Unruptured Second Trimester Pregnancy in a Rudimentary Horn of Unicornuate Uterus: A Diagnostic Dilemma

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Abstract: Unicornuate uterus with rudimentary horn is a rare mullerian anomaly of the uterus and pregnancy in a non-communicating rudimentary horn is very rare. In most of the cases, it is ruptured before diagnosis in first and second trimester due to poor distensibility of the myometrium. Catastrophic hemorrhage results due to rupture which further results in increased maternal and perinatal mortality and morbidity. Diagnosis before rupture is a challenge due to diagnostic dilemma. Expertise in ultrasonography is required and early resort to surgical management is lifesaving in such cases. Here is a case of unruptured ectopic pregnancy in the rudimentary horn of gestation 17 weeks which was diagnosed by ultrasound and MRI and patient was operated before any symptoms appeared.

Keywords: Unruptured, Unicornuate uterus, Non-communicating Rudimentary Horn, Mullerian duct anomaly, Pregnancy.

INTRODUCTION
Mullerian duct anomalies are encountered infrequently in routine gynecological practice. The prevalence of congenital uterine anomalies in the general population is 6.7% while that of unicornuate uterus with or without rudimentary horn is 0.4% [1]. Unicornuate uterus with a rudimentary horn results from the defective fusion of the malformed duct with the contra-lateral duct. A fibrous or fibro-muscular band usually connects the horns of the ducts but in 80 - 90% of cases there is no communication [2].

Rudimentary horn pregnancy is very rare and occurs in approximately 1/76,000 to 1/140,000 pregnancies [1, 3-5]. It occurs following transperitoneal migration of sperm or zygote from the contralateral side. But, it is associated with high rate of morbidity and mortality as a sequence of rudimentary horn rupture and massive hemoperitoneum.

Incidence of uterine rupture is observed in 90% of cases, mostly in the second trimester [6]. Only 8% of rudimentary horn pregnancies are diagnosed before the symptoms appear [7,8]. Pre-rupture diagnosis is possible with high index of suspicion in the early pregnancy.

Various imaging modalities have been used in the diagnosis and evaluation of mullerian duct anomalies. Ultrasonography (USG) and hysterosalpingography may suggest a Mullerian duct anomaly, further confirmation by 3-D ultrasound, MRI, hysteroscopy and laparoscopy is required. MRI is especially useful in preoperative assessment when the ultrasonography is not able to confirm or rule out an ectopic pregnancy in rudimentary horn [1].

The objective of this report is to present a case of unruptured non-communicating rudimentary horn of a unicornuate uterus in a woman at 17 weeks of gestation, highlighting the importance of ultrasound and MRI in the diagnosis and the surgical management.

CASE REPORT
A 23 years old primigravidae female came for routine booking at our institution with amenorrhea of four months. On per abdomen examination uterus was deviated to left side and was of 16 weeks size. There was no significant past or family history. She was advised ultrasound to rule out any congenital anomalies and it was done at period of gestation of 17 weeks.

On ultrasound, uterus was found to be empty and measured 7.1*8.8*5.2 cm. There was a gestational sac with a dead fetus corresponding to gestational age of 15th weeks on left side of uterus (Fig. 1). The differential diagnosis of pregnancy in rudimentary horn of unicornuate uterus and intra-abdominal pregnancy was made. With such anticipation, second opinion ultrasound and magnetic resonance imaging (MRI) was

advised which confirmed the diagnosis of pregnancy in non-communicating horn of unicornuate uterus.

**Fig. 1: Ultrasound showing gestation sac in rudimentary horn. Uterus was empty.**

Routine investigations were done and patient was kept for elective laprotomy after arranging for blood. Preoperative consent was taken and antibiotic were given on the day of surgery. Abdomen was opened with midline infraumblical vertical incision. On opening abdomen, there was evidence of unicornuate uterus with a rudimentary horn on left side which was globular in shape with a smooth surface and enlarged to size of 10*8 cm (Fig. 2).

**Fig. 2: Unicornuate uterus with intact left rudimentary horn. Tubes and ovaries were grossly normal.**

It was attached to uterus with a fibrous band and left tube and ovary were attached to the horn. There was no communication of the horn with the uterus and cervix. Bilateral tubes and ovaries were grossly normal in size and shape. The pregnant rudimentary horn on left side was excised intact and salpingectomy was done on the same side. On cut section of the horn, there was a gestation sac which had fully formed dead macerated fetus with a grossly normal looking placenta (Fig. 3). Post operative period was uneventful and patient was discharged on fourth post operative day.
DISCUSSION

Pregnancy in the rudimentary horn is a very rare clinical presentation. Pregnancy in a non-communicating uterine horn is possible by intraperitoneal sperm and ovum transmigration or contralateral tubal pick-up of the fertilized ovum within the peritoneal cavity [6]. Rudimentary horn has variable degrees of thickness of musculature, dysfunctional endometrium and poor distensibility of the myometrium which results in the rupture of the rudimentary horn and this is the most dreaded and life threatening condition for the mother. This complication is usually seen in the 2nd trimester and results in hemoperitoneum and hemorrhagic shock which requires immediate laprotomy and blood transfusions [2].

Mullerian duct anomalies may be diagnosed in the prepregnancy workup for complaints of dysmenorrhea, endometriosis, and infertility, and for various pregnancy complications like recurrent miscarriages, preterm labor and malpresentations [6]. High index of suspicion should be kept in teenagers presenting with dysmenorrhea and every effort should be done to exclude the condition by conducting relevant investigations in suspected cases. If in patients presenting with infertility, hysterosalpingography shows that the uterus is deviated to one side and there is unilateral tubal block, this condition should be strongly suspected [2]. Bimanual palpation of a mass extending outside the uterine angle i.e., Baart de la faille”s sign or displacement of fundus to contra-lateral side with rotation of uterus and elevation of the affected horn (Ruge Simon Syndrome) and deviation of uterus to one side with adnexal mass in pregnancy may indicate rudimentary horn [9]. Literatures show very low preclinical (8%) and preoperative (29%) detection rates [7, 11].

A diagnosis of rudimentary horn prior to the rupture is unusual and challenging, but it could be made with ultrasonography and MRI. Ultrasound has a sensitivity of 33.3% for diagnosing this anomaly, and sensitivity reduces with advancing pregnancy [6]. Sonographic diagnostic criteria suggested by Tsafir are — (a) Presence of pseudopattern of an asymmetrical bicornuate uterus, (b) Absent visual continuity between the cervical canal and the lumen of the pregnant horn, (c) Presence of myometrial tissue surrounding the gestational sac [7, 10]. Magnetic resonance imaging has proven to be a useful, noninvasive tool for the diagnosis of Mullerian abnormalities.

Chances of placental adherence are increased due to poorly developed musculature, scant decidualization and small size of the horn. Magnetic resonance imaging is a useful preoperative tool for the diagnosis of pregnancy in the rudimentary horn and any abnormal placentation [6].

A rudimentary horn pregnancy can never be delivered vaginally. The mode of delivery is laparotomy, either due to ruptured horn or if the pregnancy continues as abdominal post-rupture [6]. The definitive management for both ruptured and unruptured rudimentary horn pregnancy is surgical removal of the pregnant horn [15]. We performed an elective laparotomy and successfully excised the intact unruptured rudimentary horn. Ipsilateral salpingectomy was also done alongwith to prevent further occurrence of tubal ectopic pregnancy.

Laparoscopic excision of the horn prior to rupture has successfully been reported, but at earlier gestational age [12]. Management with methotrexate during early pregnancy in the rudimentary horn has also been reported [13]. Moreover, conservative management can be done till the period of viability in selected cases with larger myometrial mass, if emergency surgery can be performed anytime and the patients are well-informed [14].

Term pregnancies with a live fetus have been rarely reported in this unusual and life-threatening condition [7]. Neonatal survival in rudimentary horn pregnancy is poor, occurring in only 11% cases [4].

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The associated urologic anomalies are reported to be as high as 50-80% due to close approximation of the two systems, and must be diagnosed either at laparotomy, by palpation, or postnatally by magnetic resonance imaging or intravenous urogram [6].

CONCLUSION

Non-communicating rudimentary uterine horn pregnancy is a rare entity associated with life threatening consequences for the mother. Early diagnosis and early interventions will avoid maternal morbidity and mortality. But, the antenatal diagnosis of this condition remains difficult. If ultrasonography remains inconclusive, the use of magnetic resonance imaging is suggested.

REFERENCES