Case Report/Caso Clínico DOI: 10.69729/aogp.v20i1a06

Congenital Uterine Anomalies: Diagnostic challenges and implications on pregnancy outcomes. A Case Report

Anomalias Uterinas Congénitas: Desafios diagnósticos e implicações no desfecho da gravidez. Caso Clínico

Filipa Ladislau[1], Ana Elisa Sousa[1], Florência Sepúlveda[1], Marta Luísa Rodrigues[1]
Unidade Local de Saúde Amadora/Sintra – Hospital Professor Doutor Fernando Fonseca

Abstract

Congenital uterine anomalies (CUAs) are often asymptomatic but can significantly impact on reproductive outcomes. We report the case of a 33-year-old multiparous woman who presented to the emergency department with pelvic pain and vaginal bleeding. Transvaginal ultrasound suggested a rudimentar horn ectopic pregnancy, prompting surgical intervention. Intraoperative findings revealed a complete bicornuate uterus, with the pregnancy located in the left hemicavity. Delivery was planned by cesarean section due to breech presentation and complicated by postpartum hemorrhage. This case highlights the diagnostic challenges of different CUAs, with a direct impact on clinical management, frequently only recognized during surgical exploration.

Keywords: Bicornuate uterus; Congenital uterine anomalies; Pregnancy; Ultrasonography.

Resumo

As anomalias uterinas congénitas (ACUs) são frequentemente assintomáticas, mas podem ter impacto significativo na gravidez. Este caso descreve uma multípara de 33 anos que recorreu ao serviço de urgência por dor pélvica e hemorragia vaginal. A ecografia transvaginal era sugestiva de gravidez ectópica num corno uterino rudimentar, tendo sido decidido tratamento cirúrgico. Intra-operatoriamente, identificou-se um útero bicórneo completo com uma gravidez na hemicavidade uterina esquerda. Foi proposta cesariana eletiva por apresentação pélvica, complicada com hemorragia pós-parto. Este caso ilustra os desafios diagnósticos entre as diferentes ACUs, com implicações diretas na decisão clínica, muitas vezes identificadas apenas em contexto cirúrgico.

Palavras-chave: Útero bicórneo; Anomalias congénitas uterinas; Gravidez; Ecografia.

INTRODUCTION

Congenital uterine anomalies (CUAs) are often asymptomatic and detected incidentally during routine gynecological examinations or imaging. Nevertheless, they can have profound implications for reproductive outcomes. Despite refinements intended to improve the specificity of CUA classification, the lack of a universally accepted framework continues to hinder consistency in clinical practice and scientific communication. Diagnosis relies primarily on imaging; however, limited access to three- dimensional (3D) ultrasound and overlapping clinical presentations can obscure findings, occasionally necessitating surgical

1. Hospital Prof. Doutor Fernando Fonseca, Amadora, Portugal.

exploration for definitive confirmation. We report a case initially suspected as an ectopic pregnancy that ultimately proved to be an intrauterine gestation within one hemicavity of a complete bicornuate uterus.

CASE REPORT

A 33-year-old multiparous woman (gravida 3, para 2) with no relevant personal or family medical history presented to the emergency department with a 24-hour history of vaginal bleeding accompanied by pelvic pain. Speculum examination revealed a normal-appearing vagina and cervix, without evidence of active hemorrhage or visible lesions. Only scant traces of old blood were noted, although the exact source of bleeding could not be determined. Transvaginal ultrasound revealed a viable embryo with cardiac activity inside a gestational sac, apparently located outside the uterine cavity, with a crown-rump length consistent with approximately eight weeks of gestation (Figure 1). Visualization of the left ovary was impeded by an interposed mass corresponding to the gestational sac, raising initial suspicion of a left tubal or cornual ectopic pregnancy.

Despite maintaining hemodynamic stability, the patient experienced persistent abdominal pain, and the presence of a viable embryo significantly heightened the risk of tubal rupture and clinical deterioration. Laparoscopy was therefore selected as the most appropriate approach, offering both diagnostic confirmation and immediate therapeutic intervention if necessary. However, the uterus exhibited an atypical morphology, making it unclear whether the case represented an ectopic pregnancy within a communicating left rudimentary uterine horn or an intrauterine pregnancy in the left uterine hemicavity of a bicornuate uterus. Due to this uncertainty and the technical difficulty of the procedure, the surgery was converted to an exploratory laparotomy. This approach allowed for definitive identification of a bicornuate uterus, with the left hemicavity slightly enlarged and exhibiting a soft consistency, consistent with an intrauterine pregnancy. In contrast, the right hemicavity demonstrated a firm texture, characteristic of a non-gravid uterine horn (Figure 2). There were no complications in the post-operative period.

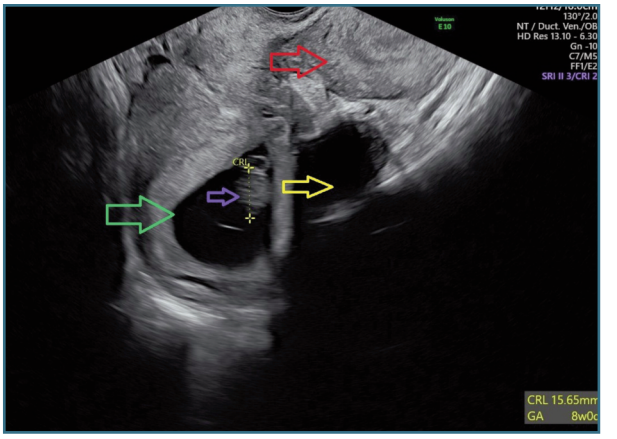

FIGURE 1. Ultrasound image of a bicornuate uterus with an eight-week embryo located in the left uterine cavity; Initial suspected ectopic pregnancy in a rudimentar horn. (Red arrow – Uterus; Green arrow – Initial suspected ectopic pregnancy; Purple arrow – Embryo; Yellow arrow – Free fluid in the pouch of Douglas).

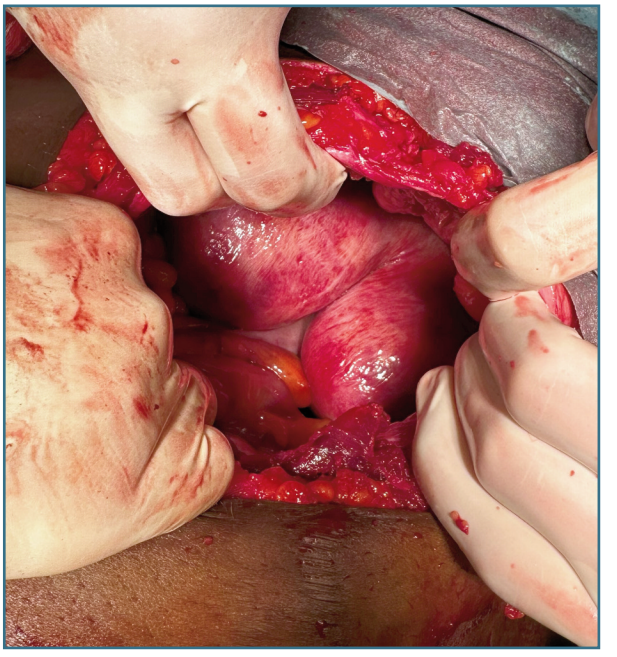

FIGURE 2. Exploratory laparotomy showing a bicornuate uterus.

The pregnancy was followed up in the high-risk obstetrics clinic. The only notable complication, during the course of the pregnancy, was mild idiopathic polyhydramnios, diagnosed in the third trimester. This condition, characterized by normal fetal morphology, negative viral serologies, negative indirect Coombs test

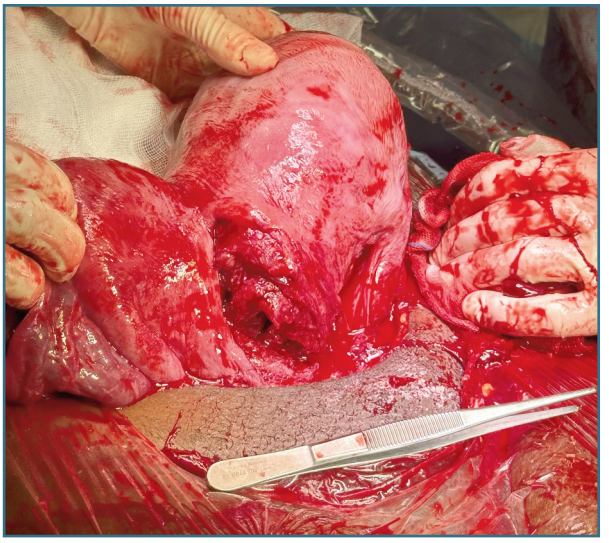

FIGURE 3. Bicornuate uterus at cesarean section: anterior view before hysterorrhaphy.

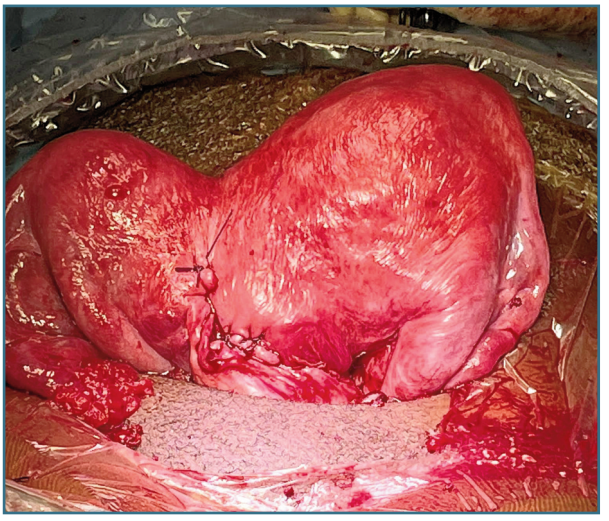

FIGURE 4. Bicornuate uterus at cesarean section: anterior view after hysterorrhaphy.

and no criteria for gestational diabetes, resolved spontaneously before term.

At 38 weeks and 5 days of gestation, an elective lower-segment transverse cesarean section was performed due to breech presentation (Figures 3-5). A baby was delivered from the left hemicavity, weighing 3390 g, with Apgar scores of 9, 10 and 10 at 1, 5, and 10 minutes, respectively. The procedure was complicated by postpartum hemorrhage secondary to uterine atony and laceration of the left angle of the hysterotomy. Hemostasis was achieved successfully with the administration of uterotonic agents (oxytocin and sublingual misoprostol) and hemostatic sutures. Aside from the expected postpartum anemia, the patient experienced no further complications and was discharged in good health together with her healthy baby.

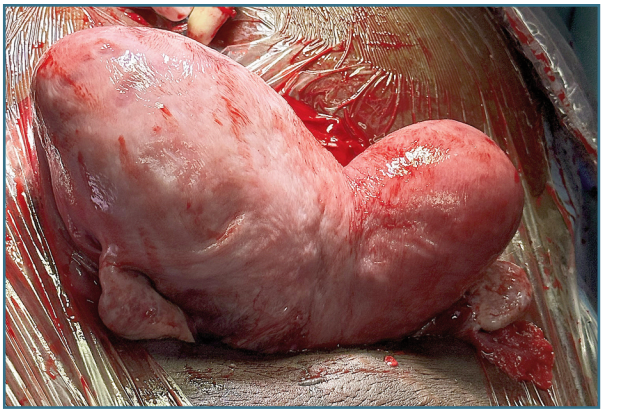

FIGURE 5. Bicornuate uterus at cesarean section: posterior view.

Postpartum abdominal-pelvic computed tomography (CT) was performed to rule out surgical complications and evaluate potential urinary tract anomalies.

DISCUSSION

The female reproductive tract develops through fusion of the Müllerian ducts between the 6th and 14th weeks of gestation. Embryologically, the Mullerian ducts fuse to form the fallopian tubes, the uterus and the upper two-thirds of the vagina. The lower third of the vagina originates from the urogenital sinus, which forms two solid evaginations, the sinovaginal bulbs, that fuse with the caudal portions of the mullerian ducts[1].

Defective lateral fusion of the Müllerian ducts is the primary defect responsible for a bicornuate uterus. This results from partial fusion of the ducts, leading to varying degrees of separation of the uterine horns, from complete to minimal division[2]. The prevalence of bicornuate uterus among patients with normal reproductive outcomes is approximately 5%[3], which places our patient within the spectrum of women who can achieve successful pregnancies despite this anomaly.

Given the shared embryological origin of the urinary and reproductive tracts, renal anomalies are present in up to 30% of patients with Müllerian anomalies[4]. In concordance with this association, postpartum CT of our patient identified left renal agenesis. This finding underscores the importance of incorporating renal imaging in the evaluation of patients with uterine anomalies, typically performed through renal ultrasound. In this case CT was chosen to rule out postpartum complications, such as pelvic hematoma, particularly in the context of significant hemorrhage, hysterotomy laceration, and postpartum anemia.

The two most widely applied classification systems are those of the European Society of Human Reproduction and Embryology with the European Society for Gynaecological Endoscopy (ESHRE/ESGE)[5] and the updated American Society for Reproductive Medicine (ASRM) Müllerian Anomalies Classification[6].

The ESHRE/ESGE system defines anomalies according to anatomical deviations stemming from their common embryological origin, categorizing them into seven major uterine anomalies, and classifies cervical and vaginal anomalies as additional independent subclasses[5]. Notably, some studies suggested that the prevalence of septate uterus has been overestimated when assessed with this classification, as it relies on the internal indentation-to-wall-thickness ratio without requiring external fundal assessment, raising concerns about diagnostic accuracy and consistency[7].

The new ASRM 2021 classification, which revised the 1988 American Fertility Society system, expanded to nine distinct groups: Müllerian agenesis, cervical agenesis, unicornuate uterus, uterus didelphys, bicornuate uterus, septate uterus, transverse vaginal septum and longitudinal vaginal septum[6]. According to this classification, our patient's anomaly corresponds to a complete bicornuate uterus, defined by the presence of a serosal fundal indentation greater than one centimeter, resulting in full separation of the cavities, rather than only the upper third as would be expected in a partial bicornuate uterus[8]. The main differential diagnosis would be a didelphys uterus, however, this was excluded as the patient presents a single cervix and no vaginal septum.

Despite the existence of various classification systems, Müllerian anomalies remain largely misunderstood, due to their rarity and the limited clinical exposure of healthcare providers, often leading to delayed diagnosis, suboptimal management or ongoing issues such as chronic pelvic pain or compromised reproductive function[6]. While many patients are asymptomatic, obstructive anomalies may cause cyclic pelvic pain, amenorrhea, or abnormal bleeding patterns. Patients with a longitudinal vaginal septum may present dyspareunia or persistent bleeding despite tampon use, as separate tampons may be required for each vaginal canal[9].

Two-dimensional (2D) ultrasound is the initial imaging modality of choice for evaluating Müllerian anomalies due to its wide availability, noninvasiveness and cost-effectiveness. Diagnosis is best achieved with 3D ultrasound, which allows simultaneous evaluation of the endometrial cavity and the external uterine contour, essential for differentiating between septate and bicornuate uterus[10]. Magnetic resonance imaging remains the gold standard when 2D and 3D ultrasound findings are inconclusive or limited[10]. Hysterosalpingography provides excellent information about the interior contour of the uterine cavity, but not the external uterine contour, rendering it inadequate for distinguishing between certain anomalies[11].

Women with CUAs, including bicornuate uterus, face increased risks of miscarriage, preterm birth, malpresentation, and cesarean delivery[12]. In our case, breech presentation led to cesarean delivery, aligning with published data on higher incidence of malpresentation[13].

Postoperative complications, particularly those related to uterine anomalies, are well documented, especially during and following cesarean delivery[13]. Our patient experienced postoperative hemorrhage and uterine atony, both of which were successfully managed. These complications underscore the elevated risk associated with surgical interventions in women with bicornuate uterus, corroborating previous reports of increased surgical challenges in such cases[14].

In summary, this case highlights that CUAs, though rare, should be considered in the differential diagnosis of pregnancies located in atypical anatomical positions, as they may mimic ectopic implantation. Maintaining diagnostic vigilance and considering Müllerian anomalies in the clinical decision-making process are

essential to avoid unnecessary surgical procedures and to optimize maternal and fetal outcomes.

REFERENCES

1. Dietrich JE, Millar DM, Quint EH. Obstructive reproductive tract anomalies. J Pediatr Adolesc Gynecol. 2014;27(6):396-402.

2. The American Fertility Society. The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, müllerian anomalies and intrauterine adhesions. Fertil Steril. 1988;49(6): 944-955.

3. Simón C, Martinez L, Pardo F. Müllerian defects in women with normal reproductive outcome. Fertil Steril. 1991;56(6):1192--1193.

4. Oppelt P, von Have M, Paulsen M, Strissel PL, Strick R, Brucker S, et al. Female genital malformations and their associated abnormalities. Fertil Steril. 2007;87(2):335-342.

5. Grimbizis GF, Gordts S, Sardo ADS, Brucker S, Angelis C, Gergolet M, et al. The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies. Hum Reprod. 2013;28(8): 2032-2044.

6. Pfeifer SM, Attaran M, Goldstein J, Lindheim SR, Petrozza JC, Rackow BW, et al. ASRM müllerian anomalies classification. Fertil Steril. 2021;116(5):1238-1252.

7. Jayaprakasan K, Ojha K. Diagnosis of congenital uterine abnormalities: practical considerations. J Clin Med. 2022;11(5):1251.

8. Troiano RN, McCarthy SM. Müllerian duct anomalies: imaging and clinical issues. Radiology. 2004;233(1):19-34.

9. Mazouni C, Girard G, Deter R, Haumonte JB, Blanc B, Bretelle F. Diagnosis of Müllerian anomalies in adults: evaluation of practice. Fertil Steril. 2008;89(1):219-222.

10. Bermejo C, Martínez P, Cantarero R, Pedregosa JP, Barrón E, Labrador E. Three-dimensional ultrasound in the diagnosis of Müllerian duct anomalies and concordance with magnetic resonance imaging. Ultrasound Obstet Gynecol. 2010;35(5):593-601.

11. Pellerito JS, McCarthy SM, Doyle MB, Glickman MG, DeCherney AH. Diagnosis of uterine anomalies: relative accuracy of MR imaging, endovaginal sonography, and hysterosalpingography. Radiology. 1992;183(3):795-800.

12. Hua M, Odibo AO, Longman RE, Macones GA, Roehl KA, Cahill AG. Congenital uterine anomalies and adverse pregnancy outcomes. Am J Obstet Gynecol. 2011;205(6),558.e1-5.

13. Venetis CA, Papadopoulos SP, Campo R, Gordts S, Tarlatzis BC, Grimbizis GF. Clinical implications of congenital uterine anomalies: a meta-analysis of comparative studies. Reprod Biomed Online. 2014;29(6),665-683.

14. Rackow BW, Arici A. Reproductive performance of women with Müllerian anomalies. Curr Opin Obstet Gynecol. 2007;19(3):229-237.

AUTHOR CONTRIBUTIONS

Filipa Ladislau: Contributed to the collecting and analysis of the data, reviewing the literature, and drafting the article.
Ana Elisa Sousa: Contributed by taking intraoperative photographs and revision of the article.
Florência Sepúlveda: Contributed with performance and interpretation of ultrasonography images, supervision and revision of the article, final approval of submitted version.
Marta Luísa Rodrigues: Contributed ensuring the supervision, critical review of the article and final approval of the submitted version.

CONFLICTS OF INTEREST

The authors have no conflicts of interest to declare.

CONSENT

Informed consent was obtained in written form from the patient for publication of this case report.

CORRESPONDENCE TO:

Filipa Ladislau
E-mail: filipa.ladislau@ulsasi.min-saude.pt
https://orcid.org/0009-0003-4036-3068

RECEIVED: 29/06/2025
ACCEPTED: 11/12/2025