A case of pregnancy in the rudimentary horn of unicornuate uterus

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ABSTRACT

A unicornuate uterus with a rudimentary horn is a uterine anomaly resulting from the incomplete development of one of the Müllerian ducts and an incomplete fusion with the contralateral side. Pregnancy in a rudimentary horn of the uterus is a rare clinical condition with a reported incidence of 1 in 100,000 to 140,000 pregnancies. The majority of cases are diagnosed late, after the rupture has occurred and can present as an emergency with hemoperitonium. The use of ultrasonography helps clinicians to diagnose uterine malformations earlier, which can then be confirmed by a magnetic resonance image (MRI) or a laparoscopy. The standard treatment is the surgical excision of the horn. Our case is a gravida 2 abortion 1 patient presented at 18 weeks’ gestation with ultrasonography suggestive of fetal demise. Repeated failed attempts at induction of labour raised the suspicion of an abnormally located pregnancy. The diagnosis was confirmed at laparotomy. She underwent a laparotomy with right rudimentary horn excision. This case highlights the importance of high clinical suspicion and the need to keep an open mind about the possibilities of uterine anomalies.

Keywords: Uterine anomalies, Rudimentary horn pregnancy, Ultrasound, Laparotomy

INTRODUCTION

A unicornuate uterus with a rudimentary horn is a uterine anomaly resulting from the incomplete development of one of the Müllerian ducts and an incomplete fusion with the contralateral side. 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%.1 A unicornuate 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.2 The anatomical variations of a rudimentary horn serve as the basis for the classification of a unicornuate 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.3 This paper presents a case from class II, and would be classified as class IIB according to the ASRM classification.

CASE REPORT

A 21 years old female presented at 18 weeks’ gestation following a routine antenatal ultrasonography which was suggestive of intrauterine demise and came to clinic at Shastrinagar Hospital Dombivali, India. There was no history of abdominal pain or vaginal blood loss at any time.

The patient was gravida 2, abortion 1, with one previous spontaneous abortion at 1 and half month’s gestation. She did not undergo any undergo any surgical procedure following spontaneous abortion. 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 till now.

At admission, the patient’s general condition was good and her vital signs were normal. A physical examination of the abdomen revealed a relaxed, non-tender uterus palpable to the level of the umbilicus. A trans-abdominal ultrasound showed a single, non-viable, intrauterine fetus with fetal parameters corresponding to 18 weeks’ gestation. The amniotic fluid was normal and the placenta was anteriorly. 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 AB 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.

Figure 1: The left pregnant rudimentary uterine horn after the delivery of the fetus. The fallopian tube is engorged. Unicornuate uterus is seen on the right.

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. Pregnancy in a rudimentary horn of the uterus was suspected with a differential diagnosis of an abdominal pregnancy.

The patient underwent a laparotomy through a transverse suprapubic incision. The findings included a normal uterus with a normal ovary and fallopian tube on the right side. The pregnancy was in a rudimentary horn on the left 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 180 g was delivered. The horn was then excised, along with the left fallopian tube. The left ovary was preserved. The patient lost 150 ml of blood.

The post-operative period was uneventful and the patient was discharged on the fourth post-operative day. Post-operative ultrasound was done which showed the presence of two kidneys. The patient was advised to follow up for intravenous urogram after 6 weeks but she didn’t come.

DISCUSSION

Uterine anomalies result from the failure of complete fusion of the Müllerian ducts during embryogenesis. Maricau and Vassal published the first description of a rudimentary horn pregnancy in 1669, and 600 cases have since been described.¹ Pregnancies occur in both communicating and non-communicating horns in proportion to their relative incidence and are equally likely to rupture.² The rupture occurs because of the underdevelopment of the myometrium and a dysfunctional endometrium.³ 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%.⁴ Placenta percreta can be confirmed by a histopathology examination from as early as seven weeks.⁷

![American Fertility Society classification of müllerian duct anomalies](https://example.com/image)

Figure 2: American Society of Reproductive Medicine (ASR) classification of uterine müllerian anomalies.

Most of the cases have poor fetal outcome as large number of cases present with fetal demise or rupture uterus at 2nd trimester of pregnancy.⁵,⁶ Maternal mortality is low (0.5%).² But morbidity is very high in view of massive blood loss and morbidity adherent placentation.²,⁸,⁹

The prerupture diagnosis of pregnancy in rudimentary horn can drastically reduce maternal mortality and a high index of clinical suspicion is required in such case.³ A pre pregnancy history of severe dysmenorrhea may be a clue in this regard. But dysmenorrhea may be absent if the rudimentary horn in underdeveloped and endometrium
nonfunctional. Routine pelvic examination of the pregnant uterus in the first trimester can also give a clue regarding the diagnosis as a deviated uterus with an adenexal mass. Ultrasound and MRI can help to confirm the diagnosis. But the sensitivity of ultrasound to detect pre-rupture rudimentary horn pregnancy is very poor (30%) probably because of rarity of the diagnosis and non-familiarity of the radiologists about this potentially lethal condition.8,9 Tsafir et al. suggested the following criteria for diagnosing a pregnancy in the rudimentary horn:

- A pseudo pattern of asymmetrical bicornuate uterus
- Absent visual continuity between the cervical canal and the lumen of the pregnant horn
- The presence of myometrial tissue surrounding the gestational sac.1,2,10

Ultrasound sensitivity remains only 26%.6 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.11 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 favors 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.10

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 exact diagnosis and type of attachment was established 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. In all such cases, the patient should be informed of the risks of the condition as well as their management options.11

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REFERENCES
