Sultan Qaboos University Med J, August 2015, Vol. 15, Iss. 3, pp. e429–432, Epub. 24 Aug 15. doi: 10.18295/squmj.2015.15.03.021.
Submitted 30 Nov 14
Revisions Req. 21 Jan & 4 Mar 15; Revisions Recd. 17 Feb & 31 Mar 15
Accepted 2 Apr 15

ONLINE CASE REPORT

Ruptured Rudimentary Horn Pregnancy Diagnosed by Preoperative Magnetic Resonance Imaging Resulting in Fetal Salvage

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تشخيص تمزق الحمل في قرن الرحم البدائي قبل الجراحة عن طريق الرنين المغناطيسي نتج عنه ولادة طفل حي

سيلجا اروموجام بيلاي، مريم ماثيو، نورين اشرات، انوبام كاكاريا، عاصم قريشي، جاوري فيديانيثان

ABSTRACT: Pregnancy in a rudimentary horn is very rare. The rupture of the horn during pregnancy is an obstetric emergency which can be life-threatening for both the mother and fetus. Preoperative diagnosis of such pregnancies can be challenging and they are usually diagnosed intraoperatively. We report a unique case of a 31-year-old multiparous woman who presented to the Sultan Qaboos University Hospital in Muscat, Oman, in January 2013 at 32 gestational weeks with abdominal pain. Ultrasonography was inconclusive. A rudimentary horn pregnancy was subsequently diagnosed via magnetic resonance imaging (MRI). An emergency laparotomy revealed haemoperitoneum and a ruptured rudimentary horn pregnancy. A live baby with an Apgar score of 2 at one minute and 7 at five minutes was delivered. The rudimentary horn with the placenta *in situ* was excised and a left salpingo-oophorectomy was performed. The postoperative period was uneventful. The authors recommend MRI as an excellent diagnostic modality to confirm rudimentary horn pregnancies and to expedite appropriate management.

Keywords: Uterus, abnormalities; Pregnancy; Magnetic Resonance Imaging; Live Birth; Case Report; Oman.

الملخص: يعتبر الحمل في القرن البدائي من الحالات النادرة، ويعد تمزق هذا القرن الرحمي أثناء الحمل هو أحد الحالات الطارئة والمهددة لحياة الأم والجنين معا. ويعتبر تشخيص مثل هذه الحالات قبل إجراء الجراحة تحديا كبيرا في أغلب الحالات التي يتم تشخيصها أثناء الجراحة فقط. نعرض هنا هذه الحالة المميزة والتي تضم مريضة في الواحدة والثلاثين من عمرها والتي حضرت إلى مستشفى جامعة السلطان قابوس بمسقط–عمان في الثاني من يناير 2013 م، وهي في الأسبوع الثاني والثلاثين من الحمل وتعاني من أوجاع في البطن. ولم يكن فحص الموجات الصوتية حاسما في التشخيص. وتم تشخيص حالة الحمل في القرن البدائي في وقت لاحق عن طريق فحص الرنين المغناطيسي. وتم إجراء عملية طارئة لفتح البطن وجد فيها سائل متراكم في الصفاق، وتمزق القرن البدائي، وولادة طفل حي. وكان معامل ابجار نقطتين بعد دقيقة واحدة، وسبعة بعد خمس دقائق من الولادة. وتم استئصال القرن الرحمي البدائي متضمنا المشيمة. كما استأصلت قناة فالوب والمبيض من الجانب الأيسر. وتخطت المريضة فترة ما بعد الجراحة دون معوقات. توصي المراجع الطبية باستعمال الرنين المغناطيسي باعتباره الوسيلة الأفضل لتأكيد تشخيص حالات الحمل في القرن البدائي ولتسرع من إمكانية اتخاذ التدابير العلاجية الصحيحة.

مفتاح الكلمات: رحم، غير الطبيعية؛ رنين مغناطيسي؛ مولود حي؛ تحرير حالة؛ عمان.

Pregnancy in a rudimentary horn is extremely rare and is reported to occur only in one in 76,000 pregnancies.[1] Rupture of a rudimentary horn pregnancy is a surgical emergency which is usually diagnosed intraoperatively, with high maternal and fetal morbidity and mortality.[1] Fetal salvage is rare as these pregnancies seldom reach the third trimester with a live fetus,[2] emphasising the importance of the current case. The sensitivity of ultrasonography in the diagnosis of uterine anomalies is low, especially in advanced gestation, and can often be missed.[3] A unique case of rudimentary horn pregnancy is presented, which was diagnosed preoperatively via magnetic resonance imaging (MRI) as the ultrasonography was inconclusive. Prompt surgical intervention resulted in a good maternal and fetal outcome.

Case Report

A 31-year-old *gravida* 5 *para* 2 woman of >32 gestational weeks presented to the Department of Emergency Medicine at the Sultan Qaboos University Hospital in Muscat, Oman, in January 2013. She complained of generalised abdominal pain of one week's duration, which had worsened overnight prior to presentation. There was no nausea, vomiting or vaginal bleeding or leaking. She had previously delivered two babies

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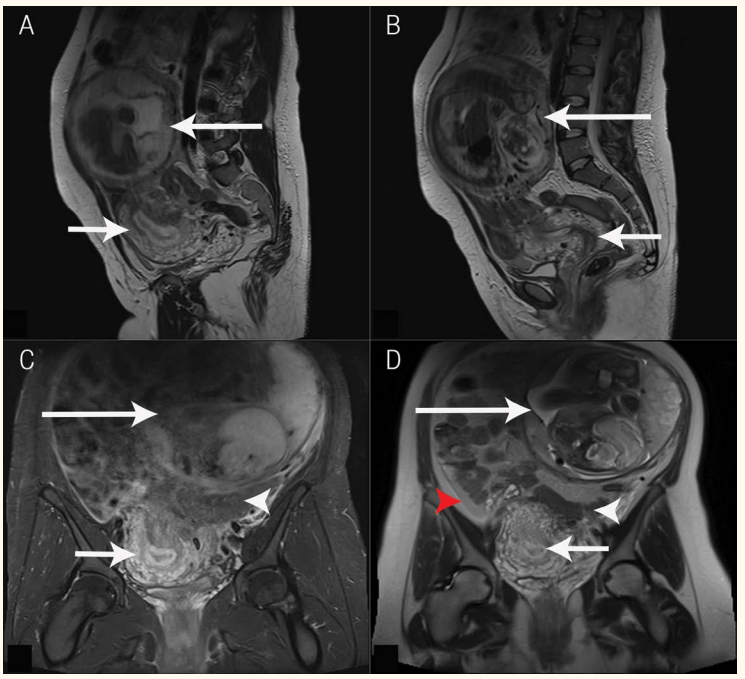

Figure 1A–D: T2-weighted fat-saturated (**A & B**) sagittal and (**C & D**) coronal magnetic resonance imaging (MRI) of the abdomen and pelvis of a pregnant patient. There was evidence of a pregnancy apparently remote from the cervix, with a very thin hypointense wall in the left hypochondrium (long arrows). On the sagittal MRI images, an (**A**) empty normal-looking uterine body (short arrow) could be observed, which was (**B**) connected to the cervix (short arrow). The (**C & D**) bowel loops seemed to be interpositioned between the gestational sac and the cervix (white arrowheads). There was (**D**) free fluid in the abdomen (red arrowhead).

vaginally (both cephalic presentations), followed by two first trimester miscarriages. During her current pregnancy, an ultrasound scan performed at a local health centre at 24 gestational weeks had reported an appropriately grown fetus with no anomalies and a cervical fibroid measuring 4.7 x 6 cm. She gave a vague history of abdominal cramps continuing without relief throughout her pregnancy.

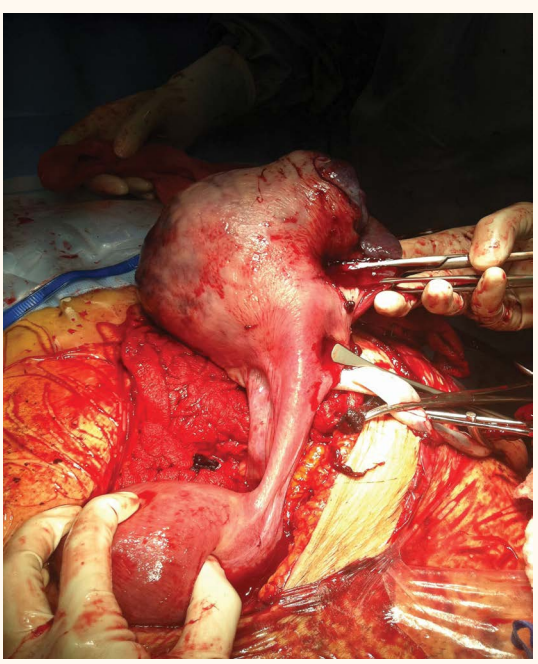

Figure 2: Photograph of the fibrous band connecting the uterus and the rudimentary horn.

On admission, the patient was haemodynamically stable but displayed diffuse abdominal tenderness. The uterine contour appeared globular and the height of the uterus corresponded to the gestation. A cardiotocogram revealed a reactive fetal heart with no uterine contractions. Vaginal examination showed a long closed uterine cervix. An abdominal ultrasound revealed an appropriately grown fetus, the presence of minimal free fluid and a possible cervical fibroid. An urgent MRI was arranged within 24 hours; this gave differentials of an abdominal or rudimentary horn pregnancy [Figure 1]. The cervical cavity was continuous with an empty uterus lying on the right side of the pelvis. The endometrium was thickened and prominent and there was evidence of a gestational sac in the left lumbar region. The sac was surrounded by a thin hypointense rim, without signal intensity features characteristic of the myometrium. It seemed to be connected to the left side of the uterus by a narrow pedicle. There did not seem to be any cervical canal connected to the gestational sac. There was a moderate amount of free fluid in the lower abdomen and pelvis. Both kidneys and the collecting system were normal. An abdominal pregnancy was ruled out due to the

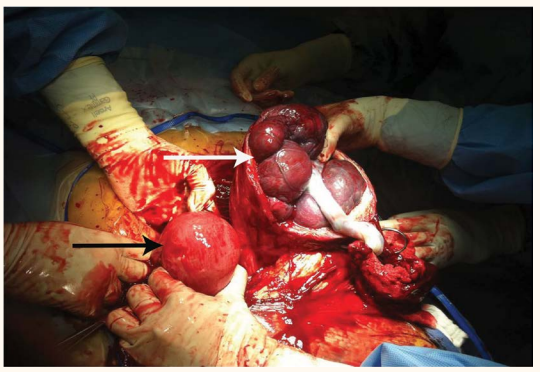

Figure 3: Photograph of the normal empty uterus showing the rudimentary horn (black arrow) with the attached placenta (white arrow).

presence of a rim of uterine muscle surrounding the gestational sac.

Haemoglobin showed a progressive drop from 9 to 7.8 g/dL. An emergency laparotomy was performed, which revealed haemoperitoneum (800 mL) with clots. The pregnancy was located in the left horn with active bleeding from a 1 cm rent in the posterior surface where the placenta was implanted. The unicornuate uterus with the right tube and ovary was normal and was connected to the rudimentary horn by a fibrous band [Figure 2]. A vertical incision was made in the horn and a baby girl weighing 1,510 g with Apgar scores of 2 at one minute and 7 at five minutes was delivered. The left tube was attached superolaterally to the rudimentary horn and the ovary was closely attached to the posterior aspect of the bleeding rudimentary horn. The placenta did not separate and appeared to be adherent to the uterine wall. The rudimentary horn with the placenta was excised [Figure 3] and a left salpingo-oophorectomy was performed. The fibrous band had a *lumen* confirmed by histopathology to be a communicating horn. The fibrous band appeared stretched due to the advanced pregnancy; however, it connected the unicornuate uterus to the rudimentary horn. A pelvic drain was inserted and the abdomen was closed. The estimated blood loss was 2,300 mL, including the haemoperitoneum. The patient received four units each of packed red blood cells, fresh frozen plasma and cryoprecipitate.

The postoperative period was uneventful and the patient was discharged on the sixth postoperative day. The histopathology report of the placenta was consistent with placenta *increta*. The baby was in good health, despite some postural deformities due to the limited space in the horn, and was discharged on the 16th postnatal day.

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

Discussion

Congenital anomalies of the female genital tract are relatively common; however, as they are often asymptomatic, they may go unrecognised. While the exact incidence of genital tract anomalies is difficult to determine, it is estimated to occur in 2–4% of fertile women with normal reproductive outcomes.[4] In patients with adverse pregnancy outcomes, the prevalence could be much higher.

The modified American Fertility Society classification by Rock *et al.* correlates anatomical anomalies with arrested embryologic development.[5] Thus, utero-vaginal anomalies are categorised as dysgenesis disorders or vertical or lateral fusion defects. These are further subdivided into obstructive or non-obstructive forms. Unicornuate uteri account for 5% of all Müllerian anomalies and are an example of a class 3b disorder of lateral fusion.[5] These result from a complete or partial failure of development of one Müllerian duct and an incomplete fusion with the contralateral side. One cavity is normal while the defective side may exhibit varying degrees of disruptive development. Müllerian anomalies are *de novo* congenital abnormalities, although familial aggregates of such conditions have rarely been reported.[6]

The connection of the rudimentary horn with the uterus may be fibrous or fibromuscular. In 72% of cases, there is no communication between the two cavities and pregnancies in the horn occur following the transperitoneal migration of the sperm or of the fertilised ovum.[1] The overall rupture rate in rudimentary horn pregnancies is 50% and the majority of these occur in the second trimester; Nahum *et al.* reported that only 30% of such pregnancies reach term.[1] However, fetal salvage is rare, which emphasises the importance of the current case. Rupture is thought to be a result of an underdeveloped myometrium and a dysfunctional endometrium.[2] The gestational age at which such pregnancies rupture varies according to the thickness of the myometrial wall. Patients with a non-communicating horn with a functional endometrium may have a positive history of dysmenorrhoea and endometriosis.[6] Due to the close embryologic association, evaluation of the renal system is warranted in such patients. Studies show there is approximately a 31–40.5% likelihood of renal anomalies in patients with unicornuate uteri.[6]

Early diagnosis of rudimentary horn pregnancies can be quite challenging. Any ruptures will necessitate immediate surgical intervention leading to higher morbidity and mortality. The maternal mortality rate associated with rudimentary horn pregnancies has improved from 18% in the 19th century to 0.5% in the

present decade, owing to improvements in healthcare and diagnostic facilities.[7] However, Jain *et al*. reported that the fetal salvage rate is still only 2%.[2]

Common diagnostic modalities include hysterosalpingography, combined laparoscopy and hysteroscopy, ultrasonography (preferably three-dimensional) and MRI. An ultrasound scan is used as an initial imaging tool to diagnose such uterine anomalies; however, the sensitivity of this modality is only 26% and this decreases in advanced pregnancies,[3] as was found in the current case. Suggested sonographic criteria by Tsafrir *et al*. include pseudo-patterns of an asymmetrical bicornuate uterus, a lack of visual continuity between the cervical canal and the *lumen* of the pregnant horn and evidence of myometrial tissue around the gestational sac.[8] Typical hypervascularisation of placenta *accreta* may support the diagnosis. Common misdiagnoses with ultrasonography include an ectopic, cornual, intrauterine or abdominal pregnancy. In the case of the current patient, the normal uterus was mistaken for a cervical fibroid in an early pregnancy scan, whereas the scan was inconclusive once the pregnancy was advanced.

MRI has been described as a valuable tool to diagnose uterine anomalies, especially in advanced gestation. A recent systematic literature review evaluated a total of 77 cases of rudimentary horn pregnancy over a 10-year period in order to determine radiological criteria to diagnose rudimentary horn pregnancies antenatally before uterine rupture.[5] Out of the 35 cases which were imaged, MRI scans were performed in only 11 cases, whereas ultrasounds were performed in all cases. In five of the 11 cases with MRI findings, the diagnosis had been missed on the initial ultrasound;[5] this emphasises the significance of MRI in the diagnosis of such anomalies. Almost all of the women in the review were either in their first or second trimester. There was only one case at 28 gestational weeks.[5] The current case reported here involved a rare presentation at 32 weeks of gestation. The low sensitivity of ultrasound scans for such an advanced pregnancy indicated the use of MRI, which helped to expedite the necessary surgical intervention.

Abnormal placentation and adherence is reported frequently in rudimentary horn pregnancies due to the poorly developed musculature, insufficient decidualisation and small size of the horn.[1] Oral *et al*. estimated the prevalence of placenta *accreta* in rudimentary horn pregnancies to be over 10%.[9] The thinning of the myometrium and the invasive nature of the placenta predisposes the rupture and severe bleeding. In the current case, the placenta was found to invade into the myometrial layer and was reported as *increta* on histopathological examination. Kadan *et al*. reported successful laparoscopic excision of a rudimentary horn in an early pregnancy.[10] Surgical intervention and excision of the rudimentary horn should be considered if the diagnosis is confirmed, as the rupture of the horn results in serious maternal morbidity or even mortality.[1]

Conclusion

The advent of new diagnostic techniques like MRI has enabled the preoperative diagnosis of rare conditions—such as rudimentary ruptured horn pregnancies—which were previously diagnosed during laparotomies. In the current case, the use of MRI as the diagnostic modality resulted in fetal salvage; this is rare as this type of pregnancy seldom reaches the third trimester with a live fetus. Excision of a rudimentary horn is recommended, as the rupture of the horn during pregnancy can be life-threatening for both the mother and fetus. A definite preoperative diagnosis helps in counselling the patient and preparing them for an emergency surgical intervention.

References

1. Nahum GG. Rudimentary uterine horn pregnancy: The 20th-century worldwide experience of 588 cases. J Reprod Med 2002; 47:151–63.
2. Jain R, Gami N, Puri M, Trivedi S. A rare case of intact rudimentary horn pregnancy presenting as hemoperitoneum. J Hum Reprod Sci 2010; 3:113–15. doi: 10.4103/0974-120 8.69335.
3. Kanagal DV, Hanumanalu LC. Ruptured rudimentary horn pregnancy at 25 weeks with previous vaginal delivery: A case report. Case Rep Obstet Gynaecol 2012; 2012:985076. doi: 10.1155/2012/985076.
4. Iverson RE Jr, DeCherney AH, Laufer MD. Clinical manifestations and diagnosis of congenital anomalies of the uterus. From: www.uptodate.com/contents/clinical-manifestations-and-diagnosis-of-congenital-anomalies-of-the-uterus Accessed: Apr 2015.
5. Rock JA, Adam RA. Surgery to repair disorders of development. In: Nichols DH, Clarke-Pearson DL, Eds. Gynecologic, Obstetric and Related Surgery. 2nd ed. St Louis, Missouri, USA: Mosby, 2000. Pp. 780–813.
6. Reichman D, Laufer MR, Robinson BK. Pregnancy outcomes in unicornuate uteri: A review. Fertil Steril 2009; 91:1886–94. doi: 10.1016/j.fertnstert.2008.02.163.
7. Siwatch S, Mehra R, Pandher DK, Huria A. Rudimentary horn pregnancy: A 10-year experience and review of literature. Arch Gynecol Obstet 2013; 287:687–95. doi: 10.1007/s00404-012-2625-7.
8. 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.
9. Oral B, Güney M, Ozsoy M, Sönal S. Placenta accreta associated with a ruptured pregnant rudimentary uterine horn: Case report and review of literature. Arch Gynecol Obstet 2001; 265:100–2. doi: 10.1007/s004040000140.
10. Kadan Y, Romano S. Rudimentary horn pregnancy diagnosed by ultrasound and treated by laparoscopy: A case report and review of literature. J Minim Invasive Gynecol 2008; 15:527–30. doi: 10.1016/j.jmig.2008.05.010.