

## CASE REPORT

### A 30 Weeks, Impending Rupture Ectopic Pregnancy in Rudimentary Horn

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#### ABSTRACT

Rudimentary horn is one of the rarest congenital uterine anomalies. 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. It is difficult to diagnose before surgery and hazardous to maternal life as rupture of pregnant horn result in severe hemoperitoneum. The standard treatment is the surgical excision of the horn.

#### INTRODUCTION

Mullerian anomalies were first classified in 1979 by Buttram and Gibbons and further revised by the American Society of Reproductive Medicine in 1988. Rudimentary horn consists of a relatively normal appearing uterus on one side with a rudimentary horn on the other side. 72–85% of the rudimentary horns are noncommunicating with the cavity.<sup>1</sup> Unicornuate uterus with rudimentary horn may be associated with gynecological and obstetric complications like infertility, endometriosis, hematometra, urinary tract anomalies, abortions, and preterm deliveries. The clinical presentation of this entity is non specific, giving ultrasound a critical role in making the diagnosis.<sup>2</sup> Rupture during pregnancy is the most dreaded complication which can be life threatening to the mother. This case highlights the importance of early ultrasound in detecting uterine anomalies and the need for high clinical suspicion.

#### CASE REPORT

A 23 years old primigravida female presented at 30 weeks of gestation and was referred to GMERS General hospital Gandhinagar (Tertiary care centre) with diagnosis of abdominal pregnancy with fetal demise and hemoperitoneum from private hospital for further management at tertiary care centre.

The patient was primigravida, there was no past medical or surgical history. She had normal menstrual periods with no history of dysmenorrhoea. In her current pregnancy she had history of regular menstrual cycle and her LMP was 18/05/2021, so she was unaware about pregnancy.

Patient has chief complaint of difficulty in passing urine, vomiting and lower abdominal pain for that visited at private hospital. USG was done there suggestive of abdominal pregnancy with fetal demise and hemoperitoneum, for that referred at our tertiary care centre for further management.

On admission patient was in hypovolemic shock with severe pallor and rapid feeble pulse 140/min, her BP was 100/60 mmhg. The abdomen was tense and distended and the uterine size was not made out. Pelvic examination revealed fullness in the fornices with cervical movement tenderness. There was no vaginal bleeding. As the patient was in shock, she was taken for immediate Laparotomy after resuscitation. Her Hb was 4.4gm% at the time of laparotomy.

On Laparotomy there was hemoperitoneum, approximately 600 grams of clot and 1000-1200 ml blood in abdominal cavity noted. A normal uterus with normal ovary and Fallopian tube on right side. The pregnancy was in a rudimentary horn on left side, with a normal ovary and fallopian tube attached to it. The horn was connected to the uterus just above the cervix by thick fibrous band. There was erosion on anterior wall of Left rudimentary horn and bleeding was from that erosion. A small incision was made over the rudimentary horn and dead female fetus of 1160 grams delivered. The placenta was adherent to left rudimentary horn. The horn was then excised along with Left Fallopian tube. After achieving hemostasis abdomen was closed in layers after keeping drain. The patient was transfused with 4 units of PCV and 2 units of FFP. Her post operative recovery was good. She was later investigated for urinary tract anomalies which was found to be absent. The Patient was discharged on

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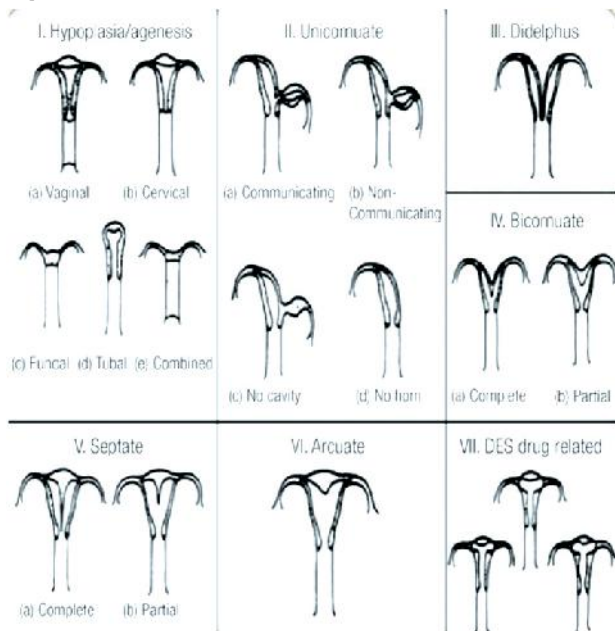
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forth day. A Histopathological examination was suggestive of placenta with mature chorionic villi ; Rudimentary horn attached with placenta showed myometrium with changes of hyperplasia and hypertrophy; No placental anomaly was observed.

### CASE 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%.<sup>3</sup> The incidence of this anomaly is approximately 0.4%.<sup>3</sup> In the majority (83%) of cases, the rudimentary horn is non-communicating.<sup>4</sup> 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). Acien 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.<sup>5</sup> The case in this would be classified as class IIB according to the ASRM Figure I.

**Figure I**



American Society of Reproductive Medicine (ASRM) classification of uterine Müllerian anomalies.<sup>4</sup>

DES = diethylstilbestrol.

Pregnancy in a noncommunicating rudimentary horn occurs through the transperitoneal migration of the spermatozoon or the transperitoneal migration of the fertilized ovum<sup>6</sup>. The first case of uterine rupture associated with rudimentary horn was reported in 1669 by

Mauriceau<sup>7</sup>. The timing of rupture varies from 5 to 35 weeks depending on the horn musculature and its ability to hypertrophy and dilate. 70–90% rupture before 20 weeks and can be catastrophic<sup>8</sup>. As the uterine wall is thicker and more vascular, bleeding is more severe in rudimentary horn pregnancy rupture<sup>9</sup>. Kadan and Romano described rudimentary horn rupture as the most significant threat to pregnancy and a life-threatening situation<sup>10</sup>. The rupture occurs because of the underdevelopment of the myometrium and a dysfunctional endometrium<sup>11</sup>. 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%<sup>11</sup>. Placenta percreta can be confirmed by a histopathology examination from as early as seven weeks<sup>12</sup>.

Early diagnosis of the condition is essential and can be challenging. Ultrasound, hysterosalpingogram, hysteroscopy, laparoscopy, and MRI are diagnostic tools<sup>13</sup>. Fedele et al. have found ultrasonography to be useful in the diagnosis<sup>14</sup>. But the sensitivity of ultrasound is only 26% and sensitivity decreases as the pregnancy advances<sup>15</sup>. It can be missed in inexperienced hands as in present case. Tubal pregnancy, cornual pregnancy, intrauterine pregnancy, and abdominal pregnancy are common sonographic misdiagnosis<sup>16</sup>. There are no definitive clinical criteria to detect this life-threatening condition in case of emergency, and diagnosis can be difficult because the enlarging horn with a thinned myometrium can obscure the adjacent anatomic structures. The key for diagnosis prior to the rupture is a high index of clinical suspicion. A history of severe dysmenorrhoea may be a clue for diagnosis. However, the rudimentary horn may be underdeveloped and its endometrium nonfunctional, 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. Tsafir 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<sup>3,4,17</sup>. Ultrasound sensitivity remains only 26%<sup>11</sup>. The enlarging horn with the thinned myometrium can obscure the adjacent anatomical structures and the sensitivity further decreases as the gestation progresses. In this case, the diagnosis was

**Figure II:- Uterus with Rudimentary horn**



**Figure III:- A dead female child**



initially missed probably due to the advanced gestational age and a lack of clinical suspicion. MRI has proven to be a very useful diagnostic tool. Renal anomalies are found in 36% of cases<sup>15</sup>; hence it is mandatory to further assess these women.

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<sup>12</sup>. Some authors have described systemic methotrexate administration or feticide with intracardiac potassium chloride as alternatives or adjuncts to surgery in early gestation<sup>12</sup>. 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.<sup>4</sup>

### CONCLUSION

Despite advances in ultrasound and other diagnostic modalities, prenatal diagnosis remains elusive, with confirmatory diagnosis being laparotomy. The diagnosis can be missed in ultrasound especially in inexperienced hands. A high index of clinical suspicion for uterine malformations early in the gestation can reduce the mortality rate, along with early intervention. Timely resuscitation, surgery, and blood transfusion are needed to save the patient. 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.

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