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Case Report

Variations in clinical presentation of unicornuate uterus with non-communicating rudimentary horn (class IIB of the American Fertility Society classification)

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ABSTRACT

Objective: The unicornuate uterus is a rare uterine malformation (2.4–13.7% of all uterine malformations (Engmann et al., 2004)) which usually features a rudimentary accessory horn in more than 75% of the cases. Pregnancy in the rudimentary horn is rare, but the complications attached to such pregnancies could be defined as the first clinical manifestation of rudimentary horn.
Case Reports: We hereby describe five cases of unicornuate uterus with rudimentary horn (UUWRH), each one with a different clinical presentation and without any correct preoperative diagnosis, and henceforth reflect on the practical aspects learnt about the differential diagnosis and management of this rare malformation.
Conclusion: Our experience with UUWRH is that perhaps asymptomatic cases are not as rare as reported in medical literature. We highlight the need for a greater awareness of the differential diagnosis of genital malformations and accurate in the exact subtype and their correct treatment.

Introduction

The unicornuate uterus prevalence fluctuates between 2.4% and 13.7% of all uterine malformations [1], although its true incidence rate is unknown. In more than 75% of the cases of unicornuate uterus, we appreciate a contralateral rudimentary horn, and most of these horns bear functioning endometrium which is non-communicating with the other horn. The prevalence of unicornuate uterus with rudimentary horn (UUWRH) in fertile women is approximately 1 in 100,000 [2]. The pregnancy incidence rate in a non-communicating rudimentary horn is approximately 1 in 76,000–150,000 pregnancies and only 14% of the cases are correctly diagnosed before clinical manifestations make themselves apparent [3]. The majority of pregnancies (80–90%) occur in a non-communicating rudimentary horn, although there are exceptional cases of twin pregnancies with one twin in each horn. UUWRH is associated with certain pregnancy risks including preterm delivery, miscarriage and intrauterine fetal death. Gestation in rudimentary horn occurs by sperm or egg transperitoneal migration during the impregnation period. We describe five clinical cases with final diagnosis of UUWRH with different clinical presentations in each case and analyze the problem in the diagnosis and how they should be properly handled. Written consent was obtained from all the women in question.

Case Reports

Case 1 (Fig. 1)

A 31-year-old woman, in her ninth week of pregnancy, was mentioned having lower abdominal pains. Pelvic ultrasound scan showed a 4 cm solid para-adnexal mass with a live embryo leading to a diagnosis of left tubal ectopic pregnancy. At laparoscopy, a UUWRH was diagnosed with pregnancy in the non-communicating rudimentary horn (NCRH). The surgical technique included transection of the round and uteroovarian ligaments and the ipsilateral fallopian tube of the NCRH, which contained the gestational sac.

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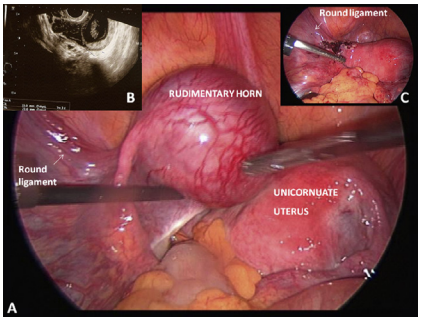

Fig. 1. (Case 1): A: Laparoscopic image of case 1, showing a unicornuate uterus with pregnant rudimentary horn. Note that the link between the two horns is thin. B: Ultrasound image showing the gestation of a 23-mm CRL fetus with positive heartbeat that was initially diagnosed as an ectopic tubal pregnancy. A and C: Note that the round ligament is observed coming out of the rudimentary uterine horn.

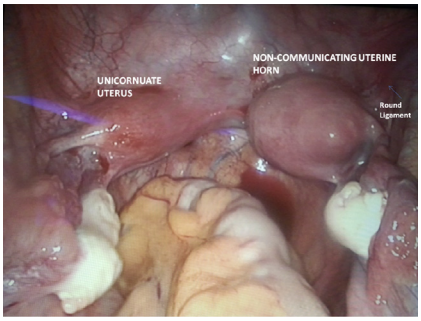

Fig. 2. (Case 2): Laparoscopic image of case 2, showing a unicornuate uterus with rudimentary horn. Note that the link between the two horns is thin.

The extraction was done with suprapubic laparoscopy port expansion and the uterus was not sutured. She remained asymptomatic five years later.

Case 2 (Fig. 2)

An 18 year old nulligravid woman was struck with sudden onset of pain in the left iliac fossa. Ultrasound revealed a suspected uterine malformation. MRI was performed in light of a possible pedunculated fibroid. Diagnostic hysterosalpingography and laparoscopy confirmed a UUWRH with a lack of communication between the unicornuate uterus and rudimentary horn. Hemihysterectomy of the NCRH was subsequently performed (as described for Case 1) and 50 cc of hematometra was drained when opened. In the medical follow-up, she remained asymptomatic and went on using contraceptive methods.

Case 3 (Fig. 3)

A 32 year old nulligravid patient was admitted to hospital with an acute abdomen pain with no vaginal bleeding but severe anemia (hemoglobin: 8.1 g/dl). She denied amenorrhea. Abdominal ultrasound brought to light an empty uterine cavity, hemoperitoneum in the pelvis and an extrauterine 20-week dead fetus. The initial tentative of diagnosis was with respect to abdominal ectopic pregnancy revealed *versus* uterine rupture in a malformed uterus. An emergency laparotomy confirmed haemoperitoneum and an unviable fetus in the abdominal cavity. The placenta was accreta and inserted into the ruptured rudimentary horn of the unicornuate uterus. Both cavities were connected by a 3–4 cm band at the isthmus, without any communication between them. The placenta was removed and the rupture of the rudimentary horn was sutured with one layer of interrupted sutures of vicryl 1/0 and peritonalization was accomplished with vicryl 2/0. NCRH resection was postponed because the patient was hemodynamically unstable but following discharge she failed to re-attend.

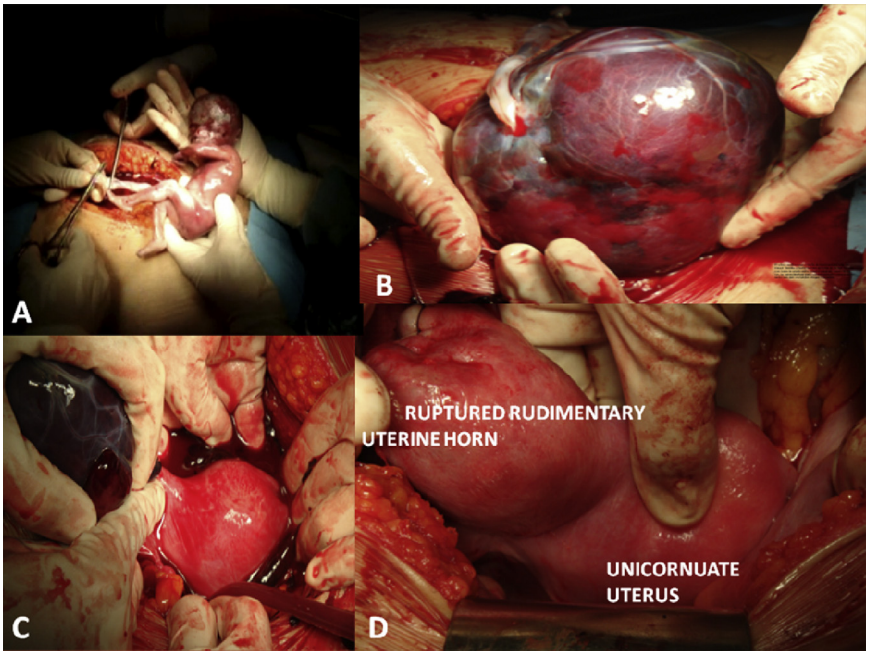

Fig. 3. (Case 3): A: Extraction of the 20-week fetus that was free in the peritoneal cavity. B: Manual extraction of the placenta. C: Manual separation of the placenta accreta with the insertion of the rudimentary uterine horn. D: Suture of the ruptured rudimentary uterine horn. Note that the zone of union with the other horn is wide.

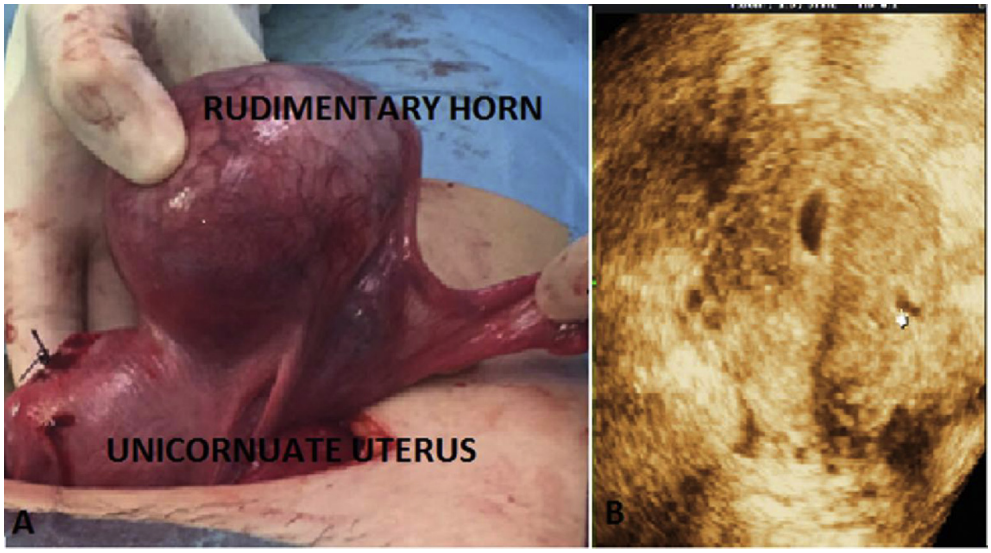

Fig. 4. (Case 4): Perforated unicornuate uterus with a pregnant rudimentary horn. 3D Sonography after surgery showing the unicornuate uterus.

Case 4 (Fig. 4)

A 28 years old woman was diagnosed having a 12-week miscarriage in a bicornuate uterus. After the failure of misoprostol treatment, surgical evacuation was performed on the patient, but there was uterine perforation, henceforth urgent laparotomy was necessary and a unicornuate uterus with a pregnant rudimentary horn was discovered. The perforated uterus was sutured as a result the rudimentary horn was removed. The patient remained asymptomatic one year later.

Case 5 (Fig. 5)

A 38-year-old woman had a miscarriage after 8 weeks of gestation. A malformation uterus was suspected in the ultrasound routine scan of the miscarriage (UUWRH or bicornuate uterus) as oppose to a pedunculated fibroid. MRI was not conclusive. Hysterosalpingography and laparoscopy confirmed the UUWRH with the lack of communication between the unicornuate uterus and rudimentary horn. Hemi-hysterectomy of the NCRH was subsequently performed (as described in the other cases).

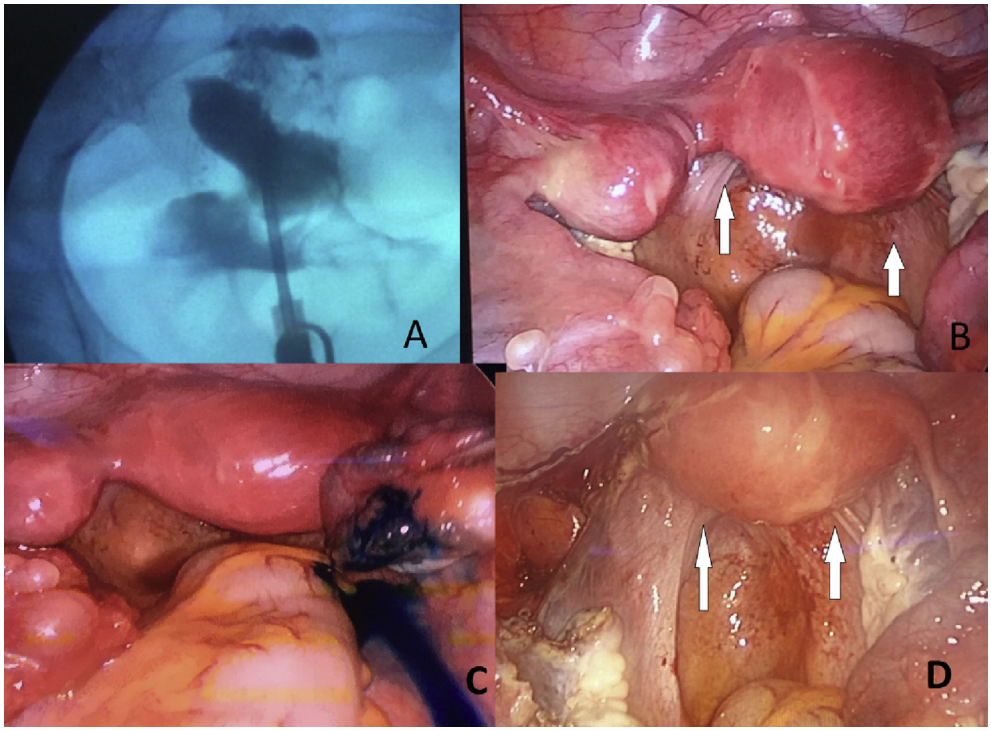

Fig. 5. (Case 5): A: hysterosalpingogram showed the rudimentary left horn was not communicating. B: Laparoscopic image of case 5 showing right unicorn uterus with left rudimentary horn. C: Methylene blue test also showed the passage through the right uterus and the right fallopian tube. D: Laparoscopic view after extirpation of the rudimentary left horn with ipsilateral salpingectomy. Both uterosacral ligaments (white arrows) are inserted into the principal horn.

Table 1
Summary table: symptoms, signs, diagnosis and management of the 5 cases described in the text.

	Symptoms	Clinical management	Diagnosis made by	Management
Case 1	Pain in a first trimester pregnancy	Solid para-adnexal mass in ultrasound; ectopic pregnancy suspected.	Surgery (Laparoscopy)	Excision of rudimentary horn with embryo inside by laparoscopy.
Case 2	Pain in an nulligravid woman	Suspected uterine malformation in ultrasound. Pedunculated fibrioid in MRI.	Hysterosalpingography and laparoscopy	Excision of rudimentary horn by laparoscopy.
Case 3	Acute abdomen in a second trimester pregnancy	Hemoperitoneum and extrauterine death fetus in ultrasound. Urgent laparotomy. Uterine rupture.	Ultrasound	Fetus and placenta extraction. Suture of NCRH by laparotomy.
Case 4	12 weeks miscarriage	Failed misoprostol treatment. Uterine perforation during surgical evacuation	Surgery (Laparotomy)	Perforated uterus was sutured rudimentary horn with embryo inside was removed by laparotomy
Case 5	8 weeks miscarriaged	Hysterosalpingography and laparoscopy	Ultrasound	Rudimentary horn was removed by laparoscopy

Discussion

Diagnosis of asymptomatic UUWRH is a challenge (Table 1), due to low prevalence and a lack of awareness and suspicion among clinicians and radiologists. Often, the patient's medical history is completely normal and symptoms depend on the presence of an obstructive anomaly, with the possibility of developing pain due to hematometra, hematosalpinx or endometriosis. This condition is usually associated with infertility, urinary tract abnormalities, recurrent abortions and adenomyosis. It should be detected during routine gynaecological ultrasound examination (case 2). To analyze the embryological origin of the female genital tract might help understanding the malformation. Cases of non-communicated and cavitated unicornuate uterus (case 2), also present a differential diagnosis with didelphys uterus with right cervico-vaginal agenesis and ipsilateral renal agenesis; despite this, our patient had two normal kidneys. The other entity that yields a differential diagnosis is segmentary atresias in Müllerian malformations [4], which presents as uterus didelphys with segmental atresia of the lower half of one of the hemiuteri, resulting in a non-communicating cavitated uterine horn with hematometra; in this type of malformation, both kidneys are normal (Fig. 6), as in our patient, but in this case, the insertion of the uterosacral ligaments provides the key because if the structure is a uterus didelphys, each uterosacral ligament would be inserted into each uterus, whereas if the structure is unicornuate, both uterosacral ligaments would be inserted into the principal horn (Fig. 5). It is especially important not to perform tubal ligation at the tube ipsilateral to the rudimentary horn because it will trigger obstructive symptoms, as retrograde menstruation from this horn is not possible. When non-communicating, functioning, rudimentary uterine horn is diagnosed, surgery will be needed to remove it [5], specially to prevent pregnancies in this rudimentary horn. Transperitoneal migration of either spermatozoa or fertilized ova from the contralateral side is the hypothesized method of conception in UUWRH. In the case of suspected tubal ectopic pregnancy [6] (case 1) it has been proposed diagnostic ultrasound criteria for gestation in unicornuate uterus [7]: pseudo pattern of asymmetrical bicornuate uterus, absence of visual continuity of the tissue around the gestational sac and the cervix, and the presence of myometrial tissue around the gestational sac. However, ultrasound sensitivity ranges from 26% to 29%. In suspicious cases, an MRI study should be completed, which also serves to diagnose possible associated urinary abnormalities (36%) [8]. It may also be associated with serious pregnancy-related complications and, indeed, this is sometimes the first clinical manifestation of the condition. The differential diagnosis includes a bicornuate uterus [6] and this is particularly relevant because of the need to avoid unnecessary surgical interventions, such as the one that occurred in case 4. If a NCRH is diagnosed, its extirpation with ipsilateral salpingectomy should be performed [5,8] even when found unexpectedly (cases 1 and 4). It is important to emphasize that the risk of rupture of a pregnant rudimentary uterine horn and placenta accreta in the second trimester is very high (case 3) [9] and, therefore, if an early diagnosis is made, excision of the rudimentary horn and ipsilateral tube with or without previous medical treatment (i.e., methotrexate, feticide via potassium chloride or gonadotropin-releasing hormone (GnRH) analogues) is recommended [10]. If the diagnosis is delayed and the patient is asymptomatic, the risks and the small possibility of term pregnancies with live birth should be fully explained [11]. When a uterine rupture occurs, urgent intervention should take place (case 3). If the patient's condition allows it, excision of the rudimentary horn with ipsilateral salpingectomy should be performed immediately. Pregnancies in women after laparoscopic excision of broadly attached rudimentary horns should be considered as high-risk cases [12].

To sum up, our experience with UUWRH is that perhaps asymptomatic cases are not as rare as reported in medical literature because these are the cases we have treated in two years in our hospital. We highlight the need for a greater awareness of the diagnosis of genital malformations and accurate in the exact subtype.

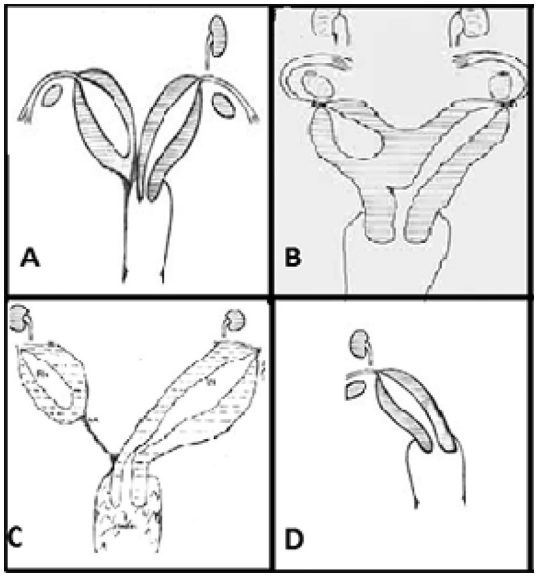

Fig. 6. Differential diagnosis of unicornuate uterus IIB. Importance of renal presence/agenesis: A. Didelphys uterus with right cervico-vaginal agenesis and ipsilateral renal agenesis. B: Unicornuate uterus with rudimentary horn (classification IIB of the AFS); note the presence of both kidneys. C: Didelphys uterus with segmental atresia of the lower half of one of the hemiuteri, resulting in a cavitated non-communicating uterine horn with hematometra; in this type of malformation, both kidneys are normal. D: Unicornuate uterus IID of the AFS (note agenesis of all the derivatives of the urogenital ridge on one side, with corresponding renal agenesis).

Declaration of interest

The authors report no declarations of interest. The authors have not any potential financial and non-financial conflicts of interest.

Précis

We herein describe a series of cases related to unicornuate uterus with rudimentary horn, by explaining its differential diagnosis based on the embryological origin and a discussion of its optimal clinical management.

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References

[1] Engmann L, Schmidt D, Nulsen J, Maier D, Benadiva C. An unusual anatomic variation of a unicornuate uterus with normal external uterine morphology. Fertil Steril 2004;82:950–3.

[2] Handa Y, Hoshi N, Yamada H, Wada S, Kudo M, Tsuda K, et al. Tubal pregnancy in a unicornuate uterus with rudimentary horn: a case report. Fertil Steril 1999;72:354–6.

[3] Arslan T, Bilgic E, Senturk MB, Yucel N. Rudimentary uterine horn pregnancy: a mystery diagnosis. Fertil Steril 2009;92:2037e1–3.

[4] Acién P, Acién M, Fernandez S. Segmentary atresias in Müllerian malformations. Eur J Obstet Gynecol Reprod Biol 2008;141:186–91.

[5] Falcone T, Gidwani G, Paraiso M, Beverly C, Goldberg J. Anatomical variation in the rudimentary horns of a unicornuate uterus: implications for laparoscopic surgery. Hum Reprod 1997;12:263–5.

[6] Nahum GG. Rudimentary uterine horn pregnancy. A case report on surviving twins delivered eight days apart. J Reprod Med 1997;42:525–32.

[7] Tsafrir A, Rojansky N, Sela HY, Gomori JM, Nadjari M. Rudimentary horn pregnancy: first-trimester prerupture sonographic diagnosis and confirmation by magnetic resonance imaging. J Ultrasound Med 2005;24:219–23.

[8] Reichman D, Laufer MR, Robinson BK. Pregnancy outcomes in unicornuate uteri: a review. Fertil Steril 2009;91:1886–94.

[9] Oral B, Guney M, Ozsoy M, Sonal S. Placenta accreta associated with a ruptured pregnant rudimentary uterine horn. Case report and review of the literature. Arch Gynecol Obstet 2001;265:100–2.

[10] Contreras KR, Rothenberg JM, Kominiarek MA, Raff GJ. Hand-assisted laparoscopic management of a midtrimester rudimentary horn pregnancy with placenta increta: a case report and literature review. J Minim Invasive Gynecol 2008;15:644–8.

[11] Ejnes L, Desprez B, Bongain A, Gillet JY. Twin pregnancy in a unicornuate uterus with a rudimentary horn. Gynecol Obstet Fertil J 2003;3:627–8.

[12] Pados G, Tsolakidis D, Athanatos D, Almaloglou K, Nikolaidis N, Tarlatzis B. Reproductive and obstetric outcome after laparoscopic excision of functional, non-communicating broadly attached rudimentary horn: a case series. Eur J Obstet Gynecol Reprod Biol 2014;182:33–7.