded (Fig. 3, A). A surgical end-to-end anastomosis was performed via the following steps: (1) resect the both ends (lower uterine end and upper cervical end) about 5 mm separately until the both ostia were observed; (2) insert a 16F Foley catheter through the external cervical os into the endometrial cavity; (3) interrupted suture the 2 cut edges to each other with Ethicon 6 stitches of absorbable suture No. 2.0 around the Foley catheter (Fig. 3, B); (4) anchor the Foley's catheter with 4 ml of fluid pumped into the catheter balloon; (5) suture 1 stitch at the external cervical os and bind the suture to the Foley's catheter with Ethicon absorbable suture No. 2.0; and (6) suture the bladder-peritoneal reflection and close the incision. The procedure was uneventful. The Foley catheter was cut off at 1 cm from the external cervical os at the 5th day after operation and not removed until the next menstruation. Antibiotics were administered postoperatively for 3 days. There were no postoperative complications. The patient was discharged. Twenty days later she had normal menstruation. To date, she has had 8 normal menstruation cycles without lower abdominal pain or infection.

Discussion

According to ESHRE/ESGE (European Society of Human Reproduction and Embryology, European Society for Gynaecological Endoscopy) consensus, this case is classified as U0C4V0 (normal uterus and vagina, cervical aplasia).[3] According to the American Fertility Society 1988 classification, cervical congenital dysgenesis is type Ib and can be subdivided into the following 3 types: cervical obstruction, cervical fibrous cord and cervical fragmentation. In the first type, a well-formed cervix exists, but a portion of the endocervical lumen is obliterated; in the second type, a cervical cord of variable length and diameter is present with a completely obliterated endocervical canal; in the third

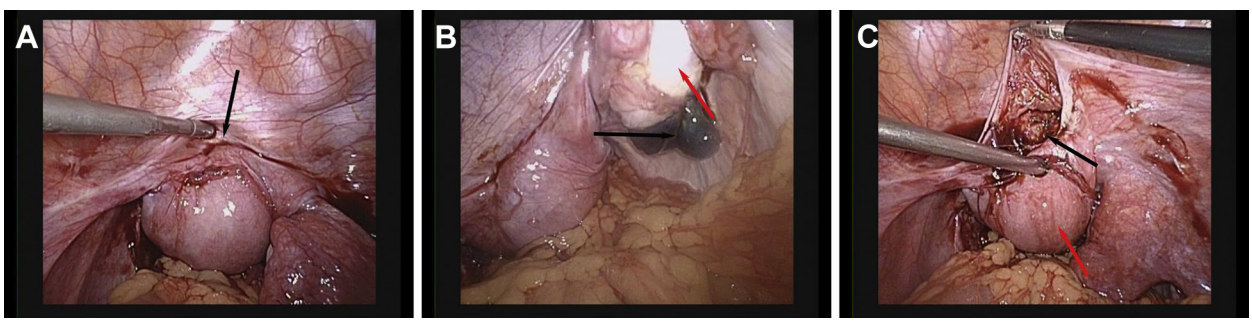

Fig. 2. Laparoscopy findings: (A) high bladder-peritoneal reflection (*arrow*); (B) 2 chocolate cysts (*black arrow*) in the right ovary (red arrow); (C) cervix (*black arrow*) disconnected from the uteri corpus (*red arrow*) after dissection of the bladder-peritoneal reflection space.