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A Case Report on Pregnancy with Systemic Sclerosis

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Abstract:- Pregnancy outcomes of patients with systemic sclerosis includes abortion, miscarriage, preterm birth, preeclampsia, iugr, oligohydramnios and perinatal death. Besides these poor outcomes, women with systemic sclerosis can safely have healthy pregnancy. The aim of this study is to evaluate a pregnant women with systemic sclerosis. A year old G2A1 women with a history of systemic sclerosis for last 12 years was admitted At Government Medical College Srinagar, with 37 weeks 6 days of pregnancy. On admission she fad severe degree of hypertension with proteinuria. She also had restricted mouth opening, salt pepper pigmentation, barnett sign, sclerodactyly, thickening of skin. She was given anti-hypertensive. Her all routine blood investigations including antitopoisomerase, Ab SCL70 and ultrasonography for fetoplacental profile sent. After stabilization of blood pressure she was prepared for emergency caesarean section as USG showed 37 weeks 6 days foetus, Oligohydramnios with breech presentation. She underwent emergency LSCS and delivered a healthy girl baby of weight 3.351kg. Her postoperative period was uneventful. Rheumatologist, dermatologist consultation taken for her further management in the postpartum period. She was discharged after 1 week. In conclusion many women with systemic sclerosis may have successful pregnancy, but pregnant women with systemic sclerosis should be followed up by a interdisciplinary medical team to control the disease activity and avoid complications.
Keywords-systemic sclerosis, pregnancy, preeclampsia.

I. INTRODUCTION

Scleroderma is a multisystem disorder of unknown etiology characterized by fibrosis and thickening of the skin resulting from accumulation of connective tissue and by involvement of visceral organs causing skin thickening, polyarthritis and joint stiffness. The hallmark of the disease is overproduction of normal collagen. Visceral involvement may lead to dysphagia, reflux esophagitis, pulmonary fibrosis, cor pulmonale and myocardial fibrosis, fibrinoid necrosis of renal arteries and renal failure with or without malignant hypertension. The prevalence of the disease is 1 in 10,000 with a 3-to1 female dominance[1] pregnancy associated with scleroderma is an uncommon condition and if multiple organs involved the prognosis is poor. Scleroderma may appear for the first time during pregnancy or postpartum, but it does not usually deteriorate during pregnancy if the condition is stable at conception[2]. A pregnant patient with scleroderma should be followed up by an interdisciplinary medical team including obstetrician with experience in high risk pregnancy and rheumatologist to control disease activity and avoid possible complications. In the current study we presented a pregnancy associated with a rare disease scleroderma.

II. CASE PRESENTATION

A 32 year old married female G2A1 married since 18 months abortion (medical management) women with a history of systemic sclerosis for last 5 years was admitted with 37weeks 6 days of pregnancy. On admission she had severe degree of hypertension with proteinuria. She also had restricted mouth opening, salt pepper pigmentation, barnett sign, sclerodactyly, thickening of skin. her vital signs including temperature, blood pressure, pulse rate, and respiratory rate were; 36,6 ºc, 160/100 mm of hg, 92 beats/min and 18/min, respectively. The examination of heart, chest, lymph node, neurological system was unremarkable. Initial laboratory values were all in normal ranges. But previously it was detected that antinuclear (ANA) and antitopoisomerase i (anti-scl 70) were positive, anti-RNA polymerase iii and anti-centromere antibodies were negative. She was given injection labetalol for lowering her blood pressure .her all routine blood investigations including antitopoisomerase, ab scl-70.

- 9Ma with systemic Sclerosis with Raynauld phenomenon since 2011 with digital ulcer.. With HRCT DOCUMENTED ILD mild PAH Gerd and ana and anti scl 70 positive.
- Patient is vaccinated with PCV 13 I/M FLU VACCINE AND CORONA VACCINE
- 2D ECHO was done before pregancy shows NO PAH REST ECHO NORMAL AS SUCH..
- PFT FVC 71%
- After patient get pregnant
- ON EXAMINATION RAYNAULD NOTICED WITH PITTING SKIN WITH SCLERODACTLY WITH MILD CHEST CREPITUS With DRY CORNEA.. In pregancy patient done 2 D ech shows mild TR with RSVP 32 MMHG +RAD
- FETAL ECHO GROSSLY NORMAL WITH NORMAL 1:1 AV CONDUCTION...

Ultrasonography for fetoplacental profile sent. After stabilization of blood pressure she was prepared for emergency caesarean section as USG showed 37 weeks 6 days foetus, oligohydramnios with breech presentation. She underwent emergency LSCS and delivered a healthy girl

baby of weight 3.367kg. Her postoperative period was uneventful. Rheumatologist, dermatologist consultation taken for her further by management in the postpartum period. She was discharged after 1 week. Follow up with rheumatologist, dermatologist, taken.

III. DISCUSSION

Scleroderma is a multisystem disorder that has a prevalence rate of approximately 1 in 10,000 with a 3-to-1 female dominance, and pregnancy with scleroderma is rare depending on the potential negative effects on fertility of the disease. Previously it had been reported that infertility was higher in patients with scleroderma than in healthy controls before the diagnosis was made[3]. But subsequent studies have refuted this argument[4]. For years, scleroderma had been considered a strict contraindication for pregnancy, because patients were thought to be at high risk for poor fetal and maternal outcome, including maternal death[5]. Although some studies demonstrated that women with scleroderma have acceptable pregnancy outcomes, scleroderma pregnancies were considered as high risk pregnancies. Indeed, there is an increased risk for premature and small fullterm infants in these patients. Reported preterm delivery rates arose up to 39% in patients with scleroderma and most of them being observed on or after gestational age 34[6]. Therefore, they should be followed by an in obstetrician experienced in high-risk pregnancies. At the onset of pregnancy, a patient who has scleroderma should be carefully evaluated to determine the type of disease, duration of symptoms, and the extent and severity of visceral involvement. Women who have less than 4 years of scleroderma symptoms, those who have diffuse cutaneous scleroderma, or those who have anti-topoisomerase or RNA polymerase iii antibodies are at greater risk for having more active, aggressive disease than are those who have longstanding disease with anti-centromere antibody[7].

It is generally suggested that no woman with scleroderma should attempt to get pregnant within the first 3 years of diagnosis, because disease related complications are more likely to appear within these years of the disease and could complicate pregnancy. The frequency of miscarriage, preterm birth and small gestational weight[8] babies increased in pregnant women with scleroderma. In a retrospective case control study, it was shown that in women who later developed scleroderma, there was an increased incidence of pregnancy complications, such as hypertension and intrauterine growth restriction. Women with diffuse scleroderma are at greater risk for developing serious cardiopulmonary and renal problems. At early stages of disease, they should be encouraged to delay pregnancy until the disease 2 stabilizes.

The worst life-threatening complication of a pregnancy is scleroderma renal crisis. In case of renal crisis, angiotensin converting enzyme inhibitors are recommended, despite the fact that[10] they are associated with congenital abnormalities and are relatively contraindicated in pregnancy. During pregnancy, scleroderma remains clinically stable in 40-60% of patients, worsens in 20%, and 11 improves in 20% related to the heterogeneous nature of the disease. Our case had emergency cesarean section when she was 36 weeks. She had no multiple visceral involvements and duration between diagnosis and pregnancy was around 5 years. Her previous laboratory parameters including immunologic markers were as follows; anti-topoisomerase i (anti scl-70) positive, anti-rnapolymerase iii and anti-centromere antibodies negative. Maybe these conditions have allowed us to achieve good pregnancy outcome in our patient. The optimal mode of delivery in patients with scleroderma is still controversial. Vaginal delivery is associated with lesser blood loss which not causes hemodynamic instability, but has a prolonged second stage of labor and issues regarding[12] increased pressure with contractions. The soft tissue thickening caused by scleroderma is also required abdominal delivery. But, tracheal intubation for general anesthesia has a special concern1because of limited ability of these women to open their mouths widely. We performed a successful delivery without any complications in our patient.

IV. CONCLUSION

Many women with systemic sclerosis may have successful pregnancy, but pregnant women with systemic sclerosis should be followed up by a interdisciplinary medical team to control the disease activity and avoid complications. although pregnant patients with scleroderma are thought to be at high risk in terms of poor. fetal and maternal outcome, they can have healthy babies and successful pregnancies. however, a pregnant patient with scleroderma should be followed up by an interdisciplinary medical team including obstetrician and rheumatologist to control disease activity and avoid.

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