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Successful management of SLE with lupus nephritis with APLA (Antiphospholipid Antibody) complicating pregnancy a case report

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Abstract : Almost 90 of lupus cases are in women and its prevalence in those of child bearing age is 1 in 500. And 50 of patients have renal involvement.5 have associated thrombophilia. Active nephritis is associated with adverse outcome. However if the disease is in remission phase have good pregnancy outcome. Newly diagnosed lupus in pregnancy have adverse outcome. The maternal mortality is 1.3. A 22 yr old GRAVIDA 2 PARA1 LIVE 0 previous term intrauterine death delivered vaginally two year back with LMP -23315 EDD-301215 a known case of SLE with lupus nephritis in remission phase with APLA was successfully monitored and managed by a multidisciplinary approach in a tertiary set up.

Keyword : Systemic lupus erythematosus, Lupus Nephritis, Antiphopsolipid Antibody Syndrome.

INTRODUCTION:

Lupus is a Heterogeneous autoimmune disease with complex pathogenesis. Affecting 1 in 1250 pregnancy. Immune system abnormality include overacting B lymphocytes that are responsible for autoantibody production that act against tissue and cellular nuclear component resulting in damage. Therefore its variable presentation is due to its multisystem involvement. It results in 7% risk of major morbidity and 1.3% of maternal mortality on pregnancy. Important factors for pregnancy outcome include whether disease is active at the beginning of pregnancy, age, parity, co-existing medical obstetrical disorders, whether APLA detected or not.

CASE REPORT:

A 22yrs old G2P1L0 previous term intrauterine death (dec 2012) delivered vaginally, from lower socio economic status class IV with regular menstrual period, married since 3yrs with LMP 23/3/2015 EDD 30/12/2015 a known case of SLE with lupus nephritis on remission phase with APLA positive came for regular antenatal check up.

PAST OBSTETRIC AND MEDICAL HISTORY:
1st PREGNANCY:

She conceived spontaneously within 3 months of marriage. Ist, IInd trimester uneventful. IIIrd trimester she was admitted with complaints of decreased fetal movements for 3 hrs and lower abdominal pain for 1hr. she was diagnosed as Abruption Grade II / fetal heart rate absent and she delivered a fresh dead born term male baby of weight 2.85Kg no obvious external congenital anamolies. Immediate postpartum she developed PRES (Posterior Reversible Encephalopathy Syndrome), RD (Retinal Detachment) with elevated Blood Pressure (200/120 mm Hg) and elevated Renal parameters (sr. urea:52mg/dl, sr. creatine 2.0mg/dl, urine protein: 3gm/l by dipstick). She was shifted to RGGH (Rajiv Gandhi Government General Hospital for AKI (Acute Kidney Injury) for IMCU care. There she was diagnosed as ANCA positive lupus nephritis (Diffuse Glomerulonephritis C3, C4 (low)) active phase and was started on immunosuppressive therapy. T. Prednisolone 50mg once daily for 8 weeks then tapered to 10mg once daily maintenance therapy. She was started on T.Mycophenolate mofentil 500mg BD, T.Nifidipine 1omg 1 Tab thrice daily, T. Enalapril 2.5mg once daily (OD) and T.Calcium 1 Tab thrice daily(TDS). She was on regular follow up in nephrology department as an outpatient. In Dec 2014 (2yr after previous conception) patient was anxious to conceive. Since patient was in remission phase she was asked to stop T.Mycophenolate mofentil and T.enalapril.. She was advised to take T.Azathioprine 50mg 2HS, T.Hydrochloroquine(HCQ) 200mg 2HS and continue all other drugs. APLA screening was done. Since she was found to be APLA positive (IgM-30 MPL UNITS). she was started on low dose Aspirin 75mg once daily.

PRESENT PREGNANCY:
Ist TRIMESTER:

She conceived 2 yrs after 1st pregnancy. Confirmed by urine pregnancy test on may 2015, LMP 23/3/2015, EDD 31/12/2015. Since she is APLA positive patient was started on Inj. Heparin 5000IU s.c BD after checking platelet count, APTT and INR.

She was said to continue low dose aspirin 75mg OD, T.Azathioprine 50mg 2HS, T.Hydrochloroquine 100mg 2HS and T. Prednisolone 10mg OD. Her platelet count was checked 3 days and 1 week later for any evidence of early and late onset HIT(Heparin Induce Thrombocytopenia). Other base line investigations, complete blood count with peripheral smear, renal function test, liver function test , urine spot PCR done and found to be normal. Nucal translucency (NT) scan was done at 12wks to rule out aneuploidy. Nephrologist, Rheumatologist opinion obtained and they advised to continue the same line of management.

IInd TRIMESTER:

Iron folic acid supplementation given. Serial BP monitoring done.Anomaly scan done, uterine artery, umbilical artery Doppler study done. Hemoglobulin, platelet count, APTT, INR,BT/CT , total protein, OGCT (oral glucose tolerance test), urine sugar, urine protein, spot PCR done found to be normal. Fetal ECHO, maternal ECG and ECHO done to ensure the feto maternal cardiac status. Nephrologist, Rheumatologist opinion reviewed at 24wks and they advised to continue the same line of management. She was advice to come every 3rd wk from 28wk for interval growth and liquor status. She was warned of and advised admission if she developed symptoms and signs of imminent eclampsia, lupus flares, infection, any cardio respiratory symptoms, decreased fetal movement, abdominal pain, draining, bleeding.

IIIrd TRIMESTER:

Interval growth, modified biophysical profile and serial BP monitoring was done every 2 weeks. Nephrologist/Rheumatologist/ ophthalmologist opinion reviewed for any end organ failure. Aspirin was stopped at 34wks and asked to get admitted in case she developed lower abdominal pain, bleeding draining decreased fetal movements, symptoms of imminent eclampsia, infection ,lupus flare, symptoms of end organ failure. She got admitted in causality at 36wks+4days with complaints of decreased fetal movements and lower abdominal pain for 3 hrs. With baseline Hemoglobulin, platelet count, APTT, INR and with 1 packed cell , 4 fresh frozen plasma cross matched patient was taken up for emergency caesarean section under spinal anaesthesia.No intra operative hemorhage. She delivered an alive male baby, late preterm of weight 2.9kg with Apgar 6/10 and 8/10.No obvious external congenital anamolies of baby. Caesarean section was uneventful. No postpartum hemorrhage.

IMMEDIATE POST PARTUM:

1. Hourly vital monitoring (BP, PR, Temp), hourly urine output, urine protein monitored. Monitoring for post partum hemorrhage done.
2. Hemoglobulin, packed cell volume, total count, platelet count, APTT, sr. urea, sr. creatine, sr transaminase was checked and found to be normal.
3. Thromboprophylaxis was started 12 hrs after surgery (Heparin 5000IU s.c BD given for 5 days).
4. Intravenous glucocorticoids given for 3 days and then T. Prednisolone 20mg OD for 2 wks then tapered to 10mg OD thereafter to withheld lupus flares.
5. To continue other immunosuppressive drugs Azathioprine and HCQ. patient asked to review in neprology OPD.
6. To continue T. Enalapril 2.5mg once daily(OD) and T.Calcium 1 Tab thrice daily (TDS), and low dose aspirin 75mg OD patient asked to review in Rheumatology OPD.

DISCUSSION:

SLE is autoimmune disease that results in interaction between susceptibility gene and environmental factors. There are overactive B-lymphocyte and abnormal regulatory T cell function result in production of auto-antibodies. The relative risk of disease is increases if there is inheritance of the autoimmunity gene on chromosome 16 that predispose to SLE, rheumatoid arthritis, crohn's disease and psoriasis. Susceptibility gene such as HLA A1, B8, DR3, DRB1 and TET 3 explain only a portion of genetic inheritability. It is a disease of multisystem involvement with common symptoms of malaise, fever, rash, arthritis, pleuropericarditis, photosensitivity, anaemia and cognitive dysfunction. During pregnancy lupus improve in1/3rd, unchanged in 1/3rd worsen in 1/3rd patients. Pregnancy outcome is best in whom

- Lupus activity is quiescent for past 6 months before conception
- No lupus nephritis manifest with proteinuria/ renal insuffiency
- No evidence of APLA/Lupus anticoagulant
- Superimposed preeclampsia does not develop

ACTIVE NEPHRITIS: has higher incidence with associated gestational hypertension and preeclampsia. 30% suffered lupus flare, 40% had renal insufficiency, 5% had associated thrombophilia

QUIESCENT NEPHRITIS: had nonsignificant effect on preeclampsia

MONITORING AND MANAGEMENT:

Most recommended is continuation of immunosuppressive therapy for nephritis. Lupus activity monitoring and identification of lupus flare by maternal clinical and laboratorial condition. The sedimentation rate is often misleading in pregnancy due to physiological hyperfibrinogenemia. Hence falling complement components c3, c4 , increased D-dimer are associated with active disease. Serial hematological studies for hemolysis (positive comb's, anemia, reticulolysis, unconjugated hyperbilirubinemia), thrombocytopenia, leucopenia, may help in detect change in disease activity. Chronic thrombocytopenia early is due to APLA; late in pregnancy is due to pre eclampsia. There is mild increase in serum aminotransferase due to immunosuppressant therapy can also be misleading. Over proteinuria is ominous sign.

FETUS:

Fetus should be closely monitored for fetal growth restriction, oligohydramnios. Many recommend screening for Anti SSA (anti Rho), Anti SSB (anti La), Anti RNP antibodies which are associated with neonatal lupus syndrome and congenital heart block.

PHARMOCOLOGICAL TREATMENT:

Ø Azathiprine are beneficiary in controlling active disease and are safe in pregnancy.
Ø Cyclophosphamide are terotogenic although if disease is severe can be used after 12wks gestation.

Ø Mycophenolate mofentil and Methotrexate should be avoided in pregnancy
Ø Antimalarial helps in control skin disease. HCQ has not been associated with congenital malformations and discontinuation of therapy may cause flare hence most recommended its continuation in pregnancy
Ø In case of lupus flare high dose glucocorticoid therapy methylprednisolone 1000mg i.v over 90min for 3 days and then return to maintenance dose.

CONCLUSION:

Lupus is notoriously variable in its presentation, course and out-come. Findings may initially be confined to one organ system and later involve other or the disease may first manifest with multisystem involvement. All maternal deaths in SLE are attributable to lupus nephritis in active phase. Infection, lupus flare, end organ failure, superimposed preeclampsia, stroke, cardiovascular involvement account for other cause of death in SLE patients. Those in remission / quiescent phase 6months prior to conception have good pregnancy outcome. Immunosuppressive agents, i.v glucocorticoids in acute exacerbations, thromboprophylaxis, close feto materal monitoring through multidisciplinary approach in tertiary set up have good outcome.

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