A Case of Mid Trimester Pre - Rupture Non - Communicating Rudimentary Horn Pregnancy: Clinical Presentation and Management

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Abstract: Introduction: Pregnancy in rudimentary horn of the uterus is a type of ectopic pregnancy diagnosed most often at the rupture of the rudimentary horn, which is the most dreaded and life - threatening complication. Here we present a rare case of pre - rupture diagnosis and management of rudimentary horn pregnancy. Case presentation: We present a case of a 26y PGR at 24weeks gestational age with rudimentary horn pregnancy with dull abdominal pain prior to rupture of rudimentary horn. Surgical management was done and rudimentary horn with ipsilateral tube was removed. The patient was stable post - op and discharged of day – 7 Discussion: Rudimentary horn pregnancy is a rare ectopic pregnancy. Diagnosis is difficult clinically, even with diagnostic imaging modalities. Identifying both cornua systematically in all patients increases the detection rate. The absence of continuity between the gestational sac’s lumen and the cervical canal on imaging is an important finding. Conclusion: High suspicion can aid in timely diagnosis and management at an early gestation and can prevent serious morbidities and mortality.

Keywords: Rudimentary horn, rupture, ectopic, unicornuate

1. Introduction

The unicornuate uterus is formed as a result of failure in the development and elongation of the müllerian duct during embryogenesis. As a result of this defect, it usually results in a functional uterus, normal cervix, and fallopian tube with agenesis on the contralateral side or an abnormal müllerian development such as a rudimentary horn [¹]. The rudimentary horn may or may not be communicating with the normal cavity. It may have a functional endometrial cavity or it may be solid [²]. This malformation presents with different complications through a woman’s reproductive life starting from hematometra, endometriosis to the most dreaded catastrophic complication of rupture of gravid rudimentary horn [³].

Pregnancy is rare in the rudimentary horn and is a form of ectopic pregnancy with an incidence of between 1/ 76.000 to 1/140.000 [⁴]. Pregnancy in rudimentary horns can reach different gestational weeks in patients, depending on the muscular structures of the rudimentary horns. The patient usually presents to the hospital with clinical symptoms of abdominal hemorrhage and acute abdominal pain due to rupture of the uterine wall in the second trimester [⁵]. Pregnancy in a non - communicating rudimentary horn occurs by transperitoneal migration of sperms or the fertilized ovum [⁶]. Approximately 40% of women with RHP are asymptomatic, and abdominal pain is the most common clinical symptom [⁷]. The maternal and fetal prognosis in unrecognized rudimentary horn ectopic pregnancy is poor, with an average neonatal survival rate of 6% and rate of uterine rupture close to 80% [⁸]. Early surgical intervention with removal of the rudimentary horn containing the gestational sac along with the ipsilateral fallopian tube has been recommended for cases of rudimentary horn pregnancy [²].

We report a case of ruptured non - communicating rudimentary horn pregnancy in shock at 24 weeks gestation.

2. Case Report

A 26years PGR presented with dull abdominal pain from 2 days at 24 weeks period of gestation. Her vitals were stable and there were no features of shock. On clinical examination, the uterus was deviated to one side and the cervix was long and pulled up behind the cervix. Fetal heart sound was not audible. There was a suspicion of rudimentary horn pregnancy, for which ultrasound was done. Ultrasound showed a normally grown fetus in a cavity in the right fundal portion of the uterus, separated by a septum from the larger uterine cavity. There was myometrial tissue all around the gestational sac. No communication was seen between the uterus and the cavity that contained the fetus.

With the first possibility of rudimentary horn pregnancy, patient was taken for exploratory laparotomy proceed. Exploratory laparotomy with excision of rudimentary horn along with ipsilateral fallopian tube was done. Intraoperatively, the uterus was unicornuate with pregnancy in the right horn of the uterus. The myometrium of the rudimentary horn was thin and membranous. Both the ovaries and fallopian tubes were normal grossly. Resection

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of the rudimentary horn along with the product of conception was done. No communication was found between the two horns.

On cut section of the rudimentary horn after excision, a single gestational sac fetus was found along with placenta as shown in image C and D.

Postoperative recovery was good and she was discharged on post operative day - 7.

Images A - D shows (A) Intraoperative image showing uterus and right rudimentary horn with pregnancy with bilateral normal ovaries. (B) Excised rudimentary horn with fallopian tube. (C) Cut section of rudimentary horn showing single gestational sac with fetus and placenta. (D) Cut section of rudimentary horn with fetus removed showing placenta and umbilical cord.

3. Discussion

In the uterus with rudimentary horns, the rudimentary part is usually located on the right side. The reason for this is that the left müllerian canal moves caudally than right part [8]. The rudimentary horn, which is not connected to the cavity containing uterus, can cause retrograde menstruation, causing hemosalpinx and endometriosis from the same sided fallopian tubes [9]. The literature reports that 80 - 90% of rudimentary horn pregnancies rupture by the second trimester and 10% reach till term with a 2% salvage rate by cesarean section [10]. The average duration of gestation is estimated at 21.5 weeks [3]. In our case too, the patient reported at 24 weeks with rupture of rudimentary horn.

The first reported case of uterine rupture associated with a rudimentary horn was in 1699 by Mauriceau and Vassal. Maternal mortality rate before 1900 was reported 47.6%. Rupture of the horn is still common but no case of maternal death has been reported since 1960 [11].

The rudimentary horn is hard to identify, especially if not suspected, and may be misdiagnosed as pelvic mass or cervix. When pregnancy develops in a rudimentary horn, the absence of continuity between the gestational sac's lumen and the cervical canal on ultrasound is an important finding. Tsafir et al outlined a set of criteria for diagnosis of pregnancy in a rudimentary horn. They are: 1. A pseudo pattern of asymmetrical bicornuate uterus. 2. Absent visual continuity between tissue surrounding the gestational sac and the uterine cavity. 3. Presence of myometrial tissue surrounding the gestational sac [12].

Laparoscopy combined with hysteroscopy forms the gold standard of diagnosis. Sensitivity of USG is only 26% and the sensitivity decreases as the pregnancy advances. Tubal pregnancy, cornual pregnancy, intruterine pregnancy and abdominal pregnancy are common sonographic misdiagnosis [13].

MRI allows accurate classification of unicorneutre uterus. At MRI, the small, curved unicorneutre uterus is typically displaced off midline. This appearance is named “Banana” configuration. It has normal myometrial zonal anatomy, with normal endometrial - to - myometrial width and ratio. The appearance of the rudimentary horn varies by subtype. If there is no endometrium present, zonal anatomy is absent and the entire horn may demonstrate diffuse low signal intensity. A rudimentary horn without endometrium and the absent rudimentary horn subtype present minimal risk and do not usually require surgical intervention. However, the
presence of endometrium in a rudimentary horn is an important finding and should be reported. A non-communicating rudimentary horn with endometrium may manifest as a large uterine mass. If endometrial tissue is present, there may be preserved zonal anatomy [14].

In our case the patient had no previous visit to any hospital and no ultrasound was conducted prior to this. During her current visit, the patient presented with hypovolemic shock and laparotomy was done keeping in mind a high suspicion of ruptured rudimentary horn pregnancy.

During surgery, same - sided salpingectomy - to avoid following ectopic pregnancies -, excising of the rudimentary horn is recommended. Oophorectomy is not recommended. Laparoscopy can be performed in non - ruptured cases [15]. However, laparotomy was preferred because our patient presented with ruptured - rudimentary horn, and was hemodynamically unstable. Advanced gestational age also favored our decision of laparotomy.

This anomaly is highly associated with - mostly same sided - urinary tract abnormalities (incidence found to be 36%). The incidence of major renal anomalies associated with incomplete uterine duplication with non - communicating rudimentary horn varies between 31% and 100%. The most common anomaly is renal agenesis on the same side with non - communicating rudimentary horn, while the same - sided pelvic kidney is 2nd most common one [16]. In our case, no urinary tract anomalies were found.

4. Conclusion
Triad of dysmenorrhea beginning at menarche, increasing severity of dysmenorrhea and unilateral pelvic mass is a strong evidence of congenital Mullerian anomaly. MRI and diagnostic laparoscopy are tools to definitive diagnosis. Surgical removal of the rudimentary horn is the mainstay of management to prevent endometriosis, future ectopic pregnancy and maintain future fertility. Laparoscopic resection of rudimentary horn pre - rupture in expert hands is less morbid with equivalent results.

References