

Refractory SLE - A rare presentation

Dr. Vidyalekshmy R, Dr. Vijayalekshmi R, Dr. Nina Navakumar, Dr. Geetha P

KIMS HEALTH, Trivandrum

Objective

- To identify the rare presentation of SLE in pregnancy and outcome.

Case report

- 20-year old G2P1L1 at 26 +5 weeks admitted with low grade fever, myalgia, fatigue for 2 weeks. Her previous pregnancy - H/O Gestational hypertension and term LSCS.
- Was detected to be Dengue IgG weak positive with platelet of 58000 , Malarial parasite and Widal negative .
- On admission her vitals were stable .

- Labs showed low platelet count - 28000 , altered liver functions and proteinuria. ANA IF 3 +
- ANA profile -RNP / Sm, Ro + and Anti-ds DNA positive. C3 ,C4 were very low. APLA ,tropical fever workup and blood cultures were negative.

- Obstetric ultrasound showed normal growth and doppler.
- Cardiac evaluation showed normal ECG with Echo showing minimal pericardial effusion .
- Xray chest showed pleural effusion .

- Diagnosis of Active SLE with severe thrombocytopenia was made.

- MDT care was initiated and started on antibiotics. She was treated with IVIG for 5 days , Methyl prednisolone 1 gm IV for 3 days followed by T Prednisolone 60 mg daily and T HCQ 200 mg.
- At 27 +1 gestational age she had BP of 160/100 and was started on Tab Labetalol 200 mg three times a day and T Nicardia 20 mg three times a day was added subsequently.
- At 27 + 5 weeks she had severe headache and blood-stained urine . Platelet count – 11000. Started Mg SO4 for fetal neuro protection. 2 SDP and 1 RDP were given .
- Emergency LSCS was done and delivered a live female baby 995 gms. Baby was on CPAP for 4 days. Intra op ascites was present and prophylactic b/l uterine artery ligation was done.
- MMF was started due to refractory thrombocytopenia after counselling regarding lactation suppression and formula feeds. Discharged with Tab MMF, HCQ , Tab prednisolone. On day 19 , urine and blood culture showed E . Coli sensitive to IV Ceftriaxone. On follow up she had recurrent UTI and was started on Septran prophylaxis

Discussion

- SLE is a chronic multisystem inflammatory autoimmune disease characterized by relapses and remission . It has a prevalence of 28 to 150 per 1 lakh individuals.
- Diagnosis is based on clinical findings and serological parameters. Classification of SLE is based on ACR and SLICC group.
- Pregnancy in SLE can have complications such as
 - Vascular lesions like decidual vasculopathy, thrombosis and infarction
 - Pregnancy loss of 8- 32% in well controlled disease(like general population), otherwise 3 fold increased fetal loss
 - Preeclampsia 15-35%
 - FGR 6-35%
 - Preterm labor 19-49%
- Active disease at the time of conception, renal disease, chronic hypertension, high-dose prednisolone use , APL antibodies are associated with more risk .
- The majority of flares during pregnancy are mild , typically consisting of arthritis and cutaneous manifestations, and are easily treatable. 15-30% of flares occur in pregnancy (any trimester) or postpartum. Risk factors for a flare occurring during pregnancy are active disease within the 6 months prior to pregnancy, severe underlying disease, active nephritis and discontinuation of hydroxychloroquine.
- Management of SLE in pregnancy requires involvement of Maternal and Fetal Medicine specialist, Rheumatologist and Obstetrician.
- Corticosteroids are the first line agent. Azathioprine can be safely used in pregnancy. Cyclophosphamide, Methotrexate, MMF and Leflunomide should be discontinued three months prior to pregnancy. All patients with SLE other than those with quiescent disease should either continue or initiate HCQ in pregnancy. In patients with quiescent disease shared decision making is used to decide regarding initiation of HCQ.
- Patients requiring anticoagulant therapy should be changed from warfarin to LMWH prior to or as soon as pregnancy is recognized. LDASP is started at 12 weeks to prevent preeclampsia.
- Tacrolimus in lupus nephritis is more effective in inducing remission than cyclosporine. Iv Ig is used for thrombocytopenia associated with SLE.

- Prolonged use of NSAIDs(>48 hours) should be avoided during pregnancy. 1g of Calcium daily in pregnancy results in >50% reduction in risk of preeclampsia and 25% reduction in the risk of preterm delivery.

- Contraception- Progesterone only contraceptives are safe. Barrier contraceptives are less effective. OCPs have theoretical risk of flare and are contraindicated in patients with prior thrombosis or aPL antibodies. Intramuscular and implantable progestins - theoretical risk of osteopenia with the use of DMPA
Implants containing etonogestrel –the effect on bone density and thrombosis uncertain, should not be the first choice for patients on long term corticosteroids or with APS

Conclusion

- Refractory SLE may need third line agents like MMF for disease control. Timely delivery and multidisciplinary care improves outcome.

References

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