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CASE REPORT

A Case of Pregnancy with Systemic Sclerosis Presenting with Acute Dyspnoea

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ABSTRACT

Pregnancy in women diagnosed with systemic sclerosis generally has a favorable outcome. It is now known that the majority of these women undergo a normal progression of pregnancy if the right time for delivery is chosen and a close obstetric care is delivered. The obstetric risk will depend on the subtype and clinical stage of the disease, and the presence and severity of the internal organ involvement during the pregnancy. The management of these pregnancies should be provided in a specialized center, with a multidisciplinary team capable of identifying and promptly treating complications. Here we report a case of systemic sclerosis in a young woman with good pregnancy outcome.

Introduction

Systemic sclerosis (SS) is a connective tissue disease of unknown origin that is characterized by cutaneous and visceral fibrosis, production of auto-antibodies, and prominent microvascular changes. Severe organ involvement, early diffuse systemic sclerosis with rapid onset, and pulmonary hypertension ought to discourage patients from pregnancy, as these situations are at high risk of complications for both mother and fetus during pregnancy. Regular follow-up of pregnant patients with systemic sclerosis by an interdisciplinary medical team including gynecologists and rheumatologists is necessary to control disease activity and avoid possible complications.

Case report

A 30 years old unbooked, multigravida carrying eight and half month pregnancy presented with acute onset of dyspnoea and palpitation. She had history of dyspnoea, cough and dysphagia for past one month. There was no history of dyspnoea in previous pregnancy and no significant family history. On examination, she was alert, conscious, co-operative with height of 156cm and weight of 56 kg, pulse was 100/min regular, blood pressure of 110/70 mm of Hg, respiratory rate of 30 per minute, bilateral mild peripheral oedema, bilateral coarse crepitations on chest examination with no murmur. There was malar rash on face. Her per abdomen examination revealed small for gestational age foetus and presence of normal foetal heart rate. Routine investigation revealed haemoglobin of 10.6 gm/dl, total

lecocyte count of 14000 with neutrophils 62%, liver and renal functions were normal and so was urine routine/ microscopy. Patient was shifted to high dependency unit and managed conservatively. A Rheumatological consultation was taken and it was found that patient had positive raynaud's phenomena and modified rodnan skin score of 7 with hyper pigmentation of skin over both upper limbs up to elbow. Decreased wrinkling of forehead and pinched up nose was present. Oral orifice and oral mucosa were normal. The autoimmune profile revealed positive anti scl-70, antinuclear antibody and anticardiolipin antibody. 2D Echo was normal.

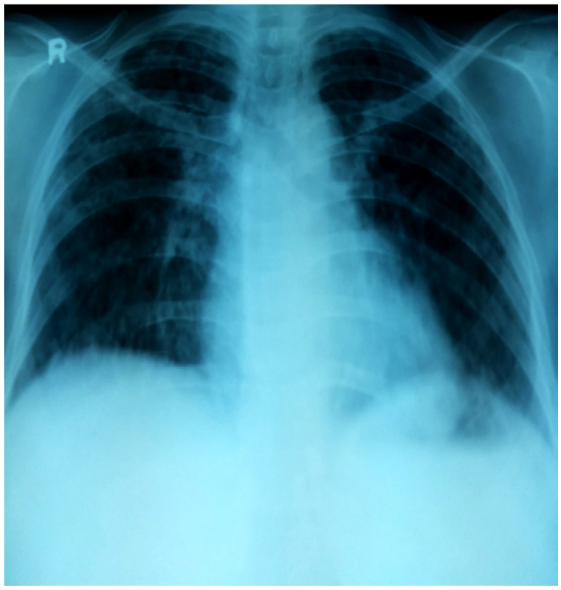

Fig. 1: Chest x-ray (P-A view)- increased bronchovascular markings

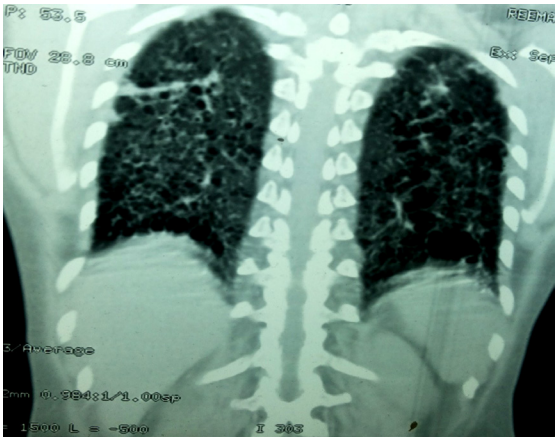

Fig. 2: HRCT of chest- Interstitial Lung disease, including alveolitis and interstitial fibrosis

Patient was started on prednisolone, ecosprin, sildenafil and N-acetylsteine with coverage of antibiotic. After 2 weeks elective LSCS was done under epidural analgesia. Single live male baby weight 2.5kg born with Apgar score 9 at the 1st and 5th minute Post op period was uneventful. Post natal chest X-ray and HRCT was done which showed alveolitis and interstitial fibrosis characteristic of Interstitial Lung disease (Fig. 1,2). Her dyspnoea improved drastically. She was discharged on post-operative day 14 with contraceptive advice and regular follow up in rheumatology clinic.

Discussion

Systemic sclerosis (SS) is a chronic autoimmune disorder characterized by progressive fibrosis of the skin and visceral tissues as well as a non-inflammatory vasculopathy manifesting as Raynaud's phenomenon, digital ulcerations, pulmonary arterial hypertension, and scleroderma renal crisis. It is a relatively rare disease, with an annual incidence 20 cases per million adults[1, 2]. SS has a strong female predominance, with an approximate female to male ratio of 5:1[1, 2]. It was reported that approximately 40% of women do not show any adverse symptoms and 40% deteriorate and 20% may improve during pregnancy[3].

Limited cutaneous SS is characterized by a more prominent vasculopathy with less cutaneous fibrosis, is associated with anti-centromere antibodies, and generally has a prolonged clinical course prior to diagnosis. In contrast, diffuse cutaneous SS more closely resembles graft-versus-host disease, has more prominent cutaneous and organ fibrosis, is associated with anti-topoisomerase I antibodies, and has a shorter period between symptom onset and diagnosis[4].

Scleroderma progression was observed in this patient during pregnancy. Physiologic hemodynamic changes are an integral part of human pregnancy. In some cases, changes in vascular blood flow and decreased vascular resistance may lead to improvement in manifestations of systemic sclerosis. In other cases, an intrinsic inability to adapt to the necessary hemodynamic changes in pregnancy, possibly due to underlying vasculopathy of systemic sclerosis, may contribute to adverse pregnancy outcomes including hypertensive disorders of pregnancy, intrauterine growth restriction preterm delivery and placental abruption. More recently, a series of retrospective and prospective studies have provided more detailed analysis of pregnancy outcomes and have demonstrated that, for most women with SS, pregnancy outcomes are reasonably good[5–10]. Reported rates of early pregnancy loss of 14% – 15%[5,6] are somewhat

increased from the estimated 10% in the general population. Late pregnancy losses were few, and generally occurred in women with severe diffuse SS[6, 8]. Preterm delivery rates have ranged from 8% to 40%[5,6,8]. The majority of preterm deliveries were on or after gestational age 34[7]. Small for gestational age infants (<10th% tile for gestational age)[11], ranged from 0% to 50%[4, 6, 8]. During pregnancy, the most frequent maternal complication is worsening of gastro-esophageal reflux, and the most severe complications are renal crisis and flaring arterial pulmonary hypertension. Angiotensin converting enzyme inhibitors are indicated at any stage of the pregnancy if renal crisis is suspected. Adverse obstetric outcomes are dominated by prematurity which may be the consequence of intra-uterine growth restriction or preeclampsia. Those complications might account for systemic sclerosis-associated vasculopathy, or for the presence of antiphospholipid antibodies. Patients with a previous history of placental insufficiency might benefit from treatment with aspirin, low dose anticoagulants or even nitric oxide donors. Overall, the majority of patients with SSc appear to have reasonable obstetric outcomes although women with rapidly progressive diffuse disease may be at higher risk for complications. 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.

Women with SS can have successful pregnancies, but they have a higher-than-normal risk of preterm delivery, intrauterine growth restriction, and babies with very-low birth weight. Progression of the disease during or after pregnancy is rare, but possible. High-risk multidisciplinary management should be standard for these patients.

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