

Spontaneous Rupture of Unscarred Uterus in a Term Primigravida with Lethal Skeletal Dysplasia Fetus (Thanatophoric dysplasia). A Case Report and Review of the Literature

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Background and Importance: Spontaneous uterine rupture, especially in an unscarred uterus, is a rare pregnancy complication that can cause severe morbidity and mortality in both the mother and the fetus. The vast majority of uterine ruptures occur in the presence of a previous uterine scar, most commonly from a previous cesarean delivery. To our knowledge, here we reported the first case of spontaneous rupture of unscarred uterus in a term primigravida secondary to lethal skeletal dysplasia fetus (Type 1 Thanatophoric dysplasia) faced by a practicing clinician in an underdeveloped country (Somalia) with a successful outcome.

Case Presentation: The patient was 24 yrs. Old Primigravida, at 40 weeks gestation by LMP, presented with abdominal pain and active vaginal bleeding; she did not receive antenatal care during pregnancy; after initial abdominal ultrasonography and vaginal examination, laparotomy was performed due to high suspicion of uterine rupture. After dead fresh fetal extraction, the uterine defect was repaired successfully, and the patient was discharged home in good condition after several days.

Conclusion: Through this case, we would like to highlight the urgent need to focus on and recognize the importance of receiving antenatal care in the community so that the burden of thousands of lives lost each year can be reduced.

Keywords: uterine rupture, Thanatophoric dysplasia, Primigravida, antenatal care, case report

Introduction

“Uterine rupture” refers to a tear in the uterine wall during pregnancy or delivery.¹ A rupture in an unscarred uterus is usually a catastrophic event that can result in the death of the mother and baby and extensive damage to the uterus. Sometimes the damage to the uterus is irreparable, and a hysterectomy will be necessary in these cases.² The incidence of uterine rupture has been reported at 0.03–0.09% of deliveries,³ although data are often unavailable from low- and middle-income countries.^{4,5}

Spontaneous uterine rupture is a rare pregnancy complication.¹ Its occurrence may result in significant morbidity and mortality in both the fetus and the mother.⁶ The diagnosis of spontaneous uterine rupture is often missed or delayed.⁷ The main risk factor for uterine rupture is a history of a previous cesarean section (the “scarred uterus”).^{8,9} Most spontaneous ruptures happen during labor and usually involve the lower segment of the uterus.^{10,11} According to research by the World Health Organization (WHO), the incidence of rupture of an unscarred uterus is 1/8000 to 15,000 deliveries or 0.006%.¹² Uterine rupture is more prevalent and remains a major obstetric complication in resource-poor countries.^{4,13} In resource-poor nations, including Somalia, uterine rupture is more frequently associated with obstructed labor, inappropriate use of uterine stimulants (often performed by untrained birth attendants), lack of antenatal care, grand

multiparity, and poor access to emergency obstetric care.^{2,14,15} In industrialized, high-resource countries, uterine rupture occurs most frequently in women who have had a previous cesarean delivery.¹⁶ Uterine rupture in a primigravida patient is rare and, in most cases, completely unexpected.^{1,12,17,18} Without precipitating factors that have either compromised the myometrium's integrity or applied abnormal physical stresses to the uterine wall, uterine rupture is unlikely to happen, and the apparent known cause of myometrial integrity loss is prior to cesarean delivery.¹⁹ We present a rare case of uterine rupture spontaneously in a primigravida mother who delivered a term fetus with congenital lethal skeletal dysplasia without apparent risk factors.

Case Presentation

A 24-year-old patient, gravida 1, at 40 weeks gestation by LMP, was brought to the emergency department by her family complaining of abdominal pain for 12 hours and active vaginal bleeding. She had no significant past medical history and never had antenatal care during pregnancy. She does not smoke or use drugs except antiemetics and analgesics (Navidoxine and Paracetamol). The patient had no significant personal or family history of congenital anomalies. She also denies a previous history of surgical operations, dilation and curettage, and abortions. Initially, the patient felt mild uterine contractions, minimal vaginal bleeding, and lower abdominal pain at night. The patient's condition deteriorated in the morning, and she was brought to the hospital. The patient denies a trail of labor with misoprostol or oxytocin at home or another hospital. On examination, she was pale and distressed, with a blood pressure of 70/40 mm Hg and a maternal tachycardia of 120 bpm. The uterus was tender to palpation, with no contractions noted. The patient was hemodynamically unstable.

An ultrasound scan showed a fetus with no heartbeat in a transverse position. The location of the placenta could not be identified due to much free fluid in the abdominal cavity. Vaginal examination revealed a 3 cm dilated cervix with 50% effacement and active vaginal bleeding. Due to the emergency, we could not perform a detailed fetal ultrasound. The

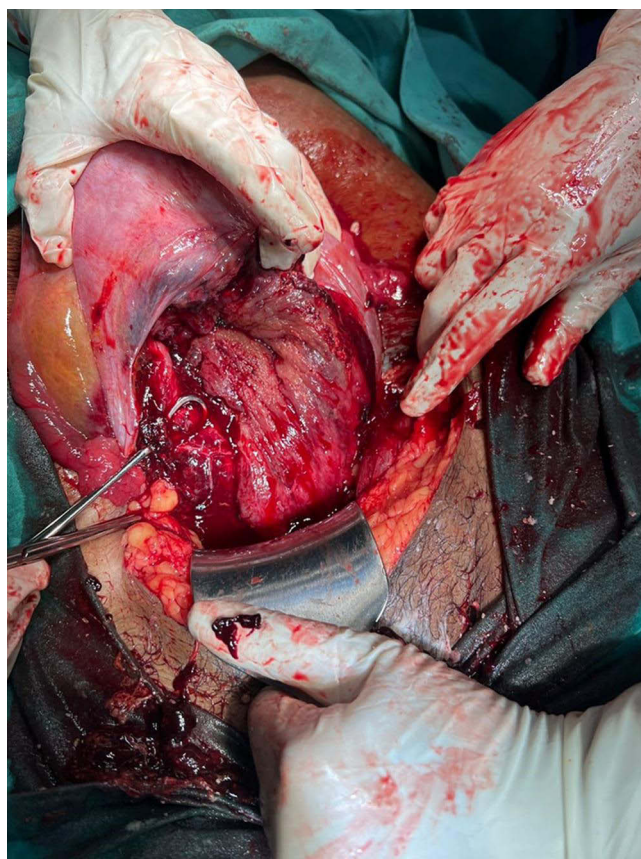


Figure 1 Rupture of the uterus from fundus down to cervix anteriorly.

hemoglobin level at arrival was 5.4 g/dl. The patient was rushed to the operating room for an emergent operation due to suspicion of uterine rupture or placental abruption. The patient had immediate laparotomy under general anesthesia through a Pfannenstiel incision. At surgical exploration, massive hemoperitoneum was evident. A dead fresh female infant weighing 3140 grams with lethal skeletal dysplasia was found extruded in the abdominal cavity and was extracted with no need for a hysterotomy incision (Figure 1).

There was marked frontal bossing with large anterior and posterior fontanelles. The nasal bridge was flat, and the ears were low set. The limbs and fingers were remarkably short, proximally and distally, with redundant skin. There was a narrow thorax with a protuberant abdomen. Based on these findings, a Type 1 Thanatophoric dysplasia diagnosis was made (Figure 2). The fetal head and abdominal circumferences were 40 cm and 55 cm, respectively. After the evacuation of the hemoperitoneum, a rupture of the uterus from the fundus anteriorly to the lower segment down to the cervix was observed (Figure 3). There was no uterine anomaly in this case, and the location of the placenta was anteriorly in the uterine fundus. The bladder was intact. The uterine defect was repaired with a continuous double-layer closure with a 1–0 vicryl absorbable suture. After obtaining hemostasis, layers were closed anatomically. The total estimated blood loss was 3000 mL; the patient was transfused with three units of red cell pack and three units of fresh frozen plasma. The patient was discharged on the fourth postoperative day with a hemoglobin level of 9.6 g/dl in good condition. After one week of follow-up, the patient was advised to use a contraceptive method for two years and prompt antenatal care for subsequent pregnancies.

Discussion

Spontaneous rupture of an unscarred uterus is extremely rare, especially in Primigravida mothers.^{18,20,21} A review of case reports of uterine rupture in primigravida women revealed an association between specific risk factors. In addition to prior uterine surgery,^{9,19} uterine rupture has been reported in women with uterine anomalies^{17,22} and abnormal

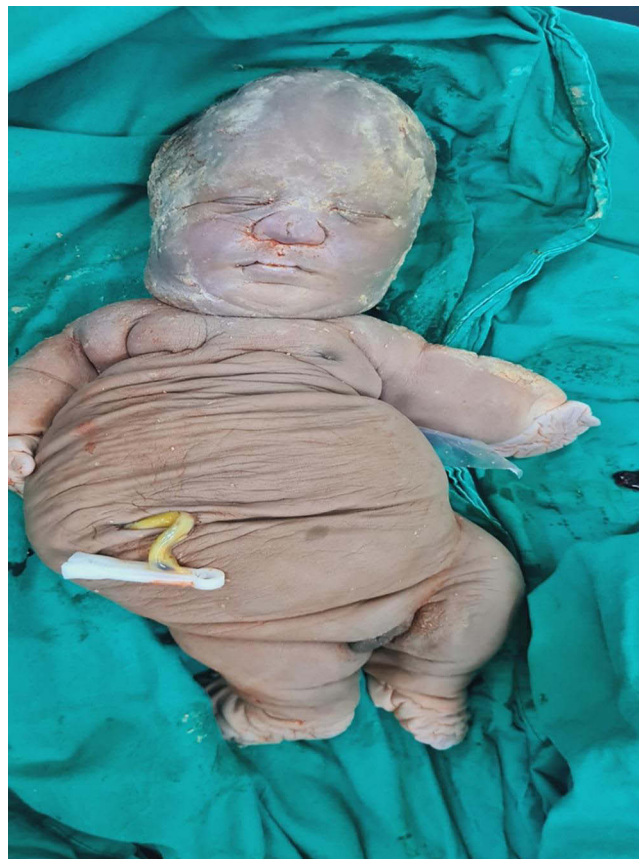


Figure 2 Female fetus at 40 weeks of pregnancy with short limbs and redundant skin folds, macrocephaly, narrow bell-shaped thorax, and protuberant abdomen.



Figure 3 Radiography of the fetus showing telephone receiver-like curved femurs and humerus with irregular metaphysis.

placentation.^{23,24} Uterine rupture was also linked to uterine adenomyosis,²⁵ dilatation and curettage,^{26,27} and maternal connective tissue disease, specifically Ehlers-Danlos syndrome.²⁸ Labor induction with misoprostol^{11,29} and labor augmentation with oxytocin³⁰ were linked to uterine rupture in unscarred women. Uterine perforation during operative hysteroscopy has been associated with the possibility of uterine rupture during subsequent pregnancies.¹⁸ However, none of the reported cases in the literature mentioned an association between uterine rupture and fetal skeletal anomalies. The current case describes spontaneous rupture of an unscarred uterus in a term Primigravida with lethal skeletal dysplasia fetus (Type 1 Thanatophoric dysplasia). In our case report, we could not pinpoint a specific condition as the cause of the uterine rupture.

Skeletal dysplasias are heritable disorders characterized by abnormalities of cartilage and bone. Thanatophoric dysplasia is the most lethal form of skeletal dysplasias caused by an autosomal dominant mutation in the FGFR 3 gene.^{31,32}

Regarding our case report, it is worth noting that the lack of antenatal care during pregnancy was an important factor that contributed to this patient's rupture of the uterus. Antenatal care is an effective method of improving pregnancy outcomes.

Conclusion

Uterine rupture is a rare complication, particularly in primigravida women and unscarred uterus. However, routine antenatal care, clinical suspicion, early detection, and prompt management should be the best options for preserving maternal and perinatal outcomes.

Abbreviations

FGFR, Fibroblast growth factor receptor; LMP, last menstrual period.

Ethical Approval

According to the guidelines of the Ethics Committee of Somali Turkey Recep Tayyip Erdogan Hospital institutional approval was not required to publish the case reports.

Informed Consent

The patient provided written informed consent for publication of the details of her case and the images shown in the figure.

Author Contributions

All authors made a significant contribution to the work reported, whether that is in the conception, Case design, execution, acquisition of data, analysis and interpretation, or in all these areas; took part in drafting, revising or critically reviewing the case; gave final approval of the version to be published; have agreed on the journal to which the article has been submitted; and agree to be accountable for all aspects of the work.

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Disclosure

The authors report no conflicts of interest in this study.

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