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SUCCESSFUL MANAGEMENT OF A CASE OF PREGNANCY WITH SJOGREN SYNDROME

Gynaecology

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ABSTRACT

Auto immune disorders are common among women of child bearing age. Pregnancies complicated by these disorders have a high clinical impact on both the pregnancy and the disease. Sjogren syndrome can either present alone as primary Sjogren syndrome or can present in association with any other auto immune disorder as secondary sjogren syndrome. Laboratory diagnosis of Sjögren syndrome is usually made by the following markers: antinuclear antibodies (most frequently detected), anti SS-A (also called anti- Ro; most specific), anti-SS-B (also called anti-La), and cryoglobulins and hypocomplementemia (main prognostic markers). Sjogren syndrome is likely to worsen during pregnancy. Pregnancy can be complicated with spontaneous abortions , fetal loss and specifically fetal congenital heart block. We have reported a case of pregnancy with bad obstetric history which was diagnosed to have sjogren syndrome and was successfully managed.

KEYWORDS

Sjogren syndrome, antinuclear antibodies, auto immune disease

INTRODUCTION:

Autoimmune diseases do not impair fertility, and women with autoimmune diseases who become pregnant are likely to experience more complicated pregnancies than are women without the disease. Pregnancies complicated by these disorders have a high clinical impact on both the pregnancy and the disease. The effect of autoimmune disease on pregnancy differs according to the type of maternal disease, disease activity, severity of organ damage, antibody profile, and drug treatment.

Sjögren syndrome is an autoimmune disease that can present either alone, as in primary Sjögren syndrome (pSS), or in association with an underlying connective tissue disease, most commonly rheumatoid arthritis or systemic lupus erythematosus (secondary Sjögren syndrome) [1]. The spectrum of clinical presentation of Sjögren syndrome extends from dryness of the main mucosal surfaces to systemic involvement (extraglandular manifestations). Dryness of mucosal surfaces occurs because of immune-mediated inflammation causing secretory gland dysfunction. Sicca features primarily affect the quality of life, whereas the disease prognosis is marked by systemic involvement. Sjögren syndrome is known to occur predominantly in women. Affected women are likely to experience more complicated pregnancies than are women without the disease.

CASE DETAILS

Our patient 31 years Gravida 4,Para 2,Live 1, Abortion 1 visted our antenatal OPD. Her first visit was at 6 weeks of pregnancy. Detailed obstetric histroy taking revealed that in her first pregnancy she had severe pre eclampsia with antepartum eclampsia at 7 months of gestational age, it was terminated , baby expired immediately after birth. During her second pregnancy which was 5 yrs back, she had early onset pre-eclampsia, LSCS was done at 8 months, female baby, alive and healthy now which is her only living issue. Third pregnancy ended in spontaneous abortion at 50 days amenorrhoea which was 3 yrs back. In view of history of recurrent preeclampsia and bad obstetric history, ANA profile was done. Her Anti – Nuclear antibodies (ANA) levels were high, Lupus anticoagulant was negative. She had symptoms of dryness of eyes and mouth. Rheumatologist opinion was sought. She was diagnosed to have Sjogren's syndrome. According to the advice of rheumatologist, she was started on aspirin, heparin and hydroxy chloroquine. At present there was no evidence of active disease. Steriods were reserved for such situations.

Pregnancy was closely monitored by 2 weekly follow up with. Her blood investigations showed Hemoglobin of 10 gm%, renal and liver function tests were normal. Thyroid function tests were also normal. She was monitored for any symptoms of pre eclampsia like excessive weight gain, rise in blood pressure, appearance of peripheral oedema, urine protienuria. Periodic consultation with rheumatologist was also taken.

During her second trimester her haemoglobin level was found to be 8 gm%, so packed cell transfusion was done. She was taking oral hematinics, calcium and folic acid. Her blood pressure measurements were normal. Growth scan was in normal range without any evidence of utero-placental insuffficiency. Aspirin and heparin was stopped at 34 weeks of pregnancy, hydroxy chloroquine was continued as per rheumatologist opinion. Betamethasone was given for lung maturity. Elective repeat Caesarean section was done at 37 weeks. An alive, term, female child 2.5 kg was delivered and handed over to paediatrician. Post operative period was uneventful, treated with iv fluids, analgesics and antibiotics. She was discharged on fourth day with well baby.

PATHOPHYSIOLOGY

The histological hallmark of the disease is the focal lymphocytic infiltration of the exocrine glands. Laboratory diagnosis of Sjögren syndrome is usually made by the following markers: antinuclear antibodies (most frequently detected), anti SS-A (also called anti- Ro; most specific), anti-SS-B (also called anti-La), and cryoglobulins and hypocomplementemia (main prognostic markers)[2] . These markers mediate the tissue damage and are thus responsible for complications in pregnancies of women with Sjögren syndrome. These antibodies cross the placenta beginning at approximately 12 weeks of gestation and may exert the following effects on the fetal tissues: 1) inducing myocarditis; 2) binding apoptotic cells, blocking presumed physiologic clearance, and diverting clearance to macrophages; and 3) producing arrhythmia.

CLINICAL MANIFESTATIONS

Effect of Pregnancy on Sjögren Syndrome

Sjögren syndrome is likely to worsen during pregnancy and more so in the postpartum period. This is because the disease is sometimes complicated by pulmonary hypertension, which frequently worsens during pregnancy and in the postpartum period[3,4].

Effect of Sjögren Syndrome on Pregnancy

Women with Sjögren syndrome are likely to experience more complications during pregnancy compared with those without the disease. Spontaneous abortions, preterm delivery, increased incidence of pre eclampsia, intra uterine growth restriction are more common in these women.

Well-known fetal outcomes in Sjögren syndrome-complicated pregnancies are neonatal lupus and congenital heart block (CHB). CHB is the most severe fetal complication and supposedly occurs because of the damage of the atrioventricular node by anti-SS-A or anti-SS-B antibodies, or both. The reported prevalence of CHB in the offspring of an anti-SS-A-positive woman is 1% to 2%. The recurrence rate in a patient with antibodies, who has a previous child affected, is approximately 10 times higher. The incidence of neonatal lupus in an offspring of a mother with anti-SS-A antibodies is estimated at approximately 1% to 2% [7].

MANAGEMENT

The outcome of pregnancies in women with Sjögren syndrome can be excellent with use of a multidisciplinary management approach involving an obstetrician who specializes in high-risk pregnancies, a rheumatologist, and a pediatrician[9].

Prenatal Management

Women with Sjögren syndrome planning to conceive must undergo good counselling regarding all specific risks and complications involved, medications that are contraindicated during pregnancy, and whether the patient is in the best condition to get pregnant according to underlying disease activity and complications. Ideally, the disease should be well under control three to six months before conception.

Antenatal Management

One of the most dreaded complications of pregnancy in patients with Sjögren syndrome is CHB. A woman is at risk of delivering a baby affected by CHB if she is anti-SS-A positive [5]. Frequent surveillance by serial echocardiograms and obstetric sonograms between 16 to 20 weeks of gestation and thereafter is required for at-risk pregnancies. The goals are early diagnosis and early treatment of incomplete CHB, thus improving the outcome for the fetus[6] . The rationales for management are 1) to decrease maternal autoantibodies and then to decrease their placental transfer and 2) to decrease the inflammation once it occurs before it leads to permanent fibrosis and irreversible CHB. Maternal treatment with fluorinated corticosteroids such as dexamethasone or betamethasone, can reduce the antibody-mediated inflammatory damage. Specific fetal risks include intrauterine growth restriction, oligohydramnios, and possibly adrenal suppression. The alternative therapies, as evidenced by a few case reports, include plasmapheresis, intravenous immunoglobulins, and β-sympathomimetics. In most cases, complete heart block requires a pacemaker implantation in the infant, preferably in the neonatal period[10].

CONCLUSION

Women with Sjögren syndrome are likely to experience more complications during pregnancy. Studies show a high incidence of poor fetal outcomes for these patients. Women with this underlying autoimmune disorder must undergo prenatal counselling explaining all the risks involved and the need to control the disease well before conception. A multi-disciplinary approach by a high-risk obstetrician, rheumatologist and paediatrician has resulted in successful outcome of case of pregnancy with sjogren syndrome.

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