Delayed Diagnosis of Uterus Didelphys Unicolis with Cervical Atresia in a Forty-Three-Year Old Unmarried Woman with Primary Amenorrhea

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Müllerian duct fusion deficiency leads to uterus didelphys. We report the first case of a didelphys-unicolis uterus with atretic cervix in a 43-year-old unmarried woman with a thirty years history of cyclic lower abdominal pain and primary amenorrhea. On abdomino-pelvic ultrasound examination, a diagnosis of pelvic mass was suspected. On exploratory laparotomy, a didelphys-unicolis uterus with atretic cervix was found. Total abdominal hysterectomy and right salpingectomy were performed. To our knowledge, such a late diagnosed uterine didelphys unicolis with cervical atresia case in a 44 years old unmarried woman with primary amenorrhea has not been previously reported

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The prevalence of congenital anomaly of female reproductive system among the general population varies 0.001-10% but the true incidence is unknown. This may be due to inaccurate diagnosis or to the fact that many of these defects go undetected during a woman's lifetime. They may be caused by an insult during the first trimester, such as exposure to sex steroids, or may be due to polygenic/multifactorial inheritance. Uterine malformation from müllerian defect is the most common. Isolated anomaly of cervix or vagina, agenesis or hypoplasia of fallopian tube or ovary is rare.1,2

Bilateral deficient müllerian duct development leads to agenesis or hypoplasia of vagina, cervix, uterine fundus, fallopian tube, and any combination there of Failure of müllerian duct fusion leads to uterus didelphys.3

In the general population, the true incidence of uterus didelphys is unknown, but has been reported between 0.1% and 3.8%.4 Some may present at menarche with pelvic pain secondary to hematocolpos, while others present with pain, fever, and abscess formation.5,6

We want to report a case of uterus didelphys-unicolis with cervical atresia which is late diagnosed in a forty-three-year old unmarried woman with primary amenorrhea. To the best of our knowledge, such a late diagnosed case has not been previously reported in a forty-three-year old unmarried woman with a diagnosis of uterus didelphys-unicolis with cervical atresia.

Case Report

A 43-year-old unmarried woman with a thirty years history of cyclic lower abdominal pain and primary amenorrhea referred to our gynecologic clinic for pelvic mass. She was complaining increasing lower abdominal pain last six months. She reported no menarche and menses for thirty years but had a normal pubertal development. She had a history of oral analgesics using in the cyclic lower abdominal pain period and she had no relevant past general medical history or previous uterine or pelvic surgery.

Physical examination revealed normal secondary sexual characteristics. On gynecologic examination, vulva, vagina and hymen were normal. On rectal examination, uterus was larger than normal size but cervix was not palpated. Also a solid, mobile pelvic mass was palpated. All main laboratory parameters were entirely uncharacteristic. Abdomino-pelvic ultrasound examination revealed the presence of a 12x9x8 cm uterus with a cavity distended to 4 cm, and a pelvic cystic mass originating from right uterine side measuring 14x11x10 cm which is likely due to a hematosalphinx and solid pelvic mass originating from left uterine side measuring 6x5x4 cm.

Based on these findings, a diagnosis of pelvic mass was suspected. She underwent a exploratory laparotomy. A didelphys-unicolis uterus with atretic cervix was found initially. The right uterus was measured 12x9 cm with hematometra and hematosalphinx. The hematosalphinx of the right uterus extended to inferior region of liver. The left uterus was measured 6x5 cm with hematometra and salpingo-agementhenal agenesis (Figure 1). Bilateral ovaries were normal in appearance. Total abdominal hysterectomy and right salpingectomy were performed and a didelphys-unicolis uterus with atretic cervix confirmed by pathologist postoperatively.

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She was discharged at hospital postoperatively without any complications on the 5th day of admission.

Discussion

Uterus didelphys and unicolis is an embryonic malformation of the genitourinary system of the female that occurs between the 12th and 16th weeks of pregnancy. It is caused by the nonabsorption of the septum formed as a result of the fusion of these two ducts. The manifesting symptoms usually appear only after menarche and consist of dysmenorrhea, severe abdominal pain, and the presence of an intraabdominal or pelvic mass, hematometra, and hematocolpos, hematosalpinx.1-5 Uterus didelphys can present with discomfort from obstructed hemivagina, agenetic cervix or obstructed rudimentary horn.6

Uterus didelphys with obstruction of hemivagina, agenetic cervix or obstructed rudimentary horn has a large variation in time to accurate diagnosis. This may be due to several factors. Since only one uterine side is obstructed, the patient menstruates regularly from the other side delaying the diagnosis of outflow obstruction. Second, it is an uncommon condition, and therefore not often thought of as a diagnostic possibility. Third, the patients are frequently sent to a general gynecologist, gastroenterologist, or pediatric surgeon who may or may not be familiar with congenital anomalies and subsequently, fail to make the correct diagnosis and also when these patients present to their pediatrician or family physician with symptoms of cyclic dysmenorrhea or severe abdominal pain they are usually given anti-inflammatory drugs and oral contraceptives.7

Wu et al. described a 17-year-old woman suffering from cyclic lower abdominal pain for 3 years with agenetic cervix and didelphic uterus.8 Lee et al. described two women with hypoplastic cervix in the didelphic uterus whom underwent uterovaginal canalization and endometrial ablation of the obstructed uterine horn.9 Sherer et al described a case of uterus didelphys with a rudimentary right horn and right cervical atresia.10 Uterus didelphys also present itself as an adnexal or abdominal mass.11,12

In our case, we report a case of a delayed diagnosis of uterus didelphys-unicolis with cervical atresia which is mimicking pelvic mass in a forty-three-year-old unmarried woman with primary amenorrhea. To the best of our knowledge, such a late diagnosed case has not been previously reported in a forty-three-year-old unmarried woman. Because of the delay in the diagnosis of uterine didelphys-unicolis with cervical atresia, the dilated hematometra and massively right hematosalpinx presented itself as a pelvic mass in a forty-three-year-old unmarried woman with primary amenorrhea. While evaluating a patient with pelvic mass and primary amenorrhea, it is important to bear in mind the possibility of uterine didelphys or other mullerian anomalies. Early diagnosis of these anomalies may lead to early treatment, which may improve patients’ reproductive performance.

References


