Posterior Uterine Wall Rupture of an Unscarred Uterus in a Woman with History of Congenital Diaphragmatic Hernia Repair

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Abstract

Background: Rupture of an unscarred uterus is an uncommon but potentially life-threatening event associated with maternal and fetal morbidity and mortality. Patients lacking common identifiable risk factors can lack appropriate cautions or lead to delayed diagnosis.

Case report: A 24-year-old woman, gravida 5, para 2, with a single gestation at 41 weeks and 1 day was admitted to the labor and delivery unit with regular uterine contractions. Her obstetrical history was significant for two previous uncomplicated spontaneous vaginal deliveries, one ectopic pregnancy and one missed abortion. Her medical history was unremarkable. Her surgical history was significant for a congenital diaphragmatic hernia repair as an infant and dilation and curettage for missed abortion. During an uneventful labor, a sudden loss of fetal station and decelerations to 80 beats/min occurred, leading to an emergent caesarean section. A 4 cm rupture site at the left lower uterine segment posterior to the uterine artery was found, and the infant was delivered through this site. The uterus was repaired without complication and maternal postpartum course was uncomplicated, leading to discharge on post-operative day #4. The neonate was discharged from the NICU on day of life #5.

Conclusion: Additional risk factors may exist in patients with a history of uterine rupture of an unscarred uterus than previously have been identified. Possible defects in formation of collagen leading to increased risk of uterine rupture should not be discounted in a patient with a history of a potential collagen vascular disorder.

Keywords
Uterine rupture, Unscarred Uterus


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150 beats/min. With additional pushing, mild variable decelerations were noted. After the 8th uterine contraction with active pushing, the fetal heart rate was difficult to monitor externally. A fetal scalp electrode was inserted into the vagina and a loss of fetal station from +3 to -1 was noted by the provider. At this time, the FHR was noted to be 60 beats/min externally. The patient was transported to the OR for an emergency cesarean section. Upon entry into the abdominal cavity, a fetal arm was noted to be protruding from a 4 cm rupture site at the left lateral lower uterine segment, posterior to the uterine artery. The rupture site was extended to 10 cm, and the infant was delivered through the posterior hysterotomy. The uterus was repaired without complication in two layers. APGARS for the neonate were 4, 7 and 8 at 1.5,10 minutes respectively. The neonate was transported to NICU for respiratory distress, but was discharged on day of life # 5. Maternal postpartum course was uncomplicated and she was discharged on post-operative day # 4. The patient was also counseled that the trial of labor should be avoided for her future pregnancies due to an increased risk of uterine rupture.

Discussion

The patient in the current case was gravida 5, para 2, with a surgical history of dilation and curettage for a missed abortion. Uterine perforation is a quite rare (0.16% occurrence rate) but serious complication of dilation and curettage [2]. Most uterine perforations following curettage may be neither suggested nor detected [3] leading to lack of provider awareness during labor. This seemingly silent risk of uterine rupture is the reason perforation, while rare, is a dangerous complication of D&C. However, it was less likely that our patient had a uterine perforation during the previous D&C due to successful vaginal delivery 1 year after D&C.

Weakness of the myometrium due to increased parity and/or connective tissue disorders is another risk factor for uterine rupture often overlooked by healthcare providers. Our patient presented with a surgical history of correction of congenital diaphragmatic hernia (CDH) as an infant. Connective tissue disorders, namely Ehler-Danlos Syndrome (EDS), have been associated with CDH and can go undiagnosed after surgical correction of the defect [4]. The signs and symptoms for individuals with EDS vary in severity, even within one sub-type, and the frequency of complications change individually. Some people have negligible symptoms while others are severely restricted in their daily life. Although our patient does not seem to have any of typical signs or symptoms of EDS, it is still possible that she may have undiagnosed EDS or another connective tissue disease and further investigation for such diseases was not undertaken after the repair of her congenital diaphragmatic hernia.

EDS is a rare heritable connective tissue disease caused by a defect in the production of collagen or proteins that interact with collagen, and it is characterized by tissue fragility, hypermobility of the joints, and skin hyper extensibility. The prevalence of EDS is approximately 1 in 5000 births, and an incidence of CDH in 22% of EDS patients has been reported [5]. Type IV Ehler-Danlos Syndrome results in decreased synthesis and/or impaired function of type III collagen, leading to increased risk of arterial dissection and uterine rupture due to increased tissue fragility [6]. Uterus atony leading to maternal hemorrhage is another complication during labor and delivery for a patient with EDS type IV. Dysfunctional type III collagen of the myometrium may lead to obstetrical complications in a patient with EDS type IV [7]. A study on mice have shown abundant levels of type III collagen between bundles of smooth muscle cells throughout the myometrium during all periods of gestation [7]. It is plausible that similarities in type III collagen synthesis in the myometrium of both mice and humans exist. Inherited defects of collagen type III synthesis and/or function could lead to uterine weakness, predisposing a patient with an unscarred uterus to uterine rupture.

The clinical presentation of uterine rupture varies widely, including abdominal pain at the site of rupture, change in fetal station and heart rate, and even maternal shock due to hemorrhage. Uterine rupture should be in the differential diagnosis for any woman with known history of previous D&C, collagen vascular disease or, by proxy, a history of congenital diaphragmatic hernia repair with abdominal pain and/or fetal heart rate anomalies. While the definite etiology of uterine rupture in this case is still unclear, awareness should be raised among obstetrical providers that rupture of unscarred uterus does occur. Due to the significant maternal and fetal morbidity and possible mortality, clinicians caring for patients with a history of collagen vascular disease, or congenital diaphragmatic hernia by proxy, should maintain a high index of suspicion for the clinical signs of uterine rupture.

References