Sjögren’s syndrome associated with antiphospholipid syndrome and fetal myocardial echogenicity: case report

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Summary

Introduction: Sjögren’s syndrome is a rare systemic autoimmune disorder associated with pregnancy (0.3-0.6%). The typical occurrence of anti-Ro/SSA and anti-La/SSB autoantibodies in the maternal serum can modify the perinatal outcome: neonatal lupus and congenital heart block are the most common fetal complications. The disease can manifest in its own right (primary form) or may develop after the onset of other rheumatic disorders (secondary form) (2). We report a case of pregnancy complicated by a secondary form of SS associated with antiphospholipid syndrome and fetal myocardial echogenicity.

Case Report

A 37-year-old pregnant woman at 25 weeks of gestation referred to our Department for threatened premature delivery. She was affected by Sjögren’s syndrome associated with Hashimoto’s thyroiditis and malignant myopia. Due to recurrent miscarriages in her obstetrical history, an antiphospholipid syndrome was diagnosed three years ago. At presentation, her management regime consisted of dalteparin sodium (Fragmin Pharmacia Italia) and folic acid. On admission, physical examination revealed a fairly good general state, a blood pressure of 130/85 mm Hg, pulse 90 beats min, with a BMI of 32, and body temperature at 36.4 °C. Abdominal examination evidenced a uterus equivalent to gestational age, a fetus in cephalic presentation and a normal fetal heart rate. Due to the presence of regular uterine activity and a modified cervix, the protocol with tocolytics and corticosteroid to prevent fetal distress was started. The woman and her husband were not consanguineous and there was no family history of congenital malformations. The conception of the pregnancy was unplanned. There was no history of drug and alcohol use and no recent infections. TORCH serology was negative and the patient had undergone first-trimester maternal serum screening for Down syndrome with a result within the normal range. A routine second trimester scan performed in a private surgery showed a male fetus with regular development of all the examined organs and biometry equivalent to gestational age. During hospitalization an ultrasound examination confirmed normal fetal anatomy but equivalent to 23 weeks of gestation not concordant with referred amenorrhea. Oligohydramnios was also noted and at careful evaluation markedly echogenic ventricular walls were evidenced (Fig. 1). This rare ultrasonographic sign was considered suggestive of fibrosis following transplacental crossing of anti-Ro/SSA antibodies. Due to the association of SS with atrioventricular heart block, a fetal ultrasonographic fol-
low up was started: this anomaly was never found and no episodes of bradyarrhythmias were detected (3). After careful consideration, a trial of dexamethasone 4 mg per os was initiated to prevent a congenital heart block. However, in consideration of the presence of antiphospholipid syndrome, a leg Color Doppler examination was performed and it evidenced a partial deterioration of the left femoral vein due to a previous thrombotic event. No new thromboembolic events were recorded. After two weeks, because of the ending of clinical symptoms, the pregnant woman was discharged: a specific maternal and fetal follow up was established fortnightly and the therapy with antithrombotic and corticosteroids was continued throughout the pregnancy. No prenatal regression of myocardial echogenicity was noted in the subsequent controls. At 36 weeks of gestation, the patient was again admitted to our Department for the premature rupture of membranes. Subsequently, due of the onset of labor, the decision for delivery was taken but the vaginal route was excluded because of the severe myopia. A caesarean section was performed and a newborn of 2070 g was delivered with Apgar score of 7 and 9 at the first and the fifth minute respectively. Maternal post operative course was uneventful. The newborn underwent a cardiac follow up: electrocardiogram in the first day after birth was performed to evidence eventual episodes of bradyarrhythmias, followed by echocardiography before discharge. Both examinations had a negative results and the mother and baby were discharged four days after.

Discussion

Some studies reported in literature show that the association of systemic autoimmune diseases and pregnancy may modify the outcome of the disease and the course of pregnancy due to the background immunologic and hormonal processes. As the great majority of patients with autoimmune diseases are young females in their reproductive age, a correct knowledge of this disease and its complications cannot be omitted by physicians. The SS is characterized by lymphocytic infiltration and destruction of the exocrine glands resulting in uncomfortable symptoms (dry mouth and eyes, gastrointestinal and urogenital discomfort), reduced health-related quality of life, disabling fatigue and increased risk of death due to non-Hodgkin’s Lymphoma (4). SS can manifest in its own right or may develop after the onset of rheumatoid arthritis, systemic lupus erythematosus, scleroderma, primary biliary cirrhosis (3). The disease remains fundamentally incurable and treatment is mainly symptomatic. Pregnancy outcome in SS has not been extensively studied, but has in general not been considered to be associated with impaired fetal outcome, although some studies have reported an increased rate of spontaneous abortion and fetal loss in pregnancies affected by SS (5). Well recognized fetal complications in SS are neonatal lupus and congenital heart block. Our case is explicative of secondary form of SS for the contemporary presence of Hashimoto’s thyroiditis and antiphospholipid syndrome. This syndrome characterized by the presence of venous or arterial thromboembolism and persistent laboratory evidence of antiphospholipid antibodies, is one of the diseases associated with the most severe thrombotic risk. Recurrent miscarriage, early delivery, oligohydramnios, prematurity, intrauterine growth restriction, fetal distress, fetal or neonatal thrombosis, pre-eclampsia/eclampsia, and HELLP syndrome are also hallmarks of antiphospholipid syndrome. Many of these complications are found in our case, but the most important sign evidenced at ultrasonographic evaluation was a myocardial hyperechogenicity. The underlying mechanism behind these complications is suggested to be due to anti-Ro/SSA and anti-La/SSB autoantibodies in the maternal serum, that yet from the early second trimester reach the fetal circulation by active transport across the trophoblast. Antibodies binding the myocardium initiate a process that starting from inflammation leads to tissue destruction and subsequent fibrosis. Interruption of conducting tissue may produce AV block while myocarditis is a possible consequence of autoantibody-mediated inflammation. In our report a fetal myocarditis was suspected by the presence of markedly echogenic ventricular walls and a trial of dexamethasone was initiated in uterus to prevent a congenital heart block and continued throughout the perinatal period.

Conclusions

In conclusion, increased attention must be paid to pregnancies associated with autoimmune disorders, since careful ultrasonographic and clinical monitoring
and preventive treatment with corticosteroids could minimize severe and common fetal complications.

Disclosure of interests

There are not conflicts of interests.

References