

Angular vs. interstitial pregnancy: A case report highlighting diagnostic nuances with stark management differences

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ABSTRACT

Background: In the literature, the terms “angular”, “interstitial” and “cornual” have often been inappropriately interchanged. The consequence is under-recognition of their differences as well as inaccurate imaging guidelines which do not reliably distinguish them as distinct entities. Angular pregnancies should be considered viable and may be managed to term.

Case: A woman at 7w5d was transferred for surgical management of a presumed interstitial ectopic pregnancy. Sonography and MRI confirmed an eccentric fundal pregnancy with a thin myometrial mantle of 2–5 mm; the diagnosis of interstitial pregnancy was favored. Upon laparoscopy, the round ligament was displaced lateral to the pregnancy bulge and the diagnosis of angular pregnancy was thus apparent. The pregnancy was continued to term and delivered via repeat cesarean section without incident.

Conclusion: Angular and interstitial pregnancies are different entities which cannot always be reliably distinguished via imaging alone. Diagnostic laparoscopy may be a final step in determining pregnancy location. Angular pregnancies should be considered potentially viable and may be managed to term.

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1. Introduction

Despite being described over a century ago [1], “angular pregnancy” remains an under-recognized entity and is often confused with the terms “cornual pregnancy” and “interstitial pregnancy”. Understanding the differences between the terms, however, is paramount, as their risks, potential for viability, and management options differ.

An angular pregnancy is an eccentric *intrauterine* pregnancy with implantation of the embryo in the lateral superior angle of the uterine cavity. It results in asymmetric enlargement of the uterus and lateral displacement of the round ligament [2]. Angular pregnancy is seldom discussed in the medical literature, with <100 cases reported, and is perceived to be rare [3]. An interstitial pregnancy is an ectopic, or *extrauterine*, pregnancy resulting when implantation occurs within the myometrial fallopian tube, or interstitium. It results in no lateral displacement of the round ligament [2]. Interstitial pregnancies comprise 2–4% of all ectopic pregnancies [4]. A cornual pregnancy historically describes the *intrauterine* fundal implantation within the anomalous bicornuate or septate uterus.

Despite these distinct definitions, examples from the current literature demonstrate inconsistent and inappropriate use, and the term “cornual” has been applied to interstitial or angular pregnancies [5,6]. Even in authoritative textbooks, the terms are often interchanged or

the concept of angular pregnancy is completely ignored. Prior to the 2014 edition of Williams' Obstetrics, the terms angular, cornual, and interstitial pregnancies were not distinguished from one another [7]. This laxity in terminology has contributed to difficulties in developing ultrasonographic diagnostic criteria to distinguish these pregnancy locations from one another and may have led to an underappreciation of the incidence of angular pregnancies.

2. Case Report

A 33yo G3P1101 was transferred at 7w5d for management of a presumed interstitial ectopic pregnancy. The pregnancy was eccentrically located in the right cornua with an overlying myometrial thickness measuring first 5 mm and then 3.5 mm on ultrasounds performed at 6 and 7wks estimated gestational age. Her prior pregnancies included a full-term NSVD followed by a 36wk IUFD of mono/di twins delivered via cesarean section.

The patient reported light spotting but no abdominal pain. Repeat vaginal sonogram demonstrated a pregnancy in the right cornual region consistent with 7w5d gestation with a heart rate of 170 bpm. Blood was noted within the endometrial cavity. The over-riding myometrial mantle was 5–8 mm and no interstitial line sign could definitively be seen (see Fig. 1). Non-contrast MRI revealed a mantle measurement as little as 2 mm (Figs. 2 and 3). The diagnosis of an interstitial pregnancy was favored.

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Fig. 1. Transverse transvaginal sonography of 7w5d gestation and line marking the right myometrial mantle thickness of only 5 mm.

The decision was made to proceed with termination. The patient emphasized her strong desire to maintain any viable pregnancy, regardless of the risk of future morbidity, if evidence was found to support an intrauterine pregnancy. A combined laparoscopic and hysteroscopic approach for termination was planned; laparoscopy to confirm the diagnosis, temporarily occlude the uterine blood supply, and monitor for perforation that may occur with our preferred route for hysteroscopic evacuation. The patient additionally gave consent for a laparoscopic cornual resection or hysterectomy, if needed, and surgery was performed the following day. The laparoscopic view of the uterus was remarkably normal (Fig. 4). A slight bulge could be seen at the right cornua which was palpably softer than the rest of the uterus. There were no abnormal vascular changes and the extrinsic right tube appeared normal (Fig. 5). Importantly, the right round ligament insertion remained lateral to the pregnancy (Fig. 5). An angular pregnancy, a lesser known and potentially viable form of intrauterine pregnancy, was thus diagnosed. The procedure was discontinued given the patient's wishes to maintain any potential viable pregnancy.

At her first trimester screen, the myometrial mantle remained thin at 5 mm. She had a normal level II ultrasound at 20wks and serial growth scans starting at 32 weeks. She developed type A1 gestational diabetes. She underwent a scheduled, uncomplicated, repeat Cesarean



Fig. 3. T1-weighted Sagittal MRI demonstrating a myometrial thickness of just 2 mm.

delivery at 37 weeks and delivered a healthy, male infant weighing 8lb8oz.

3. Discussion

Despite being described over a century ago, angular pregnancy remains an obscure entity, with <100 cases reported in the literature, and is perceived to be rare [3]. The first 39 cases were compiled in a systematic review published in 1981 and reported a 38.5% chance of spontaneous abortion, 13.6% chance for uterine rupture, and a 28% live birth rate [2]. This was updated in 2014 with the addition of 46 subsequent cases, adjusting estimates to 18% risk of spontaneous abortion and 28% risk of uterine rupture [3]. The overall live birth rate was similar at 25%, but of those pregnancies managed expectantly and not terminated, this rose to 69%. With the addition of our case, only 31 instances of expectant management to viability resulting in a liveborn fetus have been reported in the literature [3,8,9].

While angular pregnancies should be considered a potentially viable intrauterine pregnancy, there is a significantly increased risk of uterine rupture when expectant management is chosen. Spontaneous uterine

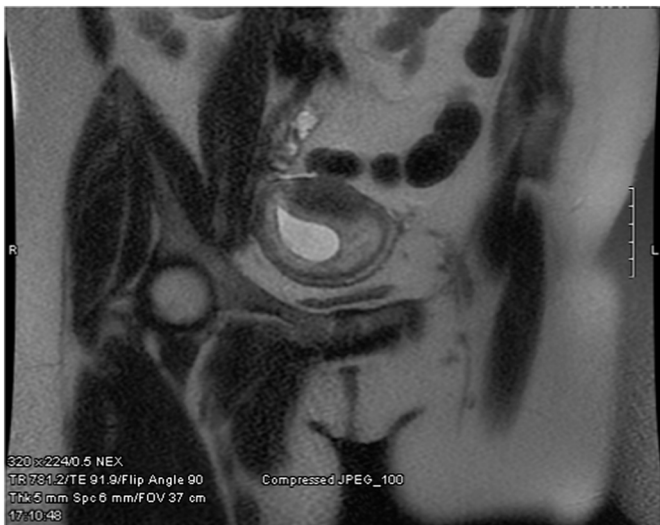


Fig. 2. T2-weighted Coronal MRI demonstrating the asymmetric location of the gestational sac which extends into the right cornua.



Fig. 4. Laparoscopic view of the uterine fundus.

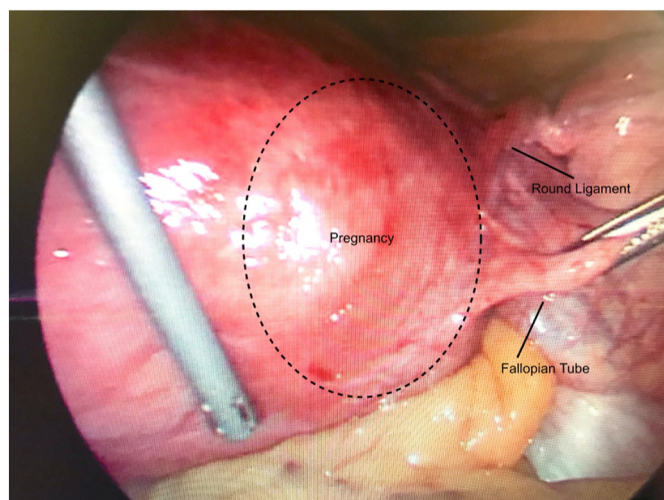


Fig. 5. Laparoscopic view of the uterine fundus. Suction is used to gently probe the uterus while the right tube is gently grasped and placed on lateral traction.

rupture has been reported as early as the 10th gestational week, and resultant catastrophic hemorrhage incurs a 5% maternal mortality rate [2]. In patients electing termination of these high-risk pregnancies, most undergo successful trans-cervical terminations including suction D&C, direct hysteroscopic resection (with and without concomitant endoscopic control), or hysteroscopic injection of methotrexate (MTX) followed by delayed hysteroscopic resection 48 h later [2,3,8,11,12]. In their retrospective review, Grant et al. [8] identified 10 patients with angular pregnancy misdiagnosed as interstitial pregnancy, 7 of whom were successfully treated with systemic MTX alone, while 2 needed additional hysteroscopic evacuation. One abstract reported a laparoscopic approach with uterine preservation (without additional details) [13], but it is likely that more angular pregnancies, misdiagnosed as interstitial, have been treated with laparoscopic cornuostomies or wedge resections.

Interstitial pregnancies are much more commonly reported in the literature and comprise 2–4% of all ectopic pregnancies [4]. They are generally considered nonviable, and their mortality rate approaches 2.5% due to the increased risk of uterine rupture and severe hemorrhage [4,10], typically occurring at <12wks of gestation [14]. Despite the predominant prevalence of interstitial pregnancies, radiologists and obstetricians must maintain vigilance in considering the possibility of angular pregnancy in the differential diagnosis of any pregnancy suspected to be interstitial.

Differentiating interstitial from angular pregnancies has proven difficult despite advances of 3D sonography and MRI. A myometrial mantle measurement of <5 mm, in addition to a gestational sac separate from the endometrium, is often denoted as indicative of an interstitial pregnancy despite low specificity [15,16]. However, a 2017 retrospective review of 44 interstitial and angular pregnancies found that myometrial thickness <5 mm was present in 96% of interstitial pregnancies but also 50% of angular pregnancies (range 3–8 mm) [8]. An “interstitial line sign” (an echogenic line connecting the endometrial stripe to an interstitial gestational sac) appears to be more specific for differentiating the interstitial pregnancy with reported rates of 100% specificity and 80% sensitivity [8,17].

A sign dubbed “surrounding endometrium” has been recently proposed by Grant as a specific sign for angular pregnancies, demonstrating specificity of 100% with excellent inter- and intra-observer agreement (kappa 0.91 and 0.95) in their retrospective study [8]. This sign is based on the hypothesis that a “double sac sign” (a layer of decidual reaction and a chorionic ring) should be seen in an angular pregnancy

given its endometrial implantation like other IUPs, while an interstitial pregnancy, in a location devoid of endometrium, should not. The sensitivity of this sign, however, is unknown. Similar findings have been noted on MRI. A gestational sac surrounded by T2 hyperintense endometrium suggests an angular pregnancy, while a sac surrounded by T2 hypointense myometrium suggests an interstitial pregnancy [18].

Our case report demonstrates the pitfalls of depending on the myometrial mantle thickness, a radiologic sign with poor specificity, combined with the higher incidence and awareness of interstitial pregnancy, leading to an under-recognition of the potentially viable angular pregnancy. Additionally, it serves to report another rare example of careful, deliberate, and successful management of angular pregnancy to delivery. While expectant management should only be undertaken after understanding the risks of miscarriage and catastrophic uterine rupture, it does remain an option. Until prospective studies validate the “surrounding endometrium sign”, lateral displacement of the round ligament remains the sine qua non of angular pregnancies. Providers should counsel patients as such and be willing to perform a diagnostic laparoscopy for definitive diagnosis.

Contributors

Cherie Q. Marfori is the primary author.

Mollie Kotzen is the secondary author.

Both authors contributed significantly to the writing of the paper.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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