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CLINICAL ARTICLE

Clinical characteristics of congenital cervical atresia and associated endometriosis among 96 patients

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ABSTRACT

Objective: To describe the anatomic variety of congenital cervical atresia and to explore the relationship between this disorder and pelvic endometriosis. *Methods:* In a retrospective study, records were reviewed for 96 patients with a confirmed diagnosis of congenital cervical atresia treated at a center in Beijing, China, between January 1984 and October 2014. Data on demographic parameters, symptoms, anatomic features, and endometriosis were obtained and analyzed. *Results:* Of the 96 patients, 54 (56%) had pelvic endometriosis, 23 (24%) had a uterine malformation, 75 (78%) had a vaginal malformation, and 12 (13%) had a urinary malformation. The patients with a delay from first symptoms to surgery of more than 1 year had a higher incidence of endometriosis than did those with a delay of 1 year or less (45/71 [63%] vs 7/23 [30%]; $P = 0.006$), and this trend was not related to the severity of endometriosis ($P = 0.658$). Among the 31 patients with unilateral endometrial cysts, 20 (65%) had left-sided cysts and 11 (35%) had right-sided cysts ($P = 0.005$). *Conclusion:* More than half of patients with congenital cervical atresia had pelvic endometriosis. Early diagnosis and surgery seem to be necessary to prevent endometriosis among patients with congenital cervical atresia.

1. Introduction

Congenital cervical atresia is a rare Müllerian malformation of the female reproductive tract that was first reported by Ludwig in 1900 [1]. Its incidence and mechanism remain unknown. Because of the absence or obstruction of the cervical canal, primary amenorrhea and cyclic abdominal pain are the most common clinical symptoms. More than half of affected patients have either partial or complete vaginal aplasia [2]. Delays in the diagnosis and treatment of this condition can result in endometriosis, which can cause irreversible reproductive damage [3].

Because congenital cervical atresia is rare, no analysis of the clinical characteristics in a large patient sample (>50) has been reported [4–7], and few studies have explored the characteristics of endometriosis in patients with congenital cervical atresia. However, larger studies would be useful to develop an increased understanding of the association with endometriosis and would also aid in better defining the anatomic variability.

Peking Union Medical College Hospital in Beijing, China, has been designated as a center for the diagnosis and treatment of female genital abnormalities in China, and approximately 1600 women with genital tract abnormalities have been admitted for treatment over the past 30 years (1984–2014), including a total of 96 women with congenital cervical atresia. The aim of the present study was to retrospectively and systematically analyze the records of these 96 patients, aiming to describe the anatomic variations of this rare disease and to explore the relationship between congenital cervical atresia and pelvic endometriosis.

2. Materials and methods

In the present single-center, retrospective study, the records of patients with a confirmed diagnosis of congenital cervical atresia treated between January 1, 1984, and October 31, 2014, at Peking Union Medical College Hospital were reviewed and their long-term follow-up data were analyzed. The study was approved by the Institutional Ethics Committee of Peking Union Medical College Hospital. All included patients provided consent to future studies (including chart review and follow-up) at the time of their treatment.

The patients received a primary diagnosis on the basis of their symptoms (cyclic abdominal pain and adolescent primary amenorrhea), gynecologic examination (normal external genitalia, normal axillary and pubic hair, and normal female karyotype), and preoperative ultrasonography or magnetic resonance imaging (Fig. 1). Standard treatment of patients with congenital cervical atresia included surgery, the timing of which depended on the type of procedure to be performed, the severity of symptoms, and the age of the patient. Pelvic endometriosis was confirmed histopathologically via laparoscopy or laparotomy, and the severity of endometriosis was rated using the revised American Fertility Society classification system [8]. Follow-up occurred 4–6 weeks after

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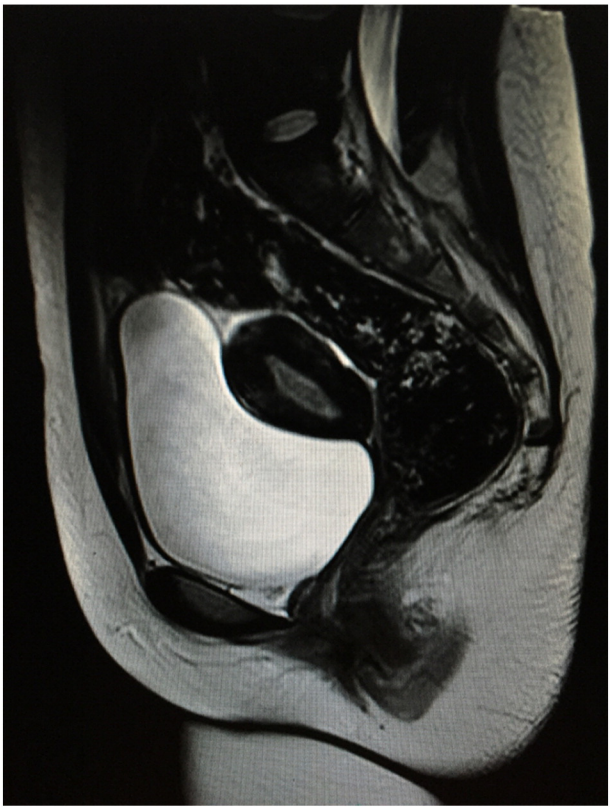

Fig. 1. Preoperative magnetic resonance imaging, showing a uterus with cervical agenesis.

Table 1
Characteristics of the patients with congenital cervical atresia (n = 96).

Anomaly observed	No. (%)
Symptoms	
Amenorrhea	96 (100)
Cyclic abdominal pain	93 (97)
Acute abdominal pain	1 (1)
Intermittent abdominal pain	2 (2)
Congenital cervical atresia	
Cervical agenesis	20 (21)
Cervical obstruction	30 (31)
Fibrous cord	25 (26)
Fragmentation	21 (22)
Associated upper-genital-tract malformations	
Left unicornuate uterus	7 (7)
Right unicornuate uterus	6 (6)
Bicornuate uterus	10 (10)
Associated upper-genital-tract lesions[a]	
Pelvic endometriosis[b]	52 (54)
Pelvic adhesions	44 (46)
None	14 (15)
Associated vaginal malformations	
Complete vaginal agenesis	34 (35)
Blind short vaginal pouches	41 (43)
Associated urinary malformations	
Kidney agenesis	9 (9)
Pelvic kidney	1 (1)
Horseshoe kidney	1 (1)
Ectopic ureteral orifice	1 (1)
Other associated malformations	
Scoliosis	2 (2)
Sixth finger malformation	1 (1)
Sacral canal cyst	1 (1)

[a] Some patients had several associated lesions.
[b] Two patients were excluded because they exhibited ovarian endometrial cysts after cervical canalization surgery.

surgery, and then annually thereafter. All patients were also followed up by telephone between February and June 2015, when they were asked about their pregnancy status.

Demographic data and information on the presenting symptoms, concomitant malformations, anatomic features, and postoperative complications were obtained from the operative notes, clinical records, and follow-up reports. The operative records were reviewed to confirm the diagnosis, and the type of cervical atresia was classified as cervical agenesis (no cervix) or cervical dysgenesis (incompletely formed cervix). Cervical dysgenesis was further subdivided into the following three categories: an intact cervical body with obstruction of the cervical os, a cervical body consisting of a fibrous band or cord, or cervical fragmentation.

The statistical analyses were performed with SPSS version 16.0 (SPSS Inc, Chicago, IL, USA). Categorical data were analyzed using either the χ^2 test or the Fisher exact test. Logistic regression analyses were used to determine the risk factors for endometriosis, which potentially included age at the onset of cyclic pelvic pain, body mass index, type of congenital cervical atresia observed, association with vaginal malformation, association with uterine malformation, and delay from symptom onset to surgery (≤1 year vs >1 year). The risk factors identified via univariate analysis were assessed via multivariate logistic regression. Odds ratios with 95% confidence intervals were calculated. $P < 0.05$ was considered statistically significant.

3. Results

Among the 96 patients—all of whom were Han Chinese—54 (56%) had pelvic endometriosis, 23 (24%) had a uterine malformation, 75 (78%) had a vaginal malformation, 12 (13%) had a urinary malformation, and 4 (4%) had another type of malformation, including two patients with scoliosis, one with a sixth finger malformation, and one with a sacral canal cyst (Table 1). The mean age at the time of symptom onset was 14.4 ± 2.1 years (range 12–20). The mean time from symptom onset to surgery was 40.2 ± 34.4 months (range 2–156). The median duration of follow-up was 62 months (range 8–371).

Among the 54 patients with pelvic endometriosis, three-quarters had ovarian endometrial cysts (Table 2). The remaining patients had peritoneal-type endometriosis or pseudo-rectovaginal endometriosis, which was defined as endometriosis below the adhesions of the posterior cul-de-sac but not at the rectovaginal septum.

Among the 31 patients with unilateral endometrial cysts, left-sided cysts were significantly more common (n = 20 [65%]) than right-sided cysts (n = 11 [35%]; $P = 0.005$). When the patients with bilateral endometrial cysts were included in the analysis, the incidence of left-sided cysts (30/51 [59%]) was higher than that of right-sided cysts (21/51 [41%]), but this difference was not statistically significant ($P = 0.075$).

Of the 41 patients with ovarian endometrial cysts, 13 (32%) were diagnosed with pelvic masses before being diagnosed with congenital

Table 2
Characteristics of the patients with endometriosis (n = 54).

Characteristic	No. (%)
Type of endometriosis	
Ovarian endometrial cyst	41 (76)
Left-sided	20 (37)
Right-sided	11 (20)
Bilateral	10 (19)
Peritoneal-type endometriosis	12 (22)
Pseudo-rectovaginal endometriosis	1 (2)
Stage of endometriosis[a]	
Minimal–mild	15 (28)
Moderate–severe	39 (72)
Type of congenital cervical atresia	
Cervical agenesis	12 (22)
Cervical obstruction	15 (28)
Fibrous cord	15 (28)
Fragmentation	12 (22)

[a] Endometriosis was staged by intraoperative exploration using the revised American Fertility Society classification system [8].

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