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Case Report

Spontaneous uterine rupture with placenta accreta at 17 weeks of pregnancy in a known case of systemic lupus erythematosus: a case report

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ABSTRACT

Over the past ten years, cases of uterine wall thinning and placental abnormalities complicated by Systemic Lupus Erythematosus (SLE) during pregnancy have been reported. Uterine rupture without prior scarring is uncommonly associated with SLE and prolonged steroid therapy. Long-term steroid use can lead to muscle degeneration; however, the exact mechanism behind myometrium thinning remains unknown. A retrospective case study of a patient with a known case of SLE admitted in a tertiary care centre of South Gujarat with spontaneous uterine rupture at 17 weeks of pregnancy. A qualitative analysis of the diagnosis and management procedures was done, offering a critical assessment of their effectiveness and the potential implications for future clinical practice. Systemic lupus erythematosus is an autoimmune disorder occurring mostly in females of reproductive age group. Genetic factors, altered B and T cell immunity, environmental factors, and production of auto-antibodies lies behind the pathogenesis of SLE. Long term steroid therapy may lead to decreased estrogen leading to uterine rupture. Extended use of systemic steroids in patients with SLE may elevate the risk of spontaneous rupture of an unscarred uterus. While abnormal placentation and uterine rupture are exceedingly rare, they necessitate timely diagnosis and early intervention

Keywords: Systemic lupus erythematosus, Spontaneous uterine rupture, Placenta accreta

INTRODUCTION

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease that is relatively prevalent among women of reproductive age. During pregnancy, SLE can lead to various obstetric complications such as miscarriage, premature delivery, and preeclampsia. Over the past decade, there have been documented cases of placenta accreta spectrum in pregnant women with SLE.[1] These cases often involved thinning of the uterine wall, resulting in uterine rupture and necessitating hysterectomy in patients on long term steroid therapy.[2] Glucocorticoids have been shown to clearly inhibit uterine growth stimulated by estrogen. Additionally, glucocorticoids inhibit the secretion of GnRH and gonadotropins, and they suppress the production of testosterone and estradiol by the testes and ovaries, respectively.[3] Pregnant women with SLE are associated with an increased risk of disease flares and adverse pregnancy outcomes. Advances in understanding the course of SLE and its management during pregnancy in recent decades have contributed to improved outcomes for both mothers and infants. The prognosis for both mother and child is optimal when SLE has been quiescent for at least six months before pregnancy and the mother's kidney function is stable and normal or near normal. Lupus nephritis can deteriorate during pregnancy. Maternal health and fetal development should be closely monitored throughout the pregnancy, with follow-up care provided by an obstetrician experienced in high-risk cases.

CASE REPORT

A 30-year-old primigravida, came to the emergency department following referral from a private hospital with complains of abdominal pain with vomiting, hypotension and difficulty in breathing with 17 weeks of pregnancy in a known case of systemic lupus erythematosus for 13 years. On general examination she was conscious, cooperative and well oriented to time, place and person. Her pulse was 136 bpm, BP was 80 mm Hg systolic on inj. Noradrenaline at 12 ml/hour with severe pallor. On examination of respiratory system, her respiratory rate was 30/min with bilateral decreased air entry with SpO2 of 92% on room air.

She was kept on NRBM mask for stabilisation and shifted to ICU. She was on tab. Hydroxychloroquine for SLE on a regular basis with history of taking steroids for 2 years continuously which she stopped a few months back and taking tab azathioprine on and off for the past 3 years. On per abdomen examination 18 weeks size uterus was felt with severe abdominal tenderness. Urgent bedside Ultrasound was done which was suggestive of a single live intrauterine fetus of 17 weeks 5 days of gestational age with moderate amount of hemoperitoneum in pelvic and peritoneal cavity with discontinuity in posterior uterine wall suggestive of uterine rupture.

Her complete blood count showed a Hb-5.9 gm/dl, WBC-28,000/mm3, PLT-290,000/mm3, HCT- 16.9%. She was shifted for an emergency laparotomy under general anaesthesia. Intra-operatively around 2l of hemoperitoneum was present with 350gms of blood clot. Multiple uterine rents were present, largest measuring around 3×4 cm2 in size at the fundus of uterus with active bleeding present from the uterine rents. Uterus was thinned out over the anterior and posterior wall around the area of fundus.

Hemoperitoneum was drained and extension of the uterine rent was done along the fundus of uterus. Foetus of around 200 gm with placenta was removed through the fundal incision. Placental tissue was found to be adherent over the thinned out uterine area. Thinned out uterine wall was removed and uterine rent was repaired in an interrupted manner. Bilateral uterine artery ligation was done and uterus was preserved. After proper haemostasis and closure, patient was shifted to ICU and kept intubated on IPPV mode of ventilation with 100% FiO2. Intra-operatively 2 units of PCV and 4 unit of FFP was transfused. Post operatively she was maintaining her vitals and was extubated the following day. Further 2 unit of PCV was transfused. Her post-op period was uneventful and she was discharged on 9th post-op day with advice from rheumatologist for continuation of Tab hydroxychloroquine. On histopathological examination of uterine tissue, it showed chorionic villi with trophoblastic proliferation within myometrium suggestive of adherent placenta.

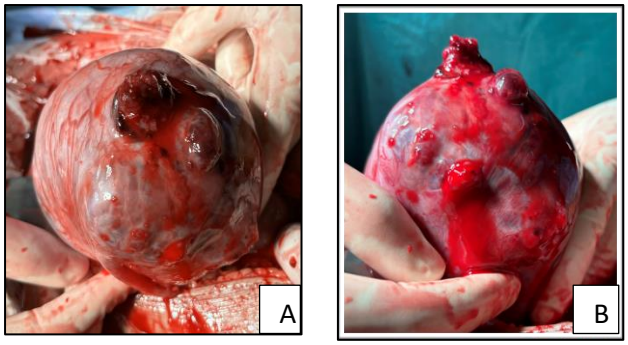

Figure 1 (A and B): Intra-operative images showing multiple rents in uterus with active bleeding present from the rents.

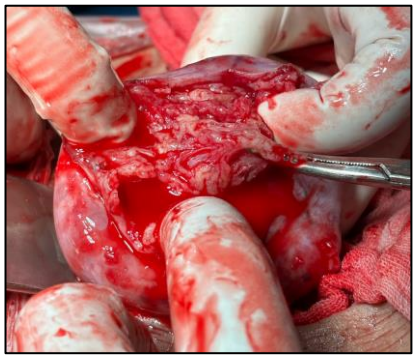

Figure 2: Placental tissue seen through the extended uterine incision.

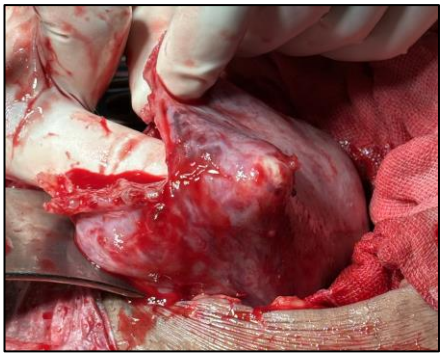

Figure 3: Extremely thinned out uterine myometrium.

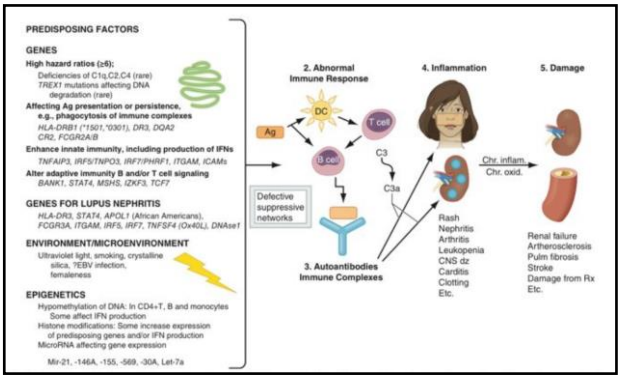

Figure 4: Pathogenesis of SLE.

DISCUSSION

Systemic lupus erythematosus (SLE) is a classic autoimmune disorder characterised by the production of antibodies against nuclear components, accompanied by a wide range of clinical symptoms. In patients with SLE, primary pathological features include inflammation, vasculitis, immune complex deposition, and vasculopathy. SLE predominantly affects females of reproductive age. Abnormal estrogen metabolism has been observed in both male and female SLE patients.[4]

Stress-induced activation of the hypothalamic-pituitary-adrenal (HPA) axis, or the systemic administration of glucocorticoids, is linked to the suppression of the reproductive axis. This leads to the inhibition of testosterone and estrogen production in the testes and ovaries, respectively. Additionally, glucocorticoids may interfere with the ability of sex steroids to act on their target tissues thus inhibiting uterine growth.[5] The exact cause of SLE remains unknown. The observed concordance of SLE in identical twins, increased prevalence among first-degree relatives, and elevated risk among siblings of SLE patients indicate a polygenic inheritance pattern.

The hypothalamo-pituitary-adrenal (HPA) axis serves as the central component of the stress response system. Stress-induced elevation in glucocorticoid levels is crucial for preventing the unchecked amplification of immune responses, which can lead to self-injury and autoimmunity. Aberrant B cell activation is observed in SLE patients, with an increased number of B cells at all stages of activation in peripheral blood, which can precede disease onset. T cell function is also affected, typically resulting in a reduced total number of T cells in peripheral blood, potentially due to anti-lymphocyte antibodies.

The primary immunological disturbance in SLE involves the production of autoantibodies. These antibodies target various self-antigens located in the nucleus, cytoplasm, and on cell surfaces, as well as soluble molecules such as IgG and coagulation factors. Antinuclear antibodies are particularly characteristic and are present in over 95% of patients with SLE. Specific antibodies like anti-double stranded DNA (ds-DNA) and anti-Sm antibodies are unique to SLE.[4]

Although genetic factors and the hormonal environment may predispose individuals to SLE, the disease likely begins due to various environmental triggers and external factors. Infectious, diet, toxins and drugs, physical and chemical agents, like UV light, these can induce inflammation, cellular apoptosis, and tissue damage. The impact of these environmental triggers on genetically predisposed individuals varies greatly, potentially contributing to the diverse nature of the disease, which manifests in alternating periods of flare-ups and remission. Pregnancy complications such as preeclampsia, preterm labor, and fetal growth restriction are known to occur in patients with SLE.[7] Patients with systemic lupus erythematosus (SLE) and on prolonged low-dose steroid therapy can experience uterine rupture with weakened myometrium with only visible serosal layer and exposed placenta and villous tissue with hemoperitoneum requiring emergency laparotomy.[8]

During pregnancy, low estrogen levels can lead to inadequate uterine growth, particularly if there is rapid fetal growth or increased amniotic fluid volume after the second trimester. Additionally, prolonged steroid use can impair protein synthesis and result in thinning of the uterine wall muscle layer. Namita et al. reported that lupus itself may significantly thin the myometrium throughout the uterus. Therefore, as pregnancy progresses, insufficient uterine expansion may contribute to uterine rupture. However, it remains uncertain whether abnormal placentation is solely due to steroids, SLE, or a combination of both.

In pregnant women with sSLE, careful consideration of abnormal placental attachment is essential, along with preparation for managing significant perioperative bleeding to ensure optimal care. There is a heightened concern regarding the potential development of placenta accreta spectrum in patients with connective tissue diseases, depending on the degree of placental attachment to the uterus.

The presence of maternal autoantibody complexes at the maternal-fetal interface is linked to complications during pregnancy. Accumulation of autoimmune complexes at the site of placental attachment disrupts normal placental development, as villous tissues attach directly to the uterus without the usual decidual cells, which are deficient. Furthermore, prolonged steroid therapy is known to reduce natural killer cell numbers in the uterus, thereby impairing uterine spiral artery maturation during placental development.

Pre-conceptional care

To optimise pregnancy outcomes in patients with SLE, careful planning is essential. The activity of the disease at the time of conception is a critical determinant of maternal health, with higher disease activity correlating with poorer outcomes. Both maternal and fetal prognoses are most favourable when SLE activity has been in remission for at least six months prior to conception.[9]

The choice of contraception for SLE patients depends on factors such as current disease activity, the presence of antiphospholipid antibodies, age, reproductive history, as well as patient preferences and cultural considerations. When preparing for pregnancy, it is crucial to review and adjust the patient's medications to minimise potential risks to the fetus while ensuring the mother's condition remains stable.[10] The longer a patient remains in remission at the time of conception, the lower the likelihood of pregnancy exacerbation. Before pregnancy, baseline laboratory

assessments should be conducted including complete blood count, renal function tests, blood urea nitrogen, liver function tests, anti-Ro/SSA, anti-La/SSB, anti-dsDNA antibodies, C3, C4, CH50 levels, 24-hour urine protein, and either creatinine clearance or spot urine protein-to-creatinine ratio. Establishing baseline complement and anti-dsDNA levels can aid in distinguishing between preeclampsia and a lupus flare during pregnancy.

SLE is a chronic inflammatory condition with implications for both the mother and foetus. Managing the mother's illness appropriately and mitigating adverse effects on the foetus are crucial considerations. Preconception planning forms the cornerstone of management.

CONCLUSION

All pregnant women with SLE should continue taking hydroxychloroquine. It is crucial to plan pregnancy carefully when the disease is well-managed with medications safe for pregnancy. Establishing a healthcare team that includes a maternal-fetal medicine specialist or obstetrician experienced in managing SLE patients is equally important. Hydroxychloroquine should generally be maintained throughout pregnancy unless there are specific contraindications.

Successful pregnancies in women with SLE require close monitoring and a collaborative approach. In our view, pregnant women with SLE who have been on long-term, even low-dose steroids should be mindful of potential complications such as abnormal placental attachment, uterine rupture, preeclampsia, preterm labor, and fetal growth restriction, even in the absence of other risk factors. Clinicians should be vigilant for signs of uterine wall thinning and structural abnormalities between the placenta and uterine wall in SLE patients without additional risk factors and on long term steroid therapy. Understanding these risk factors facilitates prompt diagnosis and management of uterine rupture.

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REFERENCES

1. Mitoma T, Hayata K, Yokohata S, Ohira A, Kashino C, Kirino S, et al. Diffuse myometrium thinning and placenta accreta spectrum in a patient with systemic lupus erythematosus (SLE): a case report and review of the literature. BMC Pregnancy Childbirth. 2022;22(1):535.
2. Noh JJ, Park CH, Jo MH, Kwon JY. Rupture of an Unscarred Uterus in a woman with long-term steroid treatment for systemic lupus erythematosus. Obstet Gynecol. 2013;122(2):472.
3. Kim HM, Chin YJ, Cha HH, Seong WJ, Kim MJ. Placenta accreta and uterine rupture of unscarred uterus in patients with systemic lupus erythematosus with prolonged steroid exposure: a report of two cases. Clin Exp Obstet Gynecol. 2020;47(5):785–8.
4. Mok CC, Lau CS. Pathogenesis of systemic lupus erythematosus. J Clin Pathol. 2003;56(7):481–90.
5. Rabin DS, Johnson EO, Brandon DD, Liapi C, Chrousos GP. Glucocorticoids inhibit estradiol-mediated uterine growth: possible role of the uterine estradlol receptor. Biol Reprod. 1990;42(1):74–80.
6. Chou D. Harrison's Manual of Medicine. New York: McGraw-Hill. 2003;7:1038.
7. Clowse MEB, Jamison M, Myers E, James AH. A national study of the complications of lupus in pregnancy. Am J Obstet Gynecol. 2008;199(2):127.
8. Zheng J, Zhou L, Hu J. Spontaneous unscarred uterine rupture at 13 weeks of gestation after in vitro fertilization-embryo transfer: A case report and literature review. Medicine (Baltimore). 2023;102(49):36254.
9. Kaur B. Systemic lupus erythematosus and preconception care. Int J Pregnancy Child Birth. 2020;6(4):99–102.
10. Yamamoto Y, Aoki S. Systemic lupus erythematosus: strategies to improve pregnancy outcomes. Int J Womens Health. 2016;8:265–72.

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