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SYSTEMIC SCLEROSIS AND ITS OBSTETRIC CHALLENGES- A CASE REPORT

Gynaecology

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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 preprondence. 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 approxi mately 1 in 22,000 pregnancies.[1]

CASE REPORT:

24 year old patient, G4P3L1IUFD1NND2 presented at 31weeks 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.

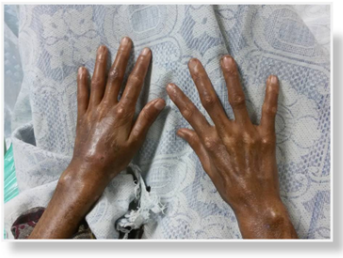

Figure 1: Thickening of skin with salt and pepper pigmentations, ulcerations and calcinosis cutis.

Figure 2: Pallor and telangiectasia of finger nails.

On systemic 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. Fever profile was normal. Serum calcium was 8.8 mg/dl. Serum uric acid was 5.1mg/dl both 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 ascites. Induction of labour was done with dinoprost gel and patient delivered vaginaly within 6 hours of induction, a female fresh still born child of 1.343 kg . There were no intrapartum or postpartum complications.

Patient was further evaluated by Rheumatologist and Dermatologist with a probable diagnosis of systemic sclerosis.USG abdomen was suggestive of Hepatomegaly with bilateral medical renal disease. Patient was started on amlodipine for hypertension, hydroxychlor oquine, aspirin, pantoprazole, pentoxifylline & topical fusidic acid for systemic sclerosis as adviced by the rheumatologist and dermatologist. Patient responded well to the treatment and improved symptoma tically. Patient was discharged on day 10 postpartum. On day 21, patient was admitted in medical ICU with complaints of difficulty in breathing. HRCT of chest was done which was suggestive of Areas of consolidation in right lung, moderate right pleural effusion, enlarged necrotic right hilar and mediastinal lymph nodes with features suggestive of infective etiology but with no signs of Interstitial lung disease. She was started on antibiotics and AKT CAT-1 for pulmonary kochs. Patient showed improvement, but took discharge against medical advice after 5 days. Patient later on discontinued her medications and started taking ayurvedic treatment and did not to follow up after this.

DISCUSSION:

Systemic sclerosis is a chronic multisystem disorder of unknown etiology. It is characterized by microvascular damage, immune system activation leading to inflammation, and excessive deposition of collagen in the skin and often in the lungs, heart, gastrointestinal tract, and kidneys. Fetal cell microchimerism is the persistence of fetal cells in the maternal circulation and organs following pregnancy. These persistent fetal cells may stimulate autoantibodies, or they may become engrafted in maternal tissues. This raises the possibility that fetal cell microchimerism is related to the predilection of autoimmune disorders for women.[2].Disease remains stable during gestation if their baseline function is good. As perhaps expected, dysphagia and reflux esophagitis are aggravated by pregnancy.[3] Raynaud's phenomenon tends to improve during pregnancy particularly with the increased cardiac output in the second half of pregnancy due to generalized peripheral vasodilatation develops. Women with renal insufficiency and malignant hypertension have an increased incidence of superimposed preeclampsia. In the presence of rapidly worsening renal or cardiac disease, pregnancy termination should be considered. Renal crisis is life threatening , it is difficult to diagnose and treat during pregnancy as it presents as an acute onset of severe hypertension, edema of the lower limbs with anemia and daily increase of serum creatinine in women who have had diffuse scleroderma for less than 5 years, which mimics preeclampsia. Pulmonary hypertension usually contraindicates pregnancy. Pulmonary hypertension is a serious complication and is associated with 50% maternal mortality. Screening should be done before pregnancy, and termination should be offered if this complication is diagnosed in pregnancy. Elevated liver function tests, which should be normal in renal crisis, and serum creatinine, which increases daily in renal crisis, are the best ways to differentiate between it and preeclampsia and is treated with ACE inhibitors, but it does not improve with delivery.[4] Despite the association of ACE inhibitors with birth defects and infant kidney dysfunction, they must be used urgently in renal crisis because they are the only drugs that successfully control hypertension in renal crisis and the only lifesaving treatment.

Vaginal delivery may be anticipated, unless the soft tissue thickening brought by scleroderma produces dystocia requiring caesarean delivery. Tracheal intubation for general anaesthesia has special concerns because of limited ability of these women to open their mouths widely.[5]Because of oesophageal dysfunction, aspiration is also more likely, and epidural analgesia is preferable

Maternal and fetal outcomes are related to the severity of underlying disease. Early pregnancy losses (14-15%), preterm delivery (8-40%), low birth weight (0-50%) are the complications seen.[6]

Fetal complications are likely related to placental abnormalities that include decidual vasculopathy, acute atherosis, and infarcts. These reduce placental blood flow and are found in many cases.[7]

For women who do not choose pregnancy, several reversible contraceptive methods are acceptable. However, hormonal agents, especially combination oral contraceptives, probably should not be used, especially in women with pulmonary, cardiac, or renal involvement. Due to the often unrelenting progression of systemic sclerosis. Permanent sterilization should also be considered.

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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