SYSTEMIC SCLEROSIS AND ITS OBSTETRIC CHALLENGES- A CASE REPORT

Dr. Chitra Champawat*
Senior Resident, Department of Obstetrics and Gynaecology Hindu Hrudya Samrat Balasaheb Thackeray Medical College and Dr. R. N. Cooper Hospital, Mumbai, Maharashtra, India. *Corresponding Author

Dr. Reena Jatin Wani
Professor Additional and Head of Department Obstetrics and Gynaecology Hindu Hrudya Samrat Balasaheb Thackeray Medical College and Dr. R. N. Cooper Hospital, Mumbai, Maharashtra, India.

Dr Rashmi Jalvee
Assistant Professor, Department of Obstetrics and Gynaecology Hindu Hrudya Samrat Balasaheb Thackeray Medical College and Dr. R. N. Cooper Hospital, Mumbai, Maharashtra, India.

ABSTRACT
Systemic sclerosis (SSc) is a connective tissue disorder that affects women of childbearing age at least five times more than men. For a long time, SSc has 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. Careful planning, close monitoring and appropriate therapy allows these patients to have a successful pregnancy.

KEYWORDS
Systemic sclerosis, pregnancy.

INTRODUCTION:
Systemic sclerosis is an autoimmune connective tissue disease. It has a female preponderance. It typically affects those aged 30 to 50 years. It can be of two types diffuse or limited. Over production of normal collagen is the hallmark. With diffuse cutaneous systemic sclerosis, skin thickening progresses rapidly, and skin fibrosis is followed by systemic involvement. In the more benign form i.e. limited cutaneous systemic sclerosis—the progression is slow. Pulmonary interstitial fibrosis along with vascular changes may cause pulmonary hypertension. Antinuclear antibodies are found in 95 percent of patients, and immunoincompetence often prevalence of approximately 1 in 22,000 pregnancies.

CASE REPORT:
24 year old patient, G4P3L1UFD1NND2 presented at 31 weeks of gestation for antenatal checkup, she was referred from a private hospital with complaints of pain in the abdomen and decreased fetal movements since one day. She also had history of sudden onset of swelling all over body, episodic burning sensation over tips of the fingers and hands and multiple joints of upper and lower limb along with febrile illness, since 6 month after her second pregnancy. She had developed decreased pitch of voice, tightening of skin all over body, difficulty in swallowing and difficulty in breathing with gradual muscle wasting over 3-4 months.

Patient had seasonal difficulty in breathing with cold clammy finger tips, uneasiness & palpitations especially during winter. She was on ayurvedic medications on & off with a little response.

She had a bad obstetric history with first child male, full term vaginal delivery, died after 20 hrs of life. Second child is 3½ year old female, full term normal vaginal delivery, healthy and alive. Third pregnancy was twin gestation, patient delivered at 8 months of gestation with first twin female IUFD, and second twin was a male child who died immediately after birth. The present pregnancy is her fourth pregnancy, spontaneously conceived.

On general physical examination, she was febrile, with pulse rate of 140 beats per minute, Blood pressure of 140/100 mm of Hg, with salt pepper pigmentation over anterior neck, chest, and bilateral upper and lower extremities. Thickened skin over bilateral distal upper & lower extremities sparing face, neck, chest and abdomen. Small ulcer over left dorsum near thumb. Bilateral elbows having lesions with chalky white discharge suggestive of calcinosis cutis, and microstomia. Pallor and telangiectasia of finger nails.

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On obstetric examination, cardiovascular and respiratory systems were normal. On per abdomen examination, uterus was 24 to 26 weeks size, with single intrauterine lie in cephalic presentation, fetal heart rate was not localized on auscultation and on doppler.

Per vaginum examination was suggestive of cervical os 2.5 cm dilated, poorly effaced, vertex presentation, station high, membranes present.

Patient was investigated. Routine blood investigations were found to be normal. Serum calcium was 8.8 mg/dl. Serum uric acid was 5.1 mg/dl. Serum uric acid was 5.1 mg/dl. Both were within normal range. ESR was 37 mm at the end of one hour. Rheumatoid factor, Serum CRP, Anti Centromere antibody and Anti SCL 70 antibody all were found to be negative.

Obstetric ultrasound was done which was suggestive of intrauterine fetal demise of 30 weeks gestation with mild fetal asceses. Induction of labour was done with dinoprost gel and patient delivered vaginally within 6 hours of induction, a female fresh still born child of 1.343 kg. There were no intra partum or postpartum complications.

Figure 1: Thickening of skin with salt and pepper pigmentation, ulcerations and calcinosis cutis.

Figure 2: Pallor and telangiectasia of finger nails.
CONCLUSION: Women with scleroderma should be encouraged to delay pregnancy until their disease has stabilized and the risk of renal crisis is less, which is usually 3–5 years from the onset of symptoms. All pregnant women with scleroderma should be monitored particularly closely and taught how to recognize early signs of labor so their physicians can try to delay delivery or prepare their infants for early birth. Women with systemic sclerosis can safely have healthy pregnancies if pregnancy is planned when the disease is stable and managed by a multidisciplinary team during pregnancy.

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