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Fate of a pregnant women with mixed connective tissue disease in a tertiary care hospital in India: A case report

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Abstract
Mixed connective tissue disease (MCTD) is a term involving the features of lupus systemic sclerosis, polymyositis, rheumatoid arthritis and high titre of anti ribonucleoprotein (RNP) antibodies however the exact etiology of the disease is not known. MCTD is characterized by microvascular damage, alongwith activation of the immune system leading to inflammation and excessive deposition of collagen in the skin, lungs, heart, gastrointestinal tract and kidneys. The females are more affected, especially after childbirth may be because of the hypothesis of microchimerism, The pathogenesis being a two way migration of fetal cells through the placenta. Although it cannot be cured completely but treatment with corticosteroids is helpful. The complications were preeclampsia, preterm labor, fetal growth restriction, eclampsia, thrombocytopenia and infections like pneumonia, sepsis like syndrome. The maternal mortality rate is approximately around 325/100000. We encountered a similar case of mixed connective tissue disorder in a patient aged 24 years at 29 weeks of gestation. The patient had conceived spontaneously after 5 yrs of diagnosis and had presented with all the known complications of the disease including lung fibrosis, FGR, preeclampsia and ascites. However she delivered and thereafter stabilized, although her baby had to be treated for a prolonged period at the NICU.

Keywords: MCTD, termination of pregnancy

Introduction
Mixed connective tissue disease is the name given to multisystem rheumatic disease with overlapping features of Systemic Lupus Erythromatosus, systemic sclerosis, rheumatoid arthritis, and polymyositis. This syndrome has been described in conjunction with ANAs and antibodies to ribonucleoprotein. Clinical features often seen with these patients include arthritis or arthralgias, lupus-like rash, telengiectasia, Raynaud's phenomenon, hypomotility of the gastrointestinal tract, myalgias, and proximal muscle weakness. The severity of these symptoms is variable, and corticosteroids often are helpful in controlling exacerbations [1]
The course of Mixed Connective Tissue Disease during pregnancy is similar to that seen in SLE patients. Some patients experience disease exacerbations, but most without any significant worsening of their condition. Although the reported experience with this condition and pregnancy is small, it is reasonable to counsel patients that they may be at increased risk for maternal and fetal complications [1].
SLE causes an increased rate of fetal death in utero and spontaneous miscarriage. Neonatal lupus is the occurrence of SLE symptoms in an infant, commonly presenting with a rash resembling discoid lupus erythematosus, and sometimes with systemic abnormalities such as heart block or hepatosplenomegaly.
Aggravation of SLE has been estimated to occur in about 20-30% pregnancies .Renal disease flare-up is the most common presentation of SLE aggravation in pregnancy. Since the prognosis of the disease in a pregnant woman in worldwide reports were quite poor, We thought of reporting our case since it had a successful maternal and fetal outcome.

Case Report
A woman aged 24 yrs, gravida 3 para 1+1,where p1-Iufd at 8 months of gestation,6yrs back, P2-spontaneous abortion at 2.5 months, 5 yrs back presented at our referral system.

She was 29 weeks pregnant and complained of decreased fetal movement. Patient had typical sclerotic skin lesions like diffuse cutaneous skin thickening, rash which was photosensitive and features of systemic sclerosis like essential vasomotor disturbances; fibrosis.(Fig-1)

She was a diagnosed case of Mixed Connective Tissue Disease from the Dept of Rheumatology previously. Patient was receiving treatment from there as follows: T. Aspirin (75mg) once daily, T. Azathioprine (50mg) once daily, T. Nifedipine (10mg) twice daily, T. Tadalafil (10mg) once daily, T. Lobetalol (100mg) twice daily, T. Prednisolone (5mg) once daily, and Multi Dose inhaler with Formeterol thrice daily and Budesonide twice daily.

She was diagnosed with MCTD after her first pregnancy in which she delivered a still born baby weighing 723 gms. After the delivery she developed swelling of her body, ascites and blackish discolouration of skin following which she was unfortunately diagnosed as sepsis however she was treated and was discharged after 10 days of conservative management.

She visited the Hospital again with the complain of rash and there, she was referred to Dept of Dermatology. She had ANA screening test, ANTI-RNP and antibodies to ANTI DS-DNA. She had a Skin biopsy done, which definitively diagnosed MCTD (Fig 2). HRCT thorax (Fig 3) showed minimal fibrosis in right upper lobe and lower lobe of lungs. Upper GI Endoscopy was normal. After the diagnosis she was referred to Rheumatology department of our Hospital for further management of Sclerosis. She was followed up for 3 yrs at Rheumatology Department and during that period, the dose of T. Prednisolone which was 25 mg was tapered down to 5 mg and her Hypertension was controlled on T. Labetolol 100mg twice a day and her chest conditions were controlled with MDI with Formeterol thrice daily and Budesonide twice daily.

The woman had a late booking at 29 wks of gestation in her current pregnancy. She presented in our Obstetric Emergency with Respiratory distress. She was then admitted in the Obstetric ward for further management of her current pregnancy.

After admission, on examination her BP was 140/90 mm of Hg, Spot urine test was trace

- Hb-11.9 g/dl, TLC-5800 cells per cubic mm, Plt-2 lac,Sr.Urea-11 mmol/l, S.Cr 0.8 mg/dl, APTT-32 sec, PT-12.5 sec, INR-1.

To know the baby status USG for growth scan and AFI with Doppler was done where IUGR was diagnosed for the baby showing Average gestational age of 23 wks, EFW 612 gms, AFI of 7 cm with gross ascites and Umbilical Artery Doppler was normal. (Fig-4)

Her routine reports i.e. Complete Blood Count with Kidney fuction tests and Liver function tests were normal. Her coagulation profile was normal at the time of admission .She was managed as a High Risk case and treated with multidisciplinary team involvement with senior Obstetrician, Rheumatologist ,Cardiologist, Chest Medicine and General physician in the team.

She developed respiratory distress with growing ascites at around 31 weeks of pregnancy and was shifted to High Dependency Unit, following which her treatment was discussed in the Multidisciplinary Team Meeting and decision for induction of labour was taken as the condition of the patient was worsening, however her medicines were continued in the same doses.

Induction of labour was done using Dinoprostone gel (0.5mg) and the patient went into labour after single dose of gel. She had a short and precipitated labour of 6-7 hrs. During the labour she was monitored throughout and analgesics were given as required to prevent further respiratory distress and exacerbation of her condition. However the intrapartum and post partum period were uneventful and active management of third stage of labour was done. She delivered a girl baby weighing 990gms and was managed in the NICU for further 2 months for further complications. The mother could feed the child on expressed breast milk going to the NICU. Estrogen containing contraceptive may flare up the symptoms so she was started on progesterone containing mini pills to prevent further pregnancy.

However the patient's general condition improved significantly following delivery and was continued on the same medications as before pregnancy along with some antibiotics. She was discharged from obstetric department after 1 week with advice to continue follow up in Rheumatology department within 6 weeks.

Discussion

The incidence of MCTD with pregnancy is 1 in 22000 pregnancies, [2] MCTD is an autoimmune disease and causes miscarriage due to the vasculopathic nature of the disease (3). The fetal wastage is increased in MCTD. Just like Neonatal lupus syndrome, due to crossing of maternal antibodies the fetus can have fetal hemolytic anemia, thrombocytopenia, leukopenia and congenital heart block [4, 5].

Hence in view of the life threatening complications in the mother as well as the fetus, termination is the best choice.

As suggested in a study on Mixed connective tissue disease in pregnancy: A case series and systematic literature review by Tardif *et al.* [6] MCTD in pregnancy puts women at risk of medical and obstetric complications, and disease activity probably increases this risk. This is in agreement with our report where symptoms exacerbted with pregnancy.

As mentioned in a study named Autoimmune diseases in pregnancy: maternal and fetal outcomes by Vengetesh *et al.* [7] Active disease at onset of pregnancy, presence of Anti-ds DNA antibodies and secondary APS were strong predictors of poor pregnancy outcomes among patients with SLE. Vigilant monitoring during pregnancy is required for favourable outcomes. Initiation of Heparin and Aspirin from early gestation in patients with autoimmune diseases has favourable outcomes. Our patient was already on aspirin when she conceived.

Another study named Pregnancy and autoimmune connective tissue diseases by Marder *et al.* [8], suggested that pre-conception counseling should be conducted routinely for reproductive age females with autoimmune connective tissue diseases, with the goal of planning for pregnancies during periods of disease quiescence in order to optimize maternal and fetal outcomes and during pregnancy and the postpartum period, frequent monitoring by a multidisciplinary team is imperative as was done in our patient during her period of stay in our hospital.

In another study named Pregnancy Outcomes in Mixed Connective Tissue Disease: Results from a Multicentre Cohort Study by Radin *et al.* [9, 10] done The observed live-birth-rate was as high as 72%, with poorer foetal outcomes observed in MCTD women with antiphospholipid antibodies and pulmonary or muscular involvement.

This multicentre retrospective cohort study describes the foetal and maternal outcomes of 203 pregnancies in 94 consecutive women ever pregnant.

Fig 1: Showing Typical Cutaneous Involvement

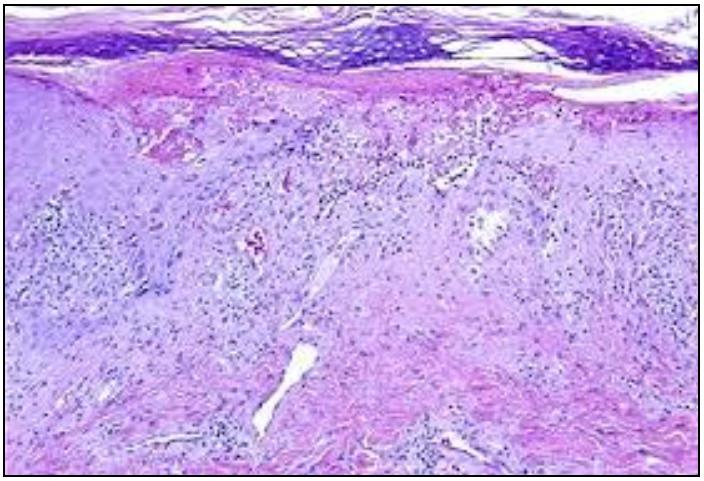

Fig 2: Of Histological Picture of MCTD

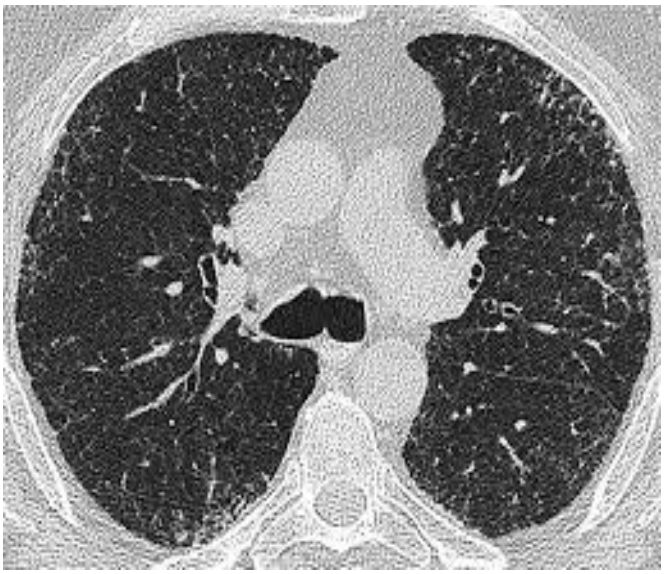

Fig 3: Of HRCT Showing Lung Fibrosis

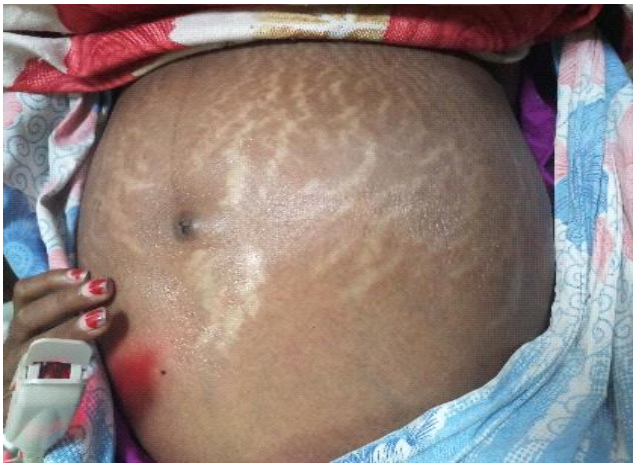

Fig 4: Showing Gross Ascites of the Mother

Conclusion

Thus in conclusion early diagnosis, multi disciplinary management and vigilant monitoring and timely intervention to deliver the baby is advocated to prevent worsening of the complications. Although the prognosis of the disease is poor worldwide, we can try to provide a better outcome to those unfortunate mothers who are having the disease, instead of terminating their pregnancy and advising against childbirth in the future.

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