CASE REPORT

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Uterus didelphys associated with ovarian endometriosis in an infertile patient

Dvostruka materica udružena sa endometriozom jajnika kod infertilne pacijentkinje

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Abstract

Introduction. Uterus didelphys results when Mullerian duct fusion is completely arrested during development. We presented a rare case of nonobstructive uterus didelphys occurring simultaneously with an endometriotic cyst of the ovary. **Case report.** A twenty-nine-year-old, nulliparous patient was admitted to our Clinic for laparoscopic treatment of an endometriotic ovarian cyst. Diagnoses of right ovarian endometriotic cyst and nonobstructed uterus didelphys were established with bimanual pelvic exam and two-dimensional transvaginal ultrasound. Diagnoses were subsequently confirmed by laparoscopy and magnetic resonance imaging. Laparoscopic incision and drainage of the endometriotic cyst were performed, followed by biopsy and coagulation of endometriotic lesions. Histopathology confirmed ovarian endometriosis. Gonadotropin-releasing hormone analogue (GnRHa) was prescribed postoperatively, for a total of 3 months. Ten months after completion of treatment, the patients was without disease recurence. **Conclusion.** Nonobstructive uterus didelphys is rarely associated with ovarian endometriosis.

Key words:
uterus; congenital abnormalities; ovary; infertility; endometriosis; laparoscopy; treatment outocme.

Apstrakt

Uvod. Dvostruka materica nastaje kada potpuno izostane fuzija Milerovih kanala. U radu je prikazan redak entitet dvostruke materice neopstruktivnog tipa, udružene sa endometriotičnom cistom jajnika. **Prikaz bolesnika**. Pacijentkinja stara 29 godina, nulipara, primljena je na našu kliniku za laparoskopsku operaciju endometriotične ciste jajnika. Bimanuelnim ginekološkim pregledom i transvaginalnim 2D ultrazvukom dijagnostikovana je endometriotična cista desnog jajnika i dvostruka materica neopstruktivnog tipa. Dijagnoza je kasnije potvrđena laparoskopijom i magnetnom rezonancom. Urađena je laparoskopska incizija i drenaža sadržaja ciste, sa biopsijom i koagulacijom endometriotičnog žarišta. Patohistološki je potvrđena endometrioza jajnika. Postoperativno pacijentkinji je ordiniran analog gonadotropin oslobađajućeg hormona, u vremenu od tri meseca. Deset meseci nakon završenog kompletnog tretmana, pacijentkinja je bila bez recidiva bolesti. **Zaključak.** Dvostruka materica neopstruktivnog tipa je retko udružena sa endometriozom jajnika.

Ključne reči:
materica; anomalije; jajnik; neplodnost; endometrioza; laparoskopija; lečenje, ishod.

Introduction

Mullerian duct anomalies arise as a result of duct development failure, incomplete duct fusion or canalization, or incomplete reabsorption of medial uterine septum. The prevalence of female genital tract anomalies in the general population varies from 0.5% to 5.0% [1]. Uterus didelphys arises due to complete failure of Mullerian ducts to fuse and differentiate to form uterus and cervix during the 8th gestational week. This is a lateral fusion defect of the Mullerian ducts resulting in symmetrical nonobstructive uterus didelphys accompanied by a complete longitudinal vaginal septum [2]. Most women with a nonobstructive uterus didelphys are asymptomatic, but some present with dysmenorrhea or dyspareunia in the presence of a longitudinal vaginal septum. If hemivaginal obstruction is present, symptoms tend to occur, and include hematocolpos, hematometra, hematometrocolpos as well as a lower abdominal pain [3]. Uterus didelphys, without an obstructed hemivagina, is rarely associated with endometriosis. Endometriosis is more commonly present in

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the obstructive forms of uterus didelphys. Uterus didelphys has also been described as being part of the Herlyn-Werner-Wunderlich syndrome (HWWS), also known as obstructed hemivagina and ipsilateral renal agenesis (OHVIRA syndrome) [4, 5]. Majority of cases of nonobstructive uterus didelphys do not require surgical management. Excision of vaginal septum is indicated in obstructed unilateral vagina while a hemihysterectomy is indicated in rare cases of cervical hypoplasia or agenesis [6, 7].

Case report

A twenty-nine-year-old nulliparous woman was referred to our clinic from a primary healthcare centre for laparoscopic surgery of a right ovarian endometriotic cyst. The gynecologic history revealed that menarche occurred at the age of 14 years and that menstrual cycles have been regular since. The patient has been married for four years, but has failed to become pregnant despite trying during the last 2 years. The obstetric history revealed a nulligravida. The patient denied a history of previous gynecologic disease. Upon admission to our Clinic, a complete preoperative work-up was performed, including the bimanual pelvic exam and two-dimensional transvaginal pelvic ultrasound. The bimanual pelvic exam revealed a partial longitudinal septum in the superior third of the vagina. The double cervix was 1.5 cm long, and contained openings to two endocervical canals. Two separate, firm and mobile pelvic masses were palpable, each mass the size of a small female fist. The left adnexa were mobile and non-tender to palpation. The right ovary was enlarged and tense on palpation. Two-dimensional transvaginal pelvic ultrasound revealed the following: two completely separate and equal hemiuteri. The right hemiuterus was anteverted, measuring 48 × 33 mm, characterized by normal contours and an endometrial thickness of 6 mm. The left hemiuterus was anteverted, measuring 50 × 33 mm, was characterized by normal contours and an endometrial thickness of 6 mm. A right ovarian cyst filled with viscous fluid was seen, and measured 42 × 40 mm. The left ovary measured 32 × 24 mm, and contained multiple follicles, which measured up to 6 mm. Double uterine cervix had two endocervical canals, and measured 28 mm in length. Abdominal and urinary system ultrasound revealed normal findings. Both kidneys were of normal size and morphology and were in their typical location. After preoperative work-up and preparation, the patient underwent laparoscopy. The following were the laparoscopic findings: two uteri of equal sizes were visualized in the pelvic cavity, each equivalent to the size of a small female fist. The two were completely separate, at a distance of approximately 3–4 cm from each other. Each uterus was associated with one Fallopian tube, and the Fallopian tubes were of normal length with mobile fimbria. An endometriotic cyst containing hemorrhagic debris was visualized on the right ovary and measured 40 × 40 mm. The left ovary was of normal size, morphology and whitish in colour. The laparoscopic finding is shown in Figure 1. The isthmus of the posterior uterine wall of the right hemiuterus had a subserous fibroid, which measured 1 cm. Endometriotic implants were seen on the right uterosacral ligament. As cystectomy, which would encompass complete excision of the cyst and its pseudocapsule, was technically difficult to perform, incision and drainage of the cyst was done instead, followed by biopsy and coagulation of endometriotic lesions. The subserous fibroid was excised. Both specimens were sent for the histopathological analysis. Chromopertubation, using methylene blue dye, was performed to assess tubal patency; spillage of the dye from the right tube into the abdomen indicated the patency of the right tube while the left tube filled up without spillage and was determined to be occluded at the uterine horn. Postoperative recovery was uneventful. The patient was discharged from the hospital on the second postoperative day. Pelvic magnetic resonance imaging (MRI) with paramagnetic contrast was performed postoperatively. The MRI findings confirmed a uterus didelphys, which consisted of two non-communicating uterine cavities and two cervices. A partial septum was found in the proximal third of the vagina. The MRI is shown in Figure 2.

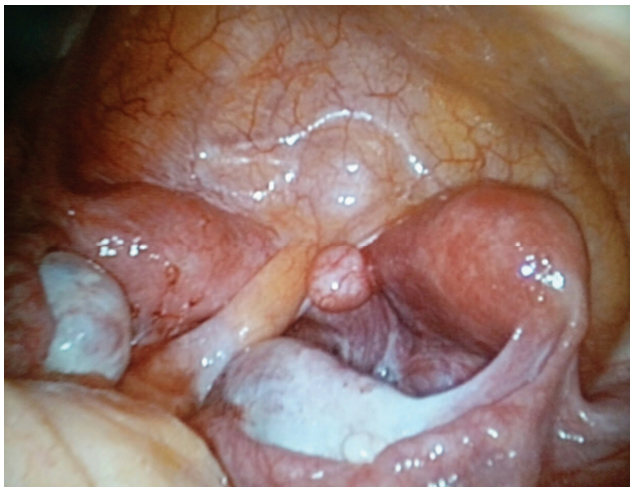

Fig. 1 – Laparoscopic findings shows two separate uterus.

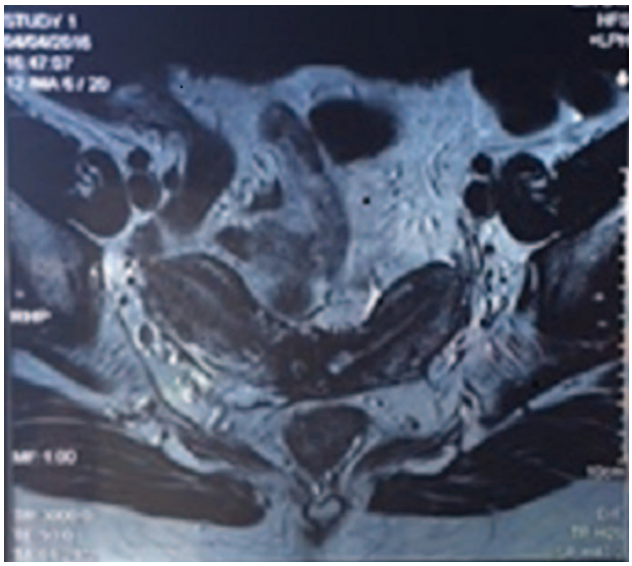

Fig. 2 – Magnetic resonance imaging (MRI) shows two separate uterus with two uterine cervix.

The final histopathological diagnosis included the following: endometriotic cyst of the right ovary; uterine fibroid. The histopathological finding is shown in Figure 3. The patient was managed with suppressive therapy using the gonadotropin-releasing hormone analogue (GnRHa), triptorelin (Diphereline® 3.75 mg), administered every 28–30 days for a total of 3 months. The patient complied with all 3 cycles of the treatment. The follow-up pelvic ultrasound and the CA-125 levels were performed and were within the normal reference range. The patient felt well and did not manifest signs of recurrent disease.

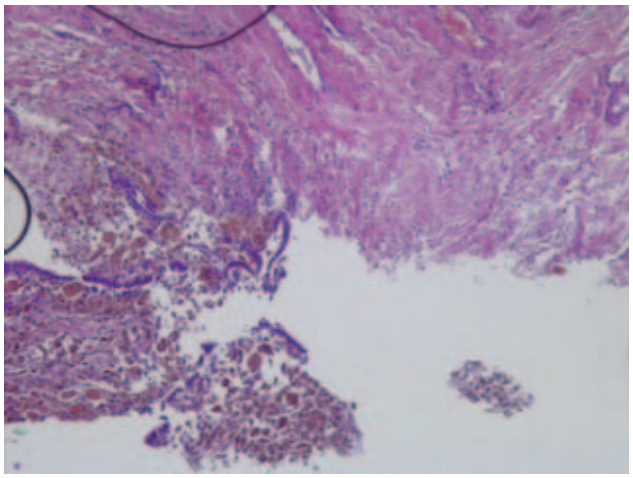

Fig. 3 – Histologic image of ovarian endometriosis (hematoxylin and eosin, ×10).

Discussion

Uterine malformations are a result of failure of Mullerian duct development, fusion, canalization or reabsorption. The prevalence of uterine anomalies in the unselected general population was 5.5%, compared to 8.0% in the population of infertile women, 13.3% in women with a history of miscarriage and 24.5% in those with miscarriage and infertility [8]. Multiple classification systems are used to classify female genital tract anomalies. The classification systems include the following: the American Society of Fertility and Sterility (AFSS); the American Society of Reproductive Medicine (ASRM); the European Society for Human Reproduction and Embryology (ESHRE) and the European Society for Gynecologic Endoscopy (ESGE) [1, 9]. Uterus didelphys constitutes 11% of all uterine malformations. Uterus didelphys arises when the Mullerian ducts completely fail to fuse leading to two separate uterine cavities and two cervices [10]. Each hemiuterus is associated with one Fallopian tube. There may be a single or double vagina. A partial or complete longitudinal septum is present in 75% of the cases [2]. Uterus didelphys was described as part of the Herlyn-Werner-Wunderlich syndrome (HWWS) [4], which is also known as obstructed hemivagina and ipsilateral renal agenesis (OHVIRA) [5, 10]. Uterus didelphys without an obstructed hemivagina is rarely associated with endometriosis. In relevant literature, there are reports of individual cases of nonobstructive uterus didelphys associated with ovarian endometriosis or peritoneal endometriosis [11]. We presented a patient with a uterus didelphys consisting of two completely separate uterine cavities and two cervices as well as a partial septum in the proximal third of the vagina, and a right ovarian endometriotic cyst. Our hypothesis is that endometriosis of the right ovary developed as a result of retrograde menstruation through the right Fallopian tube. This hypothesis is supported by the findings of chromopertubation during laparoscopy, which indicated that the right tube was patent, while the left tube was obstructed at the uterine horn. Endometriosis is more commonly present in the obstructive forms of uterus didelphys. Uterus didelphys with cervical agenesis is often associated with adenomyosis and ovarian endometriosis. Pelvic endometriosis was present in 19.15% of patients with HWWS [4]. Ipsilateral ovarian endometriosis develops almost always in cases of hemivaginal obstruction. Pelvic endometriosis is more frequent in the women with the complete hemivaginal obstruction and is found in 37% of cases, than in women with incomplete hemivaginal obstruction where it is found in 11.9% [4]. Endometriosis was found in 13.8% of women with OHVIRA, and 5.75% of these cases consisted of ovarian endometriosis [5]. The correlation between Mullerian duct anomalies and infertility is debatable. Mullerian duct anomalies occur in 3.4% of infertile women, while this prevalence increases to 4.3% in the general population, which implies that these anomalies do not negatively influence fertility [1]. Researchers reported that the women with a uterus didelphys had twin pregnancies, which developed normally within each of the uteri [12]. A term pregnancy delivered vaginally was described in the literature [13]. Most women with a nonobstructive uterus didelphys do not manifest any symptoms. If a thick longitudinal vaginal septum is present, common symptoms include dyspareunia and dysmenorrhea. Hematocolpos and hematometra arise as a result of an obstructed vaginal septum, leading to lower abdominal pain [3]. The HWWC syndrome is rare and occurs in 0.1% to 3.8% of cases. Hematocolpos and hematometra develop ipsilateral to the atretic hemivagina. This classical presentation occurs in 72.4% of HWW syndrome cases, while a rare variant of HWW syndrome occurs in 27.6% consisting of a uterine septum and cervical agenesis [14]. The diagnosis of uterus didelphys is usually established by bimanual pelvic exam, pelvic ultrasound (2D and 3D), hysterosalpingography, laparoscopy, hysteroscopy and magnetic resonance imaging [15]. The women with a nonobstructive uterus didelphys usually do not require surgical management. Excision of vaginal septum is indicated in the women with an obstructed unilateral vagina [6]. Hemihysterectomy is indicated in cases of uterus didelphys associated with unilateral cervical aplasia [7].

Conclusion

Nonobstructive uterus didelphys is rarely associated with ovarian endometriosis. Most women with a nonobstructive uterus didelphys do not manifest any symptoms. If a thick longitudinal vaginal septum is present, common symptoms include dyspareunia and dysmenorrhea. Endometriosis is more frequently associated with obstructive uterus didelphys, which manifests as hematocolpos and hematometra.

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