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CASE REPORT

Pregnancy Outcome in Young Patient with Scleroderma

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Introduction

Scleroderma is a rare progressive connective tissue disorder that more often affects women in their fourth decade of life. Hence pregnancy is rarely encountered with this disorder. This rare disorder leads to fibrosis that involves skin and various internal organs like lungs, heart, kidneys and gastrointestinal tract. Poor pregnancy outcome is reported in literature. Here a case of scleroderma in a young woman with good pregnancy outcome is reported.

Case A 28 year old primigravida presented at 9 weeks of pregnancy with symptoms of pigmentation, thickening and tightening of skin over face and limbs for 5 years with history of painful ulcers over her finger tips for 1 year. USG at 9 weeks was normal with gestational age corresponding to the period of amenorrhoea. On examination she had induration and hyperpigmentation of skin over both upper limbs up to elbow. Digital pitted scars were present. There was no induration over chest, back or lower limbs. Decreased wrinkling of forehead and pinched up nose were present. Oral orifice and oral mucosa were normal (Fig. 1). Dermatology opinion sought with a suspicion of scleroderma was confirmed after skin biopsy.

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Immunologist's opinion was taken regarding the management. She was advised to take tab. nifedepine 10 mg twice daily and tab. aspirin 75 mg once daily from 16 weeks of gestation and to continue folic acid. USG at this time did not reveal any fetal anomalies. She was hospitalized and further evaluated at 24 weeks of pregnancy as she complained of difficulty in breathing. X-ray chest P/A taken with abdominal shield did not reveal any evidence of interstitial lung disease (Fig. 2).

Haemogram, renal function tests (RFT) and liver function tests (LFT) were normal, 24 h urine protein was less than 150 mg. Anti scleroderma antibodies-70 Ig G were positive. Lupus erythematosus cell (LE), APLA (antiphospholipid antibodies), lupus anticoagulant and anticardilipin antibodies were negative. CPKT-25 IU/l (creatine phosphokinase test). ECHO was normal, USG KUB (kidney urinary bladder) was normal. Fundus did not show any evidence of vasculitis. She could not perform pulmonary function tests (PFT). She was advised to continue tab nifedepine and to take tab deriphyllin retard 300 mg twice daily.

Third trimester 2-D USG evaluation was normal. She was hospitalized at 37 weeks of pregnancy and was re-evaluated. ECHO, RFT and LFT and were normal. Her haemoglobin was 8.6 gm%. Her height was 141 cm and pelvic assessment showed pelvic outlet contraction. She underwent elective LSCS at 38 weeks under spinal anaesthesia. There were no anaesthetic complications. She was transfused with one unit of packed cells as her post-op Hb was 6 gm%. The neonate was alive, male, weighed

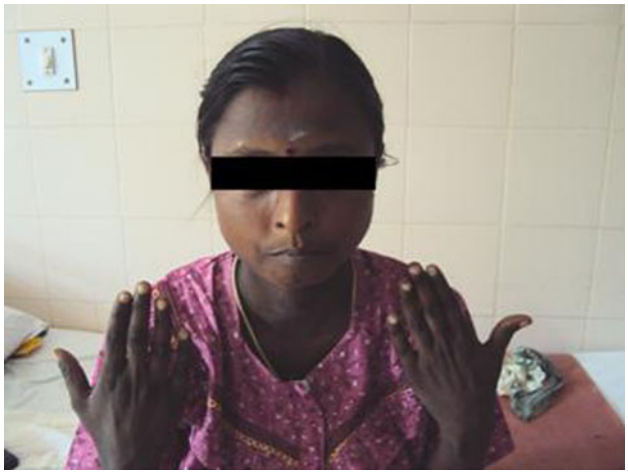

Fig. 1 Patient profile with her hands in which small healed ulcers are present

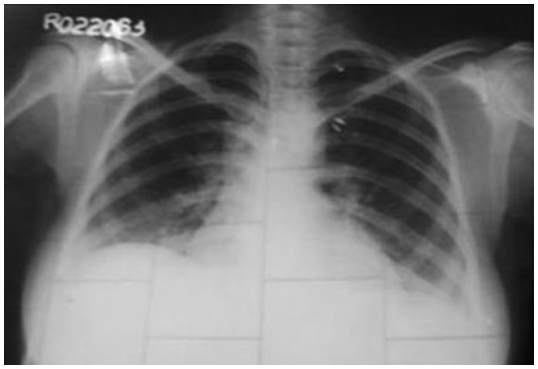

Fig. 2 X-ray chest P/A—increased bronchovascular markings

2.9 kg and showed syndactyly of hands and feet. The neonate was evaluated for the presence of congenital heart disease (ECHO) which was reported as normal. She was reviewed by the immunologist and ECG and ECHO were advised which were normal. X-ray chest did not show any evidence of interstitial lung disease. She recovered well and discharged on the 8th post-operative day with advice to continue tablet nifedepine and deriphyllin along with haematanics.

On follow up the patient conceived after 1 year and she had developed breathlessness after 32 weeks of pregnancy despite taking tablet deriphyllin. The pulmonary function tests at this time revealed restrictive lung disease (FEV1-33; FVC-46; FEV1/FVC-84; PEFR-pre and post-270 l/min). She underwent elective LSCS with sterilisation at 37 completed weeks of pregnancy under spinal anaesthesia. Alive male baby 2.8 kg did not show any anomaly. The post-operative course was normal.

Discussion

Scleroderma is progressive with exacerbations and remissions and is associated with increased maternal mortality, in the past so much so that pregnancy was advised against in these women [1]. It was reported that approximately 40% of women do not show any adverse symptoms and 40% deteriorate and 20% may improve during pregnancy [2]. A study of systemic sclerosis during pregnancy over a 10 year period (1987–1996) concluded that women with systemic sclerosis can safely undertake pregnancy. This study analysed 91 pregnancies in 59 women and found that the incidence of preterm births was high (29%) and that of miscarriages was not increased. Raynaud's phenomenon was improved but oesophageal reflex had worsened. Renal crisis occurred in three women with early diffuse scleroderma [3]. Delivery by LSCS in a woman who presented with preterm labour and cervical incompetence as she had decreased pulmonary function because of severe pulmonary fibrosis resulted in good outcome [4]. Mortality occurred in a woman who was managed in a similar way as the disease had progressed faster and multiple visceral involvements occurred, resulting in respiratory failure due to interstitial fibrosis with alveolar bronchopneumonia and renal failure [5]. The present case though complained of little breathlessness during the later half of pregnancy did not go into preterm labour; the most possible reason for this may be that she was on tablet nifedepine, which also has a tocolytic action. Other problems reported are high incidence of pre-eclampsia, intrauterine growth restriction and intrauterine death [1].

Renal crisis usually occurs during pregnancy with diffuse involvement of skin and with other visceral involvement and it is life threatening to the mother and fetus. Pregnancy should be avoided in patients of recent affection, diffuse involvement of skin and involvement of other viscera [4, 5]. The other rare complications reported in literature include spontaneous pneumo-mediastinum and subcutaneous emphysema.

Conclusion

Scleroderma progression was not observed in this patient during pregnancy. Pregnancy can be allowed in patients with systemic scleroderma involving skin. However, frequent monitoring and assessment is necessary to rule out organ involvement. Treatment with nifedepine will help to prevent preterm labour as well as to retard the progression of disease. Thus, pregnancy may be achieved with good maternal and fetal outcome with careful planning, close monitoring and aggressive management. The risk for premature and small infants may be minimised with specialised obstetric and neonatal care.

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