Case Report

Pregnancy with Systemic Lupus Erythematosus with Sjogren Syndrome and intrauterine fetal heart block: A successful pregnancy outcome: A case report:

*Hoque NS[1], Tanvir ATM[2], Fatema NN[3], Hossain S[4], Seraji SI[5]

Abstract:

Fertility is not impaired with autoimmune diseases. Pregnant women with autoimmune diseases, are likely to experience more complications than are women without the disease. Pregnancies complicated by these disorders have a high clinical impact on both the pregnancy and the disease. Sjögren syndrome is an autoimmune disease with a high prevalence of anti-SS-A (anti-Ro) and anti-SS-B (anti-La) antibodies. Anti-SS-A antibodies are associated with congenital heart block. Women with Primary Sjögren syndrome require prenatal counseling explaining the risks involved and the need to control the disease well before conception. High-risk pregnancies can be optimally managed by a multidisciplinary team. Excessive fetal morbidity and mortality have been noted in patients with systemic lupus erythematosus (SLE). The influence of anti-SSA/Ro antibodies on fetal outcome in SLE patients has rarely been reported, but its high association with congenital heart block or neonatal lupus syndrome is well known. Here we are presenting a case Mrs. Tahiya, 34 years, 2nd gravida with 37+wks pregnancy with SLE with Sjogren Syndrome with uncontrolled DM with history of previous 1 LSCS. This case was managed in multidisciplinary approach.

Key words: SLE (Systemic Lupus Erythromatosus), Sjogren syndrome, anti-SS-A(anti-Ro) antibody, anti-SS-B (anti-La) antibodies, Lupus anti-coagulant.

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Introduction

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.2 Sicca features primarily affect the quality of life, whereas the disease prognosis is marked by systemic involvement.3

The effect of autoimmune disease on pregnancy differs according to the maternal disease, disease activity, severity of organ damage, antibody profile, and drug treatment.4 Data on pregnancy outcomes in pSS are scarce, and results have been conflicting.

Case report

Mrs. Tahiya Rahman, 34-year, P-1(LSCS)+0, was presented at her 6weeks of pregnancy with uncontrolled DM. She has been prescribed short acting insulin at that time. She was under regular antenatal checkup under multidisciplinary approach from the very beginning of her pregnancy. Her pregnancy was uneventful upto 13 weeks and at that time she developed threatened abortion and was managed accordingly with oral and injectable progesterone. During her 14 weeks pregnancy she was diagnosed as a case of oligohydramnios (single pocket amniotic fluid volume <1.8 cm). AS her previous pregnancy was delivered out by emergency LSCS due sudden severe fetal distress and threatened abortion & oligohydramnios at 13 and 14 weeks of current pregnancy

Authors affiliation

1. Naima Sharmin Hoque, Associate Professor, Department of Obstetrics and Gynaecology, Z H Sikder Women's Medical College, Dhaka.
2. A T M Tanvir, Associate Professor (Rheumatology), Green life Medical College and Hospital
3. Nurun Nahar Fatema, Professor (Brig. General) and Paediatric and Neonatal Interventional Cardiologist, Lab Aid Cardiac Hospital.
4. Mohammad Shakhawat Alam, Associate Professor, Department of Paediatric Cardiology and Neonatology, BSMMU
5. Shariful Islam Seraji, Assistant Professor, Department of Anaesthesiology, Holy family Red Crescent Medical College and Hospital.

Address of Correspondence: *Dr. Naima Sharmin Hoque, Associate Professor, Department of Obstetrics and Gynaecology, Z H Sikder Women's Medical College, Dhaka. Mobile: +8801711184896. Email: drsharminhoque@gmail.com

respectively, she was advised to do ANA (Anti-Nuclear antibody) and Lupus anticoagulant. ANA screening test was strongly positive. She was then referred to Rheumatologist and she was diagnosed as a case of SLE with Sjogren Syndrome at her 16 weeks of pregnancy. On her evaluation Anti SSA (Ro) was positive and Anti SSB, Anti β2 Glycoprotein IgM & IgG, Anti-cardiolipin antibody IgM & IgG were Negative. She had history of dry eyes and positive Schirmer test on both eyes. HCV infection was excluded and there was no risk factor for HIV infection. She was managed in a multidisciplinary approach including Obstetrician, Rheumatologist and Endocrinologist.

Rheumatologist advised to do fetal echocardiogram and it was done weekly as the baby had 1st degree heart block. We also take Interventional Paediatric Cardiologist consultation and maintain doing fetal echocardiogram till last week of delivery. Gradually the mother was becoming very high-risk pregnancy. Throughout the pregnancy she was advised to take tab. Hydroxychloroquine 400mg daily (started from 23 weeks of gestation), tab. Dexamethasone 4mg daily up to 30 weeks then 6 mg daily till delivery. From 30 weeks onward, she required both high dose of short acting insulin and intermittent acting insulin and doses were increasing accordingly as she was under steroid supplementation.

Upto 34 weeks we continue tab. Ecosprin 75mg from very early weeks of pregnancy. On her serial ultrasound fetal weight was not increasing satisfactorily from 32 wks of pregnancy. During 32 wks EFW was only 1762gms (±10%) with grade II maturity placenta. We terminate the pregnancy by emergency LSCS at 37 weeks of gestation when EFW was 2379gms (±10%) with grade III placenta due to 2nd gravida with fetal distress with SLE with Sjogren Syndrome with DM (controlled with insulin) with history of previous 1LSCS. Birth weight of baby girl was 2.3 kg and her after birth echocardiogram was good and follow up remains. Now the baby is risk free according to the Paediatric Cardiologist.

Discussion

Sjögren syndrome is one of the most common autoimmune diseases, with a reported prevalence between 0.1% and 4.8%.5 It may occur at any age but affects mainly women at the fourth decade of life; the female-male ratio is estimated at 9:1.6 Affected women are likely to experience more complicated pregnancies than are women without the disease.[7,8]

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.[9]

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) Myocarditis; 2) Binding apoptotic cells, blocking presumed physiologic clearance, and diverting clearance to macrophages; and 3) Producing arrhythmia.[10,11]

Sjögren syndrome sometimes complicated by pulmonary hypertension, which frequently worsens during pregnancy and in the postpartum period.

Several studies have reported an increased rate of spontaneous abortion and fetal loss associated with Sjögren syndrome (Table 1)[7,12,13,14]

Table No.1: Pregnancy outcomes in women with Sjögren syndrome

Author	Study design	Pregnancies, no.	Spontaneous abortions, no. (%)	Still births, no. (%)	Induced abortions, no. (%)	IUGR, no. (%)	Premature deliveries, no. (%)	Live births, no. (%)	Congenital heart block, no. (%)
Skopouli et al,1994	Retrospective study with questionnaire	207	18 (9)	3 (1.5)	75 (36)	NR	2(1)	111 (54)	2 (1)
Siamopoulou-Mavridou et al,1988	Retrospective study with questionnaire	63	13 (21)	2 (3)	NR	NR	0	48 (76)	NR
Julkunen et al,1995	Retrospective study with records and interview	55	10 (18)	1 (2)	NR	1 (2)	1 (2)	44 (80)	NR
Priori et al,2013	Case-control delivery registry linkage study	45	4 (9)	0 (0)	1 (2)	NR	6 (13)	40 (89)	2 (4)
Takaya et al,1991	Retrospective study with questionnaire	39	2 (5)	0 (0)	9 (23)	NR	1 (3)	28 (72)	NR
De Carolis et al,2014	Electronic case records review	34	10 (29)	0 (0)	1 (3)	1 (3)	9 (27)	23 (68)	2 (6)

IUGR = intrauterine growth restriction; NR = not reported.

A significant increase in the rate of preterm deliveries was found in pregnant women with pSS in most of the studies.15,16 The mean neonatal birth weight was significantly lower in the offspring of women with pSS. This may be related to a pathologic intrauterine growth restriction. The mean neonatal birth weight was significantly lower in the offspring of women with pSS. This may be related to a pathologic intrauterine growth restriction and is not influenced by the timing of the delivery.13 An increased frequency of cesarean delivery was observed in patients with Sjögren syndrome.16 This might be caused by an increased risk of severe fetal outcomes in pregnancies in the Sjögren syndrome population resulting from an increased risk of fetal growth restriction. In the presenting case fetal weight was restricted from 32 weeks and birth weight was not increasing according to the expected way. We delivered the baby at 37 weeks.

Well-known fetal outcomes in Sjögren syndrome-complicated pregnancies are neonatal lupus and congenital heart block (CHB).17 CHB is the most severe fetal complication and 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.18,19,20 The incidence of neonatal lupus in an offspring of a mother with anti-SS-A antibodies is estimated at approximately 1% to 2%.21 Fetus of this case was suffering from first degree heart block and monitored weekly by fetal echocardiogram.

In our presented case she had history of dry eyes and positive Schirmer test (2mm in both eyes) plus positive anti-SS-A antibody suggestive of Primary Sjogren Syndrome as per 2016 ACR-EULAR criteria. DM was trying to control with gradual rising of insulin and was not responsible for hour dryness of eyes. HCV infection was excluded and there was no risk factor for HIV infection.

Conclusion:

Most women with Sjögren's will conceive and deliver healthy babies. However, there are potential complications. It is recommended that women consult with their obstetrician (OB-GYN), Rheumatologist prior to conceiving or early in pregnancy, and continue to monitor throughout.

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