CASE REPORT

Uterus Didelphys With Bilateral Cervical Agenesis

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Abstract

Background: The mullerian duct anomaly is a congenital abnormality of the female reproductive system caused by abnormal embryological development during pregnancy. If accompanied by cervical agenesis and infertility, intervention must be taken. Accurate diagnosis and proper treatment are very crucial to the future of reproduction and treatment of infertility in patients.

Objective: Reporting the handling of cases of uterine didelphys accompanied by bilateral cervical agenesis.

Method: Case report

Case: Reported cases of women aged 34 years with primary amenorrhea and 9 years primary infertility, not typical cyclic pain, normal secondary sex development and from gynecological examination obtained cervical agenesis. Transvaginal ultrasound examination found a mass with the appearance of adenomyosis. Laparoscopic performed show 2 masses, 1 mass resembling adenomyosis with a size of 9x6x5 cm located lateral to the left pelvis and another mass in the form of a hypoplastic uterus with a size of 2x2x1 cm visible 2 tubes with 2 ovaries within normal limits. Uterine mass resembling adenomyosis with a location far from the vagina making it difficult to do anastomoses while other uterus hypoplasia and non-functional. Hysterectomies were performed on the mass of adenomyosis with the results of PA was adenomyosis.

Conclusion: The uterus didelphys with bilateral cervical agenesis with 1 uterine adenomyosis and located in the pelvic lateral it was difficult to do uterovaginal anastomose so that hysterectomy was performed. Second uterine was hypoplasia and non-functional so that no action was taken. Need to think about “future fertility” in these patients and various options for having children.

Keywords: Primary Amenorrhea, Uterine Didelphys, Cervical Agenesis, Adenomyosis

INTRODUCTION

Amenorrhea is the absence of menstrual periods in women of reproductive age. This terminology comes from Greek: a = negative, men = month, rhoia = flow. Primary amenorrhea (the menstrual cycle never occurs) is likely to occur due to developmental failure such as the absence of a congenital uterus or failure of the ovaries to receive and maintain ovum cell.1

The characteristics of the Didelphys Uterus are due to the failure of the Mullerian Duct to fuse which results in the separation of the uterine cavity, two cervix and two vaginas. Because of the close association and developmental interrelation between the 2 tracts,
Mullerian duct abnormalities are usually related to Wolffian duct abnormalities which cause congenital anomalies in the kidneys and urinary tracts.\(^2\)

Cervical agenesis is classified as a type of Mullerian IB anomaly, according to the American Society of Fertility, is the result of an abnormal fusion of the Mullerian channel with urogenital sinus, or segment atrophy that normally forms the Mullerian system. The incidence of cervical agenesis is 0.01% in the general population. This represents about 3% of all uterine anomalies. Cervical agenesis is rarely associated with the presence of functional vagina and uterus and if associated with uterine function, hematometra will occur. It is estimated that only 4.8% of women with cervical agenesis have uterine function. This case begins with primary amenorrhea, the development of good sexual character and cyclic abdominal pain.

The main goal of the treatment of cervical agenesis is to reduce symptoms, achieve regular menstruation and restore fertility. This case management is a challenge because many cases of treatment failure and hysterectomy may be needed as well as how infertility is managed in these patients.\(^3,4\)

**CASE REPORT**

Reported case of a 34-year-old woman with primary amenorrhea and 9-year-old primary infertility, non-typical cyclic pain, normal secondary sex development (tanner 3-4) and from the gynecological examination cervical agenesis was obtained, the vagina was rather short, probe measuring obtained a 4 cm long vagina, no portio. Transvaginal ultrasound examination found hypoechoic mass with indistinct size limit 6.96 cm x 5.27 cm x 3 cm both ovaries within normal limits, right ovary measuring 2.40 cm x 1.02 cm and left ovary measuring 3 cm x 1.84 cm with the impression of adenomyosis and both ovaries within normal limits.

Laparoscopic performed show 2 masses, 1 mass resembling adenomyosis with a size of 9x6x5cm located lateral to the left pelvis and another mass of uterine hypoplasia with a size of 2x2x1cm visible 2 tubes, 1 tuba originating from the uterine hypoplasia and 1 tuba again from the mass of adenomyosis and the two ovaries within normal limits. Uterus adenomyosis is located in the pelvic lateral and far from the vagina so that it is difficult to do anastomose, other uterus hypoplasia and non-functional. Hysterectomy was performed on uterine adenomyosis with the result of PA adenomyosis.
Figure 1. A, B: External Genitalia and Stomach, C: Breasts, D: Vagina

Figure 2. Secondary Sex Development Table
Figure 3: Patient ultrasound: A: Uterine appearance 6.96 x 5.27 cm x 3 cm invisible cervix visualization, visible hypoechoic mass size 6.9 x 5.2 x 3 cm indefinitely filling the posterior wall of the uterus, feeding artery (-). C: The right ovary is 2.40x1.02 cm and the left ovary is 3x 1.84 cm. Impression: Adenomyosis and cervical agenesis

Figure 4. A. Intraoperative Pictures (Red Arrows: Uterine Possible Adenomyosis, Yellow Arrows: Nonfunctional Uterus, Ovarian Black Arrows and Left and Right Tubes out of each uterus) B. Uterine after hysterectomy (removed from the abdominal cavity with morcelator)
DISCUSSION

Diagnosis

Didelphys uterus is usually associated with renal agenesis in 25% of the cases. Isolated didelphys do not require treatment, in contrast to cervical agenesis, where 26 cases have been reported undergo hysterectomy. In view of this, an appropriate diagnosis for Mullerian duct anomalies is important for managing patients.

The diagnosis in this patient is made because the patient has Primary Amenorrhea, unclear cyclic abdominal pain, good development of secondary sexual characteristics (Tanner 3). From the ultrasound there was no cervix, intrabdominal mass was suspected of uterine adenomyosis, while both the ovaries and left and right tubes were within normal limits.

Management

Intraoperative (laparoscopic) findings show uterus didelphys, one hypoplasia, the other may be adenomyosis, and there are 2 tubes, of which 1 is from the hypoplastic uterus and 1 is the uterus from adenomyosis, 2 ovaries from each normal uterus. In the hypoplastic uterus (probe: 2x2x1cm) is a non-functional uterus so there is no point for anastomosis.

Uterus adenomyosis which is located laterally in the pelvis is technically difficult to do anastomosis because of its distance from the fornix, so a hysterectomy is performed. Open retroperitoneal space is performed to identify both ureters (eliminating the possibility of renal agenesis).

CONCLUSION

Failure to develop Mullerian ducts is often accompanied by urogenital anomalies. Accurate diagnosis determines the right and optimal management. Laparoscopic and / or laparoscopic approaches can be used in correcting this anomaly.

The goal of reconstructive surgery is to provide a channel for menstruation, to reduce pain and to maintain reproductive potential. Patients with fragmented atresia or cervix usually is a bad candidate for canalization so the total hysterectomy is a treatment option.

Hysterectomy is the treatment of choice for cervical agenesis because of the common complications of cervical recanalization and difficulty of getting pregnant.

Mullerian duct anomalies have separated into so many classifications in recent years, but this classification is to expose various clinical manifestations, management and prognosis. The case that we report is didelphys uterus with bilateral cervical agenesis in primary amenorrhea patients which is a very rare case and even few have reported the same case.

It is necessary to think about the follow-up of infertility management in these patients with bilateral cervical agenesis and uterine didelphys.
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