Case Report

A rare case of Sjögren's syndrome in pregnancy and its perinatal outcome- Fetal congenital heart block

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ABSTRACT

Sjögren's syndrome is an autoimmune disease that commonly affects women after the fourth decade of life. Management of pregnancy in women with Sjögren's syndrome poses unique challenges due to potential maternal and fetal complications. Children born to women with Sjögren's syndrome can have a higher incidence of prematurity, low birth weight, hypoxic-ischemic encephalopathy, and congenital heart block. We describe the case of 24-year-old primi at 36 weeks of gestation with Sjögren's syndrome and hypothyroidism, resulting in congenital heart block in the fetus. In this case report, we will discuss the management and perinatal outcome of pregnancy in the case of Sjögren's syndrome.

Key words: Autoimmune disease, Congenital heart block, Pregnancy, Sjögren's syndrome

Sjögren's syndrome is an autoimmune disease that usually affects women in the age group 40–60 years. The disease is characterized by an impairment of secretory gland function, affecting the oral cavity, joints, and eyes, as well as, other body tissues. Laboratory diagnosis reveals increased antinuclear antibodies, anti-Sjögren's-syndrome-related antigen A (anti-SS-A/Ro), and anti-Sjögren's-syndrome-related antigen B (anti-SS-B/La) antibodies. In pregnant women with Sjögren's syndrome, these antibodies can cross the placenta at 12 weeks of gestation and induce myocarditis and arrhythmias by acting on fetal myocardial tissue [1]. Thus, the disease can affect fetal development and increase the risk of adverse outcomes such as miscarriage, congenital heart block, and preterm delivery [2]. The incidence of adverse outcomes of pregnancies in patients with Sjögren's syndrome has been reported to be around 9% [3]. Congenital heart block specifically has an incidence of around 2% for the first pregnancy, with a recurrence rate of nearly 20% in future pregnancies [4]. It may also result in multiple pregnancy-associated comorbidities, such as preeclampsia and premature rupture of membranes, and increase the incidence of complications, such as post-partum deep vein thrombosis [5].

Sjögren's syndrome is a relatively rare condition, and its associations with pregnancy-related complications have not been widely studied. To date, there is limited literature about the prevention and management of adverse fetal outcomes in such pregnancies. Thus, each reported case can greatly help improve the understanding of the disease. Risk factors can be identified, and diagnostic techniques can be improved to ensure timely intervention. Adverse sequelae like congenital heart block can result in permanent neonatal complications or death, hence making early diagnosis and intervention critical. Reporting cases with adverse outcomes increases healthcare providers' awareness and allows them to monitor these pregnancies better.

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CASE REPORT

A 24-year-old primigravida was admitted to the hospital at 36 weeks gestation because of fetal bradycardia (fetal heart rate [FHR] of 70 beats/min) detected on antenatal scanning. Fetal echocardiography revealed cardiomegaly with mild mitral and tricuspid regurgitation and mild ventricular dysfunction with bradycardia (Fig. 1a and b). There was no sonographic evidence of fetal hydrops. The liquor was adequate, and there was no placental abnormality.

The patient's vitals were stable throughout the pregnancy, with a pulse rate between 86 and 94 beats/min, and a baseline blood pressure recording of 110/70 mmHg. Her respiratory rate was around 16 breaths/min, and she was afebrile throughout. She had no clinical features of any autoimmune disorder or any known medical or surgical illness except for hypothyroidism, for which she was on thyroid hormone supplementation.

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She was further investigated and her antinuclear antibodies, anti-SS-A, and anti-SS-B antibody tests were strongly positive. She was diagnosed with Sjögren's syndrome and subsequently, consultations were sought from a rheumatologist, neonatologist, and cardiologist.

The patient was prescribed tablets dexamethasone (4 mg) and hydroxychloroquine (20 mg) once daily, which was to be continued till delivery. She was monitored daily for the FHR which on auscultation varied between 65 and 80/min. As fetal bradycardia persisted, she was counseled regarding the possible need for a pacemaker for the baby post-delivery.

An emergency lower segment cesarean section (LSCS) was performed at 36 weeks, 4 days on account of fetal distress and a non-reassuring non-stress test. At LSCS, the liquor was meconium-stained. A male baby of 2400 g was born with an appearance, pulse, grimace, activity, respiration score of 5 at 1 min and 8 at 5 min of life. The baby cried immediately after birth and was attended to by a pediatrician and conservatively managed in the neonatal intensive care unit.

The baby's heart rate was 50 beats/min. An electrocardiogram (ECG) obtained immediately after birth was suggestive of a complete heart block (Fig. 2a and b). Conservative management was continued for the pre-maturity and low birth weight. Four months post-delivery, the infant was started on diuretics and was advised permanent pacemaker implantation as the ECG showed a persisting complete heart block with a ventricular rate of 48 beats/min.

DISCUSSION

Sjögren's syndrome is an autoimmune disease characterized by antibody-mediated damage of various body tissues, primarily the joints and the secretory glands in the eyes and oral cavity. The antibodies involved in the pathology are anti-Ro/SS-A, anti-La/SS-B, and antinuclear antibodies.

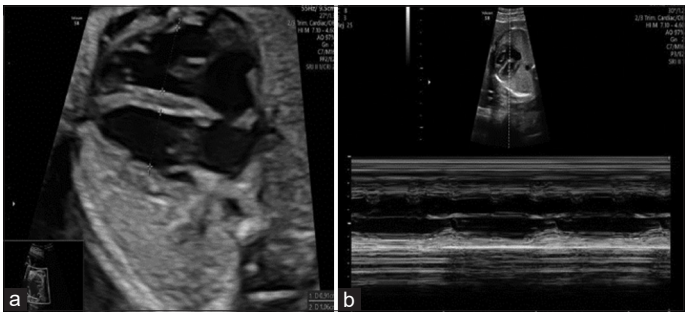

Figure 1: (a) Fetal echocardiography shows cardiomegaly with a dilated right ventricle; (b) Motion-mode image showing fetal bradycardia with a heart rate of 70 beats/min

Pregnancy can impact the progression of Sjögren's syndrome, as around 30% of patients experience an aggravation of the disease during pregnancy, and subsidence after the termination of pregnancy [2]. Women with the antibodies mentioned above may also be asymptomatic at the time of pregnancy, and develop symptoms of autoimmune disease in the future. Pregnant women with Sjögren's syndrome have a higher risk of developing hypertensive disorders, and antepartum hemorrhage, with an increase in overall maternal morbidity (e.g., respiratory failure, cerebrovascular hemorrhage, shock, and cardiac arrest) [6].

Sjögren's syndrome can also have harmful effects on the child, such as neonatal lupus (which includes congenital heart block, skin rash, hepatobiliary or hematologic manifestations), and neonatal death [4]. The occurrence of these effects depends on the antibody titers in the mother [7]. Congenital heart block occurs in children born to 2% of women with positive anti-Ro/SS-A and anti-La/SS-B antibodies and carries a high risk of intrauterine fetal death, neonatal mortality, and long-term sequelae [8].

Guidelines recommend surveillance by serial fetal echocardiography and obstetric sonograms to monitor atrioventricular time intervals during 16–26 weeks of gestation in at-risk pregnancies [9]. The goals are early diagnosis and treatment of incomplete congenital heart block, thus improving the outcome for the fetus. The basis for management is to decrease the maternal auto-antibody levels and their placental transfer and to mitigate the inflammatory response before it leads to permanent fibrosis and irreversible congenital heart block [10]. Hydroxychloroquine is the recommended treatment for all pregnant women with rheumatic diseases and positive anti-Ro/SS-A or anti-La/SS-B antibodies (when there is low disease activity) [9]. It has been found that the use of hydroxychloroquine during pregnancy in individuals with autoimmune diseases significantly reduces the incidence of pre-eclampsia and fetal loss [11].

Maternal treatment with fluorinated corticosteroids, such as dexamethasone or betamethasone, can reduce the antibody-mediated inflammatory damage of nodal tissue [12]. Fluorinated steroids should be administered under supervision as they may produce side effects such as infection, osteoporosis, osteonecrosis, and glucose intolerance in the mother, and intrauterine growth restriction, oligohydramnios, and adrenal suppression in the fetus [13]. The alternative/additional therapies include plasmapheresis, intravenous immunoglobulins, and beta sympathomimetics. In most cases, a complete heart block requires pacemaker implantation in the infant, preferably in the neonatal period [1].

Close prenatal monitoring and collaboration between the obstetrician, rheumatologist, and pediatric cardiologist is essential

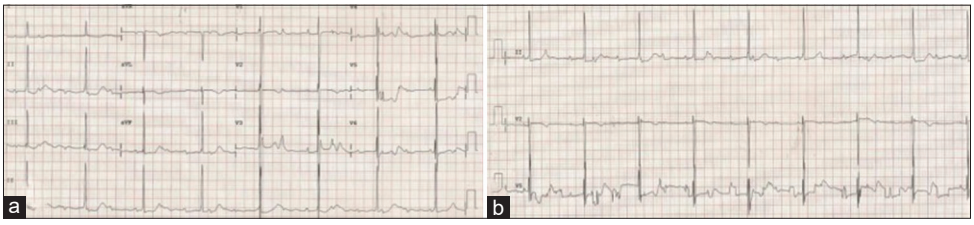

Figure 2: (a and b) Electrocardiogram showing complete heart block and bradycardia with a heart rate of 48 beats/min

for the early detection and management of congenital heart block in affected pregnancies.

CONCLUSION

Sjögren's syndrome can impact both maternal and fetal outcomes. Prenatal screening should be carried out, which includes screening for anti-SS-A and anti-SS-B antibodies, FHR monitoring, and echocardiography. The risks associated with such a pregnancy should be explained to the family. Timely intervention for the condition should be initiated, such as corticosteroid therapy for the mother and subsequent pacemaker implantation for the child. Early detection and multidisciplinary management are crucial for optimizing maternal and fetal outcomes in pregnant women with Sjögren's syndrome.

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