


CASE REPORT

Comprehensive management of pregnant woman with Sjögren's syndrome

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Article Info	ABSTRACT
<p>Received Mar 19, 2022 Revised May 17, 2022 Accepted Jun 20, 2022 Published Dec 1, 2022</p> <p>Corresponding author: Giovanny Azalia Gunawan giogunawan95@gmail.com</p> <p>Keywords: Sjögren syndrome Pregnancy Autoimmune disease Comprehensive management Preeclampsia</p> <p>This is an open access article under the CC BY-NC-SA license (https://creativecommons.org/licenses/by-nc-sa/4.0/)</p> 	<p>Objective: To illustrate the comprehensive management of pregnant women with Sjögren syndrome.</p> <p>Case Report: A 24 years old women came to Dr. Soetomo General Academic Hospital, Surabaya, Indonesia, due to Sjögren syndrome in 35/36 weeks of gestational age for routine examination. She was first diagnosed with Sjögren syndrome in February 2018 because she complained of dry eyes and hair fall out. This was her first pregnancy. Laboratory result showed positive ANA test. The patient had hypertension with controlled blood pressure and dry eyes. The patient was diagnosed with primigravida 35-36 weeks of pregnancy, single live intrauterine, head presentation, IUGR, screening preeclampsia was positive, and there was complication with Sjögren's syndrome. The patient was treated by multidisciplinary team consisting of obstetricians, internists, ophthalmologists and neonatologists.</p> <p>Conclusion: Sjögren's syndrome is a chronic autoimmune disease that attacks the exocrine glands, especially lacrimal and salivary glands. The exact cause of Sjögren's syndrome is still not known. Women with Sjögren's syndrome should have clinical and laboratory examination, risk assessment and also preconception counseling before planning pregnancy because Sjögren syndrome was a rare case during pregnancy. Close monitoring and proper management was imperative to detect the early complication.</p>

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INTRODUCTION

Sjögren's syndrome is a chronic systemic autoimmune disease caused by lymphocyte infiltration in the exocrine glands, the lacrimal and salivary glands.¹ Sjögren's syndrome is divided into primary and secondary. Secondary Sjögren's syndrome occurs with other autoimmune diseases, most commonly are rheumatoid arthritis and systemic lupus erythematosus.²⁻⁵

Sjögren's syndrome is mostly prevalent in women, with a ratio of women and men from 20 : 1 to 9 : 1. It

generally appears in the 4-5 decade, at the age of perimenopause.¹⁻⁵ Immune-mediated inflammation causes secretory dysfunction of the glands and causes sicca symptoms.⁶ The main symptom of Sjögren's syndrome is dryness of eyes caused by lacrimal hypo-function and dry mouth because of hyposalivation.^{4,7} Physical examination will reveal keratoconjunctivitis sicca and xerostomia.¹ The symptoms sometimes had extra-glandular involvement, such as joints, skin, lungs, GI tract, nervous system, and kidneys.

Hormonal and immunological changes during pregnancy can modulate the expression of autoimmune disease.⁸ Autoimmune disease itself will affect maternal and fetal development. Women with Sjögren's syndrome can get pregnant and have healthy babies but it is a good idea to get advice from a specialist because there is risk of complications for some women such as abortion, preterm delivery, preeclampsia and etc. Careful patient management and control is important to detect complications early and provide early therapy.⁹

CASE REPORT

A 24-year-old primigravida at 35-36 weeks of gestational age visited the obstetrics gynecology clinic for routine examination. The patient had no complaints.

Current medical history

The patient was diagnosed with Sjögren's syndrome since February 2018. She complained dry eyes and hair fall out. Since positive for pregnancy, she had routine examination at Dr. Soetomo General Academic Hospital, Surabaya, Indonesia, and received methylprednisolone 4 mg every 24 hours, azathioprine 2 mg every 12 hours, folic acid 1 tablet, aspirin 1 tablet every 24 hours and calc 1 tablet every 24 hours.

This was the first pregnancy for the patient. She had her first menarche at age 13, having regular cycle every month, with duration of menstruation about 5 days. The patient got cramp pain every month before and after menstruation. This was her first marriage and she never used any contraception. There was no family member who had autoimmune disease similar to the patient's.

Physical examination

From vital sign examination, the patient looked healthy with GCS 456. There was no pain with Visual Analog Scale (VAS) score of 0. The patient's body weight was 60 kg, body height 160 cm, with a body mass index of 23.4 kg/m² (normal). From the head and neck examination, there was non-anemic conjunctiva, non-icteric sclera, no cyanosis, no dyspnea. There was no increase in jugular venous pressure nor any enlargement of the neck lymph nodes.

From the chest examination, we obtained symmetrical chest wall movement, without intercostalis or supraclavicular retraction. On cardiac examination, we found single S1 and S2 heart sound, regular, without additional hearts sounds, gallop, or pericardial friction rub. In lung examination, vesicular breath sounds were obtained on both hemithorax, no additional breath sounds, either rhonchi or wheezing, in both lung fields.

Table 1. Vital sign examination

Pre gestational	Blood pressure 135/70 mmHg Pulse 99 x/minute Respiratory rate 22 x/minute Temperature 36.6 °C Saturation 98 % free air
Trimester I	Blood pressure 130/80 mmHg Pulse 91 x/minute Respiratory rate 20 x/ minute Temperature 36.9 °C Saturation 98 % free air
Trimester II	1 st Blood pressure 145/90 mmHg Pulse 88 x/minute Respiratory rate 18 x/minute Temperature 36.7 °C Saturation 99% free air
	2 nd Blood pressure 148/89 mmHg Pulse 90 x/minute Respiratory rate 21 x/minute Temperature 36.4 °C Saturation 99% free air
Trimester III	Blood pressure 140/80 mmHg Pulse 97 x/minute Respiratory rate 20 x/minute Temperature 36.7 °C Saturation 97% free air

From the abdominal examination, we found no surgery scar. From extremities examination, they felt warm, dry, and looked red. There was no edema on both extremities. From obstetric examination, we obtained protuberance abdomen with striae gravidarum. We could palpate the fetus, single live intrauterine with head presentation, fundus height 24 cm, fetal heart rate 144 x/minute, regular rhythm.

Additional examinations

From laboratory examination, we found Hb 12.6 g/dl, Hct 36/6%, leucocytes 4.870/μL, and platelets 226,000, with non-reactive HbsAg, HIV, and syphilis. An immunology panel showed high titers of antinuclear antibody levels 211,60 units (normal range <20 units), normal complement levels, C3 level of 63.1 mg/dL (normal range 50-120 mg/dL), C4 of 20.5 mg/dL (20-50 mg/dL). A diagnosis of autoimmune disease was made.

From obstetric sonography, we found intrauterine single living fetus, approximately 35/36 weeks of pregnancy, placenta on fundus, enough amnion fluid, with estimated fetal weight 2050 g. There was no congenital heart block.

Assessment and management

Based on the history, physical and additional examinations, the patient was assessed with primigravida 35-

36 weeks of pregnancy, single live intrauterine, head presentation, IUGR, positive preeclampsia, and complicated with Sjögren's syndrome. The patient was planned to have fetomaternal sonography routinely to detect any abnormality. The patient was treated by multidisciplinary team consisting of obstetricians, internists, ophthalmologists and neonatologists. The therapy given to the patient was methylprednisolone 4 mg every 24 hours, azathioprine 2 mg every 12 hours, folic acid 1 tablet, calc 1 tablet every 24 hours and aspirin 1 tablet every 24 hours. The patient was planned to have vaginal labor.

Outcome

The baby was born vaginally at 39 weeks with birth weight 2200 g with apgar score 8. There were no complications found in the mother and the baby.

DISCUSSION

Sjögren's syndrome is a chronic systemic autoimmune disease caused by lymphocyte infiltration in the exocrine glands, namely the lacrimal and salivary glands.¹⁰ This syndrome was first discovered by an ophthalmologist from Sweden named Henrik Sjögren in 1933.¹⁹ The exact cause is unknown. Hormonal and immunological changes can occur during pregnancy and affect the patient's autoimmune disease activity.²

Sjögren's syndrome patient tends to experience worsening during pregnancy and post partum mainly due to worsening pulmonary hypertension.²⁰

The most common complications in pregnancy with Sjögren's syndrome include spontaneous abortion, preterm delivery, preeclampsia, and IUGR. Neonatal lupus and congenital heart block (CHB) are the most common complications in the fetus.¹² The incidence of neonatal lupus in mothers with positive anti-SSA is 1-2%.⁴ Prenatal screening of anti-SSA/Ro and anti-SSB/La antibodies is needed before conception or as soon as possible, especially in patients with known Sjögren's syndrome.¹¹

The main symptom of this disease is Sicca syndrome.¹⁷ A person suffering from Sjögren's syndrome will feel dryness on mouth and eye. This is due to the decreasing secretions of the salivary and tear glands that act as moisturizers. In addition to the two symptoms above, there can also be complaints of dry throat and nose, numb tongue, digestive complaints, joint pain and frequent fatigue. Complaints that arise in each individual tends to be more varied.¹³

Women with Sjögren's syndrome should undergo clinical examination, laboratory examination, risk assessment and also preconception counseling before planning pregnancy.

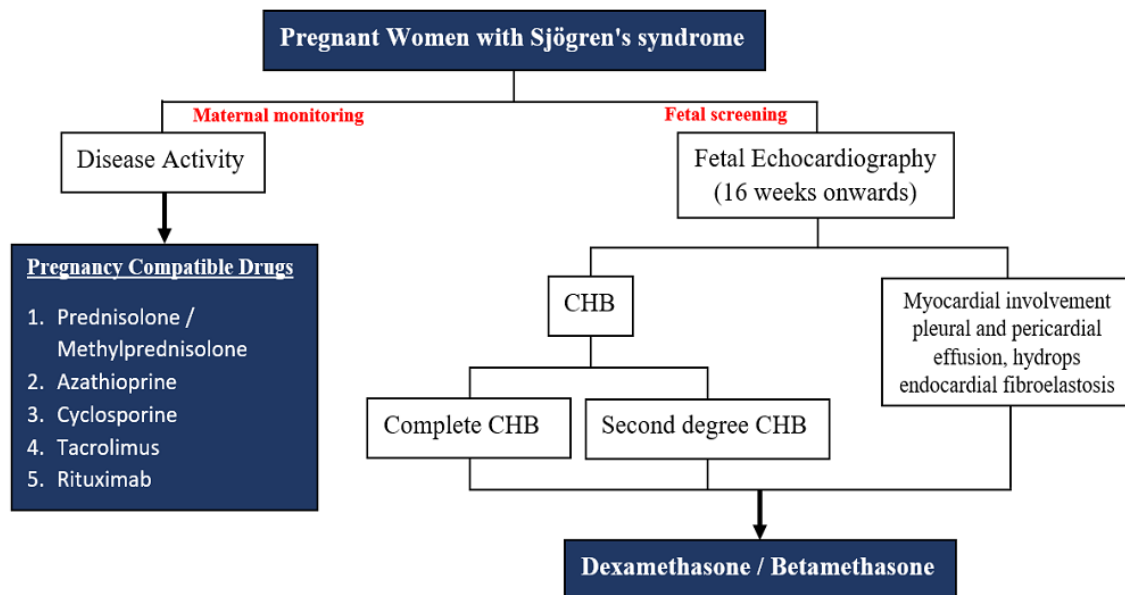


Figure 1. Overview of Sjögren's syndrome management in pregnancy.¹⁴

Ideally, disease activity should be well controlled at least 3- 6 months before conception. Teratogenic drugs such as methotrexate, cyclophosphamide, and mycophenolate should be discontinued at preconception and replaced by other drugs safe for pregnancy. Pregnancy is contraindicated in CKD, severe PAH, severe ILD, and heart failure.^{4,9}

CHB is caused by damage to the AV node by anti-SS-A and/or anti-SSB.¹⁵ Congenital heart block generally occurs at 18-24 weeks of gestation, can appear as fetal bradycardia which can be detected by routine fetal auscultation, ultrasound, or pulsed Doppler echocardiography.¹⁶ There is no guideline on the frequency of examinations, some carry out Doppler examinations for 18-26 weeks and every 2 weeks after 32 weeks of age.¹¹

The therapy is expected to reduce maternal auto-antibodies, thereby reducing placental transfer and reducing inflammation before permanent fibrosis and irreversible CHB occur. Early diagnosis and therapy are expected to improve prognosis, especially in cases of incomplete CHB.⁴ Therapy with fluorinated steroids, such as dexamethasone and betamethasone, can reduce damage caused by inflammation because of antibodies.¹⁸ Thirty percent of cases of second-degree block return to the sinus rhythm after steroid administration. If after 1 week there is progression to grade 3, then the therapy is stopped. There is still a lot of controversy for giving steroid on the 1st degree block. Few studies claim that steroid therapy can reverse to sinus rhythm. These studies are not enough and some considered conservative therapy.¹¹

CONCLUSION

Sjögren's syndrome is an autoimmune disease that attacks the exocrine glands and causes sicca symptoms. The exact cause of Sjögren's syndrome is still not known. The diagnosis begins with anamnesis of the complaints of dry eye and mouth symptoms with examination for antibody levels. Pregnant women accompanied by Sjögren's syndrome has risk of complications such as death of the fetus in the womb, spontaneous abortion, and premature birth. Close monitoring and proper management are imperative to detect early complication. Management of Sjögren's syndrome is supportive and symptomatic to prevent complications. Immunosuppressives such as corticosteroid is one of the therapies for Sjögren's syndrome. As for pregnant women accompanied Sjögren's syndrome, apart from getting corticosteroid therapy, they are advised to do regular pregnancy ultrasound to

determine the presence of organ abnormalities in the fetus (congenital heart block).

DISCLOSURES

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Conflict of interest

All authors have no conflict of interest.

Patient consent for publication

The patient signed the informed consent form and agreed that this case report is published.

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Author contribution

All authors have contributed to all processes in this research, including preparation, data gathering and analysis, drafting and approval for publication of this manuscript.

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