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A Rare Obstetric Outcome in Uterus Didelphys with Obstructed Hemivagina and Ipsilateral Renal Agenesis

Neuza Mendes, MD,^{1,2} Rita Lermann, MD,¹ Carla Francisco, MD,¹ and Fátima Palma, MD¹

Abstract

Background: The unique clinical syndrome of uterus didelphys, obstructed hemivagina, and ipsilateral renal anomaly is very rare and can be quite difficult to recognize because of the enormous heterogeneity in its clinical presentation. There are few long-term reports of the reproductive performance of women with this syndrome following treatment, or about the location of subsequent pregnancies. **Case:** A case in which two spontaneous pregnancies occurred alternatively in both hemiuteri: one despite a previous ipsilateral large hematometra and hematocolpos and the other, 8 years after, simultaneously with contralateral hematometra and hematocolpos (because of vaginal stenosis), is reported. Drainage of hematocolpos was performed at 14 weeks of pregnancy with immediate pain relief. **Results:** Pregnancy proceeded without complications. Eight months after delivery, a vaginoplasty was performed by excising the longitudinal vaginal septum, and marsupializing the vaginal cuff. **Conclusions:** This case highlights the importance of a correct and early diagnosis of developmental anomalies of the urogenital tract, as well as how a conservative approach in a Müllerian anomaly with unilateral obstruction led to two successful pregnancies occurring alternatively in the unaffected and in the previously blocked side. This is additional information supporting that every effort should be made to preserve the obstructed uterus. (J GYNECOL SURG 28:365)

Introduction

THE DEVELOPMENTAL ANOMALIES of the Müllerian duct system are some of the more intriguing and challenging disorders encountered by clinicians. They comprise a broad range of congenital defects that vary widely in clinical presentation, often leading to diagnostic difficulties.¹ The specific association of uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis was recognized and first reported in 1922 by Purslow.² The exact cause of this particular rare association is unknown, but this constellation of findings may be the result of an embryologic arrest at 8 weeks of pregnancy, which affects simultaneously the Müllerian and metanephric ducts.³

The incidence is unknown and probably underestimated.⁴

There are few long-term reports of the reproductive performance of these women following treatment, or about the location of subsequent pregnancies. Additionally, although the majority of recent articles support a conservative approach,^{5,6} there are several that report hemi-hysterectomy and ipsilateral hemicolpomy with or without concomitant salpingo-oophorectomy as a first surgical treatment.^{4,6-8}

The literature review described few women who had pregnancies in the previously obstructed uterus.^{4-9,10} An unusual case in which pregnancy occurred alternatively in both hemiuteri is presented here. Moreover, the last pregnancy (contralateral to the affected side) was simultaneous with a hematocolpos and a hematometra because of vaginal stenosis consequent to an incomplete vaginal septum resection.

Case

A 25-year-old woman presented complaining of severe right lower quadrant pain and amenorrhea.

Her medical history included a spontaneous menarche at 11 years of age with dysmenorrhea and irregular menses ever since. Thirteen months later, she sought medical help in a different medical tertiary care institution because of severe lower quadrant abdominal pain and rectal pressure. Physical examination revealed a healthy young female with normal secondary sexual development (breasts were Tanner stage 3-4, axillary hair was normal, pubic hair was Tanner stage 4-5). Pelvic examination showed normal external genitalia

¹Departments of Gynecology and Obstetrics, Centro Hospitalar de Lisboa Central-Maternidade Dr. Alfredo da Costa, Lisbon, Portugal.

²Nova Medical School, Universidade Nova de Lisboa, Lisbon, Portugal.

and normal introitus. A one-finger vaginal examination revealed a right paravaginal mass. An occluding longitudinal vaginal septum determining a right upper hemato-colpos was diagnosed after sonography. To further evaluate and characterize the vaginal septum and investigate possible associated anomalies, magnetic resonance imaging was performed. It demonstrated two separate uteri with an external contour delimiting two widely separated uterine horns, two cervixes, an occluding longitudinal vaginal septum, a right hemato-colpos and hematometra, and an absent right kidney. Therefore, the diagnosis of didelphic uterus and right renal agenesis was made. At that time, the physicians involved performed drainage of the hemato-colpos through a 2 cm laser incision in the vaginal septum. The patient abandoned medical care soon after this procedure. She reports that, thereafter, menses occurred at regular monthly intervals with normal flow without dysmenorrhea. A spontaneous pregnancy, located in the previously obstructed site, as demonstrated by the obstetric ultrasounds, occurred at the age of 18. Speculum examination revealed a cervix on the left side of the vault of the vagina and a septum with a central defect partially obstructing the right part of the vagina. Pregnancy was monitored at the same hospital where she had previously been a patient. No complications were noted. An elective cesarean section was performed at 39 weeks of gestation because of breech presentation. The newborn was a male weighing 3030 g with an Apgar score of 9/10. Unfortunately, no postpartum examination was performed because the patient abandoned medical care again.

Therefore when the patient presented at our hospital, (Centro Hospitalar de Lisboa Central-Maternidade Dr. Alfredo da Costa), 7 years later, at the age of 25, her didelphic uterus, vaginal longitudinal septum, and right renal agenesis were already known. Details described previously were subsequently investigated by contact with the previous hospital. Physical examination revealed a palpable painful mass at the right vaginal apex and a palpable uterus above the symphysis pubis. No cervix could be seen on vaginal examination and only the left one was palpable, because of the medial bulging of the right vagina. These findings, together with the amenorrhea, led to further investigation. Abdominal, pelvic, and transvaginal ultrasound revealed a second trimester pregnancy (14 weeks and 2 days) located in the left uterus, a right hemato-colpos, and a large ipsilateral hematometra, as well as an absent right kidney. The fetus had no visible malformations. An incision was made on the occluding longitudinal septum, and ~500 mL of dark brown blood was drained. Relief was immediate. Pregnancy proceeded without complications, with normal fetal growth monitored by serial ultrasound. At 36 weeks of gestation, there was a spontaneous rupture of membranes. The cervix was 1 cm dilated, 80% effaced, and vertex at station -2. Labor progressed spontaneously until the cervix was 4 cm dilated, at which time a cesarean section was performed because of suspected fetal compromise (lack of fetal heart rate variability and late fetal heart rate decelerations). The newborn was a female weighing 2475 g, with an Apgar score of 10/10. The cord blood pH was 7.23.

Eight months after delivery, a vaginoplasty was performed by excising the longitudinal vaginal septum and marsupializing the vaginal cuff. The patient was comfortable in the postoperative period, continues to have regular evaluations, and has been asymptomatic ever since.

Discussion

The condition represented by the presence of obstructed hemivagina, uterus didelphys, and ipsilateral renal anomaly is difficult to locate in an index or journal report because of different words used by different authors, who sometimes name the same set of anomalies differently. Since the mid-1980s, the triad has been called "Herlyn-Werner-Wunderlich syndrome" in several articles (first in articles in Bulgarian and then, since 2006, in articles in English).⁹ Because many authors disagree with the triad being named, this particular association of congenital anomalies is also referred to as the "no eponym" syndrome.¹¹ The acronym OHVIRA has been used to describe patients with an obstructed hemivagina and ipsilateral renal anomaly, as suggested in 2007 by Smith and Laufer.⁵ As it includes only two of the three components of the triad, it enables the inclusion of uterine anomalies, other than the more common uterus didelphys, such as a completely septated uterus (in up to 22% of cases, in some series).¹⁰ Given the lack of uniformity in the denomination of the syndrome, review of the literature is also difficult. For example, although the word "septum" usually describes the midline longitudinal and nonobstructive septum of the isolated uterus didelphys, in the context of the abovementioned triad it has been used to describe the medial wall of the upper unilateral hemato-colpos. Also, the hemato-colpos has been referred to as "blind vagina."

The greater awareness of this rare Müllerian anomaly and the higher level of suspicion among physicians has led to an increase in case reports of this congenital disturbance. This has enhanced the ability to recognize the enormous heterogeneity in its clinical presentation. In the presence of a more frequent obstructing vaginal longitudinal septum, as in the case described, the classical clinical scenario is that of a postmenarchal girl with pelvic pain and a medial bulging of a vaginal wall. Nevertheless, communication can exist between the two vaginal orifices. In these cases, menses can be regular and symptoms usually present later, the most frequent being intermittent mucopurulent discharge, because vaginal flora is allowed to grow in the accumulated menstrual fluid above the obstruction.¹²

An obstructed hemivagina should prompt the investigation of a possible renal anomaly. Therefore, the identification of an absent functional kidney in a patient experiencing pain and with a vaginal longitudinal septum should alert the practitioner to the strong possibility of the diagnosis. Unfortunately, and illustrating the difficulty in identifying a rare congenital multiple anomaly, this patient did not benefit from a correct diagnosis prior to referral. This can frequently lead, as occurred in the present case, to inappropriate care.

In this case, the obstruction was on the right side. The more common occurrence of the obstructed hemivagina and renal agenesis on the right side was noted by several authors but it is not exclusive.^{5,6,13,14} Even though a variety of urologic anomalies can be found, the more frequently reported, as in this case, is the ipsilateral renal agenesis. Several series reported other anomalies or even patients with two normal kidneys.^{5,7,8,10} Smith and Laufer, in the largest series limited to modern management described to date, including 27 cases of obstructed hemivagina and ipsilateral renal anomaly, reported: 20 cases of ipsilateral renal agenesis (74%), 1 case of

dysplastic and 1 case of polycystic kidney, 3 cases of bilateral normal kidneys, 1 contralateral duplex collecting system, 1 urethral diverticulum, and 1 rectovesical band.⁵ Other anomalies, such as an ectopic ureter, have also been described.¹⁵ The majority of case series report a double, mainly didelphic, uterus.^{4-8,16,17} However, there are cases of other types of uterine anomalies described, and the external shape of the uterus was frequently unknown, as diagnosis was generally achieved by hysterosalpingography.¹⁸

Few reports exist of the reproductive performance of these women following treatment. As expected, pregnancy occurs much more often in the uterus not previously obstructed. Nevertheless, there are at least 9 previously reported cases of ipsilateral pregnancy after surgical treatment of the obstructed site:

1. Monks reported a case of a 12-year-old girl with obstructed hemivagina, right upper hematocolpos, right hematometra, and ipsilateral renal agenesis.¹⁹ Later in life, and after resection and marsupialization of the vaginal septum, she had two successful pregnancies in the previously obstructed right uterus.
2. Stassart et al. reported a series of 15 patients with the syndrome.⁶ Six of these 15 women gave birth to a total of 9 live infants after surgical treatment. In only 1 patient did pregnancy occur in the affected side, with a cesarean section performed at term because of breech presentation. Before resection of the septum, the affected hemiuterus had been noted to be 10 cm in diameter.
3. In 1999, two distinct interesting series were published. Haddad et al. described 7 women with didelphic uterus and obstructed hemivagina who became pregnant after surgical correction, while investigating 42 cases of blind hemivagina.¹⁰ Heinonen described 9 cases of unilateral distal hematocolpos with distal vaginal agenesis and ipsilateral renal agenesis during his investigation of the clinical implications of 49 cases of didelphic uterus.⁷ In the series described by Haddad et al., three of the seven pregnancies were ipsilateral (one vaginal term delivery, one ectopic pregnancy and one early spontaneous abortion). Of the 9 women described by Heinonen, only 6 achieved delivery. Of these, 4 had pregnancies contralateral and 1 had a pregnancy ipsilateral to the affected side, and 1 had pregnancies alternately in both hemiuteri, as is described in the present case report. This is the only case previously described of pregnancies occurring alternatively in both hemiuteri.
4. In 2006, Chen and Yang reported a case of a 28-year-old woman with uterus didelphys, distal atresia of the right vagina, right hematocolpos, ipsilateral renal agenesis, and a left ovarian mature teratoma that had a pregnancy located in the right uterus 6 months after surgical treatment of the hematocolpos and teratoma.²⁰ An elective cesarean section at term was performed because of breech presentation.
5. Altchek and Paciuc reported a woman with uterus didelphys, unilateral distal vaginal agenesis, and ipsilateral renal agenesis who, 3 years after complete excision of the bulging right vaginal wall, had a voluntary early interruption of pregnancy in the previously obstructed right uterus.¹¹ Ten months later, she had an early

spontaneous complete abortion (the uterus in which it was located was not identified). Three years later she had another pregnancy in the right uterus. Pregnancy proceeded without complications and, at 36 weeks of gestation, a vacuum cup-assisted delivery was performed.

This substantiates that the functional recovery of the dilated uterus and its corresponding tube is possible once the obstruction has been corrected.

The patient described here was initially managed by another physician who had failed to recognize the true nature of the syndrome. Therefore, the patient underwent an incomplete surgical procedure (simple incision of the septum with drainage of the hematocolpos and hematometra). The spontaneous closure occurred 13 years later, during a contralateral second trimester pregnancy. There are reported cases of hematocolpos or pyocolpos after spontaneous septum closure of women treated by incision alone.^{10,14} Although there are many reports of patients managed by hemi-hysterectomy with or without concomitant salpingo-oophorectomy as a first surgical approach,^{4,6-8} at present the vast majority of physicians support an initial conservative surgical procedure. Ideally, a single-stage vaginoplasty should be performed (excising the occluding longitudinal vaginal septum and marsupializing the vaginal cuff). A two-stage vaginoplasty should be planned in the presence of infection and/or anatomic complex vaginal distortion that impedes satisfactory assessment for repair. Postoperative complications following vaginoplasty are uncommon. However, there are descriptions of postoperative stenosis.^{5,10} As previously demonstrated by several reported cases of pregnancy ipsilateral to the initially obstructed site (despite previous hematometra, hematocolpos, hematosalpinx, pelvic endometriosis, or adhesions), every effort should be made to avoid a hemihysterectomy and adnexectomy. Strassmann-type metroplasty has been used as a surgical approach in cases of uterus didelphys in the context of the syndrome described.^{6,8} Recurrent and late miscarriages are the main indications for uterine unification. Therefore, in a didelphic uterus that has not been associated with pregnancy complications or infertility, surgical intervention should not be routinely recommended.²¹

Conclusions

In conclusion, we present a case with an unusual obstetric outcome in a rare Müllerian anomaly, which highlights the importance of a correct and early diagnosis to avoid further complications and aggressive surgical treatment.

Disclosure Statement

No competing financial interests exist.

References

1. Dukovski A. Difficulties in the diagnosis and treatment of uterine malformation. *Akush Ginekol (Sofia)* 1993;32:37.
2. Purslow CE. A case of unilateral haematocolpos, haematometra and haematosalpinx. *J Obstet Gynecol Br Emp* 1922; 29:643.
3. Singh M, Geareart JP, Rock JA. Double urethra, double bladder, left renal agenesis, persistent hymen, double vagina and uterus didelphys. *Adolesc Pediatr Gynecol* 1993;6:99.

4. Zurawin RK, Dietrich JE, Heard MJ. Didelphic uterus and obstructed hemivagina with renal agenesis: Case report and review of the literature. *J Pediatr Adolesc Gynecol* 2004;17:137.
5. Smith N, Laufer M. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome: Management and follow-up. *Fertil Steril* 2007;87:918.
6. Stassart JP, Nagel TC, Prem KA, et al. Uterus didelphys, obstructed hemivagina, and ipsilateral renal agenesis: The University of Minnesota experience. *Fertil Steril* 1992;57:756.
7. Heinonen PK: Clinical implications of the didelphic uterus: Long-term follow-up of 49 cases. *Eur J Obstet Gynecol Reprod Biol* 2000;91:183.
8. Candiani GB, Fedele L, Candiani M. Double uterus, blind hemivagina, and ipsilateral renal agenesis: 36 cases and long-term follow-up. *Obstet Gynecol* 1997;90:26.
9. Kimble RMN, Kimble RM. The obstructed hemivagina, ipsilateral renal anomaly, uterus didelphys triad. *Fertil Steril* 2010;93:e15.
10. Haddad B, Barranger E, Paniel BJ. Blind hemivagina: Long-term follow-up and reproductive performance in 42 cases. *Hum Reprod* 1999;14:1962.
11. Altchek A, Paciuc J. Successful pregnancy following surgery in the obstructed uterus in a uterus didelphys with unilateral distal vaginal agenesis and ipsilateral renal agenesis: case report and literature.
12. Badawy SZA, Musa AG, Houck BM. Uterus didelphys associated with obstructing longitudinal vaginal septum and hematocolpos. *J Gynecol Surg* 2005;21:89.
13. Rock JA, Jones HJ. The double uterus associated with an obstructed hemivagina and ipsilateral renal agenesis. *Am J Obstet Gynecol* 1980;138:339.
14. Morgan MA, Thurnau GR, Smith ML. Uterus didelphys with unilateral hematocolpos, ipsilateral renal agenesis and menses: A case report and literature review. *J Reprod Med* 1987;32:47.
15. Wang ZJ, Daldrup-LinK H, Coakley FV, Yeh BM. Ectopic ureter associated with uterine didelphys and obstructed hemivagina: Preoperative diagnosis by MRI. *Pediatr Radiol* 2010;40:358.
16. Orazi C, Lucchetti MC, Schingo PM, et al. Herlyn-Werner-Wunderlich syndrome: Uterus didelphys, blind hemivagina and ipsilateral renal agenesis: sonographic and MR findings in 11 cases. *Pediatr Radiol* 2007;37:657.
17. Altintas A. Uterus didelphys with unilateral imperforate hemivagina and ipsilateral renal agenesis. *J Pediatr Adolesc Gynecol* 1998;11:25.
18. Shah D K, Laufer M. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA) syndrome with a single uterus. *Fertil Steril* 2011;96:e39.
19. Monks P. Uterus didelphys associated with unilateral cervical atresia and renal agenesis. *Aust N Z J Obstet Gynaecol* 1979;19:245.
20. Chen FP, Ng KK. Term pregnancy at the site of atresia following vaginal canalization in a case of uterus didelphys with hemivaginal atresia and ipsilateral renal agenesis. *Taiwan J Obstet Gynecol* 2006;45:366.
21. Musich JR, Behrman SJ. Obstetric outcome before and after metroplasty in women with uterine anomalies. *Obstet Gynecol* 1978;52:63.

Address correspondence to:

Neuza Mendes, MD

Rua Viriato

1069-089 Lisbon

Portugal

E-mail: neuzamdm@hotmail.com