Pregnancy in the Rudimentary Uterine Horn
Case report of an unusual presentation

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Pregnancy in a rudimentary horn is a rare clinical condition with a reported incidence of 1 in 100,000 to 140,000 pregnancies. Usually the diagnosis is missed and may present as an emergency with hemoperitoneum. The standard treatment is the surgical excision of the horn. A gravida 2, para 1 patient presented at 23 weeks' gestation with fetal demise. Repeated failed attempts at induction of labour raised the suspicion of an abnormally located pregnancy which was confirmed by magnetic resonance imaging. She underwent a laparotomy with right rudimentary horn excision. The final diagnosis of a non-communicating rudimentary horn pregnancy was made intraoperatively and was confirmed by histopathology. This case highlights the importance of an early ultrasound in detecting uterine anomalies and the need for high clinical suspicion.

**Keywords:** Uterus, abnormalities; Pregnancy; Ultrasonography; Magnetic Resonance Imaging; Laparotomy; Case Report; Oman.

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**Case Report**

A 24-year-old Omani female presented at 23 weeks’ gestation and was referred to the Royal Hospital, Muscat, with a diagnosis of intrauterine fetal demise, which had been detected on a routine antenatal ultrasound. She reported an absence of fetal movements for the previous two days. There was no history of abdominal pain or vaginal blood loss at any time.

The patient was gravida 2, para 1, with one previous vaginal delivery at term approximately 16 months previously. The third stage had been complicated by a retained placenta which was removed manually under general anaesthesia. There was no significant past medical or surgical history. She had had normal menstrual periods with no history of dysmenorrhea. Her current second pregnancy had previously been uneventful, with a normal fetal anomaly scan at 20 weeks’ gestation.

At admission, the patient’s general condition was good and her vital signs were normal. A physical
Case Report

examination of the abdomen revealed a relaxed, non-tender uterus palpable to the level of the umbilicus. A transabdominal ultrasound showed a single, non-viable, intrauterine fetus with fetal parameters corresponding to 22 weeks' gestation. The amniotic fluid was normal and the placenta was posterior in the upper segment. She was diagnosed with an intrauterine fetal death and the decision was made to induce labour. A complete blood count and coagulation profile were normal. Her blood group was B positive with negative antibody screening. She was screened for TORCH infections (toxoplasmosis, other [syphilis, varicella-zoster and parvovirus B19], rubella, cytomegalovirus and herpes) and was negative for immunoglobulin M antibodies.

A medical induction of labour with misoprostol was attempted. There was no response to the full course of 400 µg of misoprostol given vaginally every four hours for a maximum of five doses. The patient experienced uterine irritability, with minimal vaginal spotting but no cervical changes. A second course of misoprostol was repeated after 48 hours, but with no success. An abdominal ultrasound was repeated and a transvaginal ultrasound was also done. This showed a normal cervix leading to a small, normal, empty uterus just above it. The non-viable pregnancy was seen above and on the right side of the uterus. Pregnancy in a rudimentary horn of the uterus was suspected with a differential diagnosis of an abdominal pregnancy. An MRI of the pelvis confirmed the diagnosis of pregnancy in the rudimentary horn of the uterus, as normal myometrial tissue was seen around the fetus; however, pregnancy in one horn of a bicornuate uterus could not be definitively excluded [Figure 1].

The patient underwent a laparotomy through a midline infra-umbilical incision. The findings included a normal uterus with a normal ovary and fallopian tube on the left side. The pregnancy was in a rudimentary horn on the right side, with a normal ovary and fallopian tube attached to it.

The horn was connected to the uterus just above the cervix by a thick fibrous band. A small incision was made over the pregnant horn and a dead female fetus weighing 450 g was delivered. The horn was then excised, along with the right fallopian tube. The patient lost 200 ml of blood [Figures 2 and 3].

The post-operative period was uneventful and the patient was discharged on the fourth day. A histopathology examination confirmed the diagnosis. There was no infiltration of the chorionic

Figure 1 A & B: Magnetic resonance images showing (A) a normal uterus (white arrows) with the pregnancy above and to the right side, and (B) with a thin myometrium all around the pregnancy (black arrow).

Figure 2: The uterus was exteriorised after the delivery of the fetus. The image shows the right pregnant rudimentary uterine horn (white arrow). As this image was taken after delivering the fetus, the umbilical cord can be seen emerging from the incision.

Figure 3: Image taken after the excision of the right rudimentary horn, which was linked by a fibrous band to the uterus (white arrow).
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Discussion

Uterine anomalies result from the failure of complete fusion of the Müllerian ducts during embryogenesis. The incidence in the general population is estimated to be 4.3%. A unicornean uterus with a rudimentary horn is the rarest anomaly and results from the failure of one of the Müllerian ducts to develop completely and an incomplete fusion with the contralateral side.

The incidence of this anomaly is approximately 0.4%. In the majority (83%) of cases, the rudimentary horn is non-communicating. The anatomical variations of a rudimentary horn serve as the basis for the classification of a unicornean uterus by the American Society of Reproductive Medicine (ASRM). Acién et al. performed a systematic review to analyse the classification systems for uterine anomalies and concluded that an embryological clinical classification system seemed to be the most appropriate. This paper presents a case from class 1, and would be classified as class IIB according to the ASRM [Table 1 and Figure 4].

Pregnancy in a rudimentary horn was first described by Mauriceau and Vassal in 1669. The reported incidence is 1 in 100,000 to 140,000 pregnancies. The most accepted explanation is the transperitoneal migration of the sperm cells or a fertilised ovum. This explanation was supported by the observation of the corpus luteum in the contralateral ovary. It is extremely uncommon for such cases to result in a viable baby. These cases usually result in the rupture of the horn in the second or third trimester, typically between the 10th and 20th week of gestation, although a rupture has been reported at 34 weeks. Only 10% of cases such as these reach term, and the fetal salvage rate is only 2%.

The rupture occurs because of the underdevelopment of the myometrium and a dysfunctional endometrium. A rudimentary horn pregnancy can be further complicated by placenta percreta due to the poorly developed musculature and the small size of the horn; the reported incidence is 11.9%. Placenta percreta can be confirmed by a histopathology examination from as early as seven weeks.

The key for diagnosis prior to the rupture is a high index of clinical suspicion. A history

| Table 1: Embryological-clinical classification for female genito-urinary malformations |
|---------------------------------|---------------------------------------------------------------|
| **1. Unilateral genito-urinary agenesis or hypoplasia** | Cases of unicornean uterus with contralateral renal agenesis due to agenesis or hypoplasia of an entire urogenital ridge. |
| **2. Uterine duplicity (bicornean or didelphys uterus) with a blind hemivagina (or unilateral cervico-vaginal atresia) and ipsilateral renal agenesis** | This includes the Herlyn-Werner-Wunderlich syndrome and there can also be cases of resorption partial of the interstitial septum. |
| **3. Isolated or common uterine or utero-vaginal anomalies** | This includes the anomalies in the Müllerian development processes (also included in the classification of the American Fertility Society) without other associated anomalies; and also the transverse vaginal septum. |
| **4. Accessory uterine masses with an otherwise normal uterus, and other possible gubernaculum dysfunctions** | |
| **5. Anomalies of the urogenital sinus** | These include cases of imperforated hymen, vesico-vaginal fistulas, persistent urogenital sinus, cloacal anomalies and other external gastrointestinal or urinary anomalies. |
| **6. Malformative combinations** | |

villi into the myometrium. An intravenous urogram was done which showed an absent right kidney.
of severe dysmenorrhoea may be a clue for diagnosis. However, the rudimentary horn may be underdeveloped and its endometrium non-functional, so dysmenorrhoea may be absent. A careful pelvic examination in the first trimester showing a deviated uterus with a palpable adnexal mass should provoke suspicion of a Müllerian anomaly. It can be confirmed by an ultrasound or MRI. Tsafrir et al. suggested the following criteria for diagnosing a pregnancy in the rudimentary horn: (1) a pseudo pattern of asymmetrical bicornuate uterus; (2) absent visual continuity between the cervical canal and the lumen of the pregnant horn, and (3) the presence of myometrial tissue surrounding the gestational sac. Ultrasound sensitivity remains only 26%. The enlarging horn with the thinned myometrium can obscure the adjacent anatomical structures and the sensitivity further decreases as the gestation progresses. MRI has proven to be a very useful diagnostic tool.

Approximately 38% of patients have coexisting renal abnormalities. Unilateral renal agenesis is most commonly found; this is always ipsilateral with the rudimentary horn. The differential diagnosis includes a tubal, cornual or intrauterine pregnancy in a bicornuate uterus. Ultrasonographical features may help to reach diagnosis, as in the following examples. A tubal pregnancy will not show a ring of the myometrium surrounding the gestational sac. A variation in the thickness of the myometrium in two horns and a marked distance between them favour the diagnosis of a rudimentary horn pregnancy. The continuity between the endometrium lining the gestational sac and the other uterine horn is typical for a pregnancy in a bicornuate uterus.

In this case, despite the patient’s earlier ultrasound, the diagnosis was initially missed probably due to the advanced gestational age and a lack of clinical suspicion. It was only when the patient failed to respond to repeated attempts to induce labour that an abnormal pregnancy was suspected. The use of misoprostol to terminate a pregnancy in such a case can lead to the rupture of the horn. The MRI was helpful in making the diagnosis of a uterine malformation, although the exact diagnosis and type of attachment was established only by a laparotomy.

Immediate surgery is recommended whenever a diagnosis of a pregnancy in the rudimentary horn is made. The traditional treatment is a laparotomy and the surgical removal of the pregnant horn to prevent rupture and recurrent rudimentary horn pregnancies. In recent years, several cases have been treated successfully by laparoscopies using various techniques. Some authors have described systemic methotrexate administration or feticide with intracardiac potassium chloride as alternatives or adjuncts to surgery in early gestation. Conservative management, until viability is established, has been advocated in selected cases with large myometrial masses. Emergency surgery can be performed at any time. In all such cases, the patient should be informed of the risks of the condition as well as their management options.

**Conclusion**

Despite advances in ultrasound technology, the antenatal diagnosis of a rudimentary horn pregnancy remains difficult for inexperienced physicians. A high index of clinical suspicion for uterine malformations early in the gestation can reduce the mortality rate, along with early intervention. When a rudimentary horn pregnancy is diagnosed, the excision of the horn with ipsilateral salpingectomy is the recommended surgical treatment for the best prognosis. This case highlights the need for high clinical suspicion of this rare condition.

**Declaration**

Written informed consent was obtained from the patient and her husband for the publication of this report, along with the MRI images and photographs.

**References**


