FETUS' SLOW HEART BEATS SOLVING A MOTHER'S COMPLAINT: A CONGENITAL CARDIAC BLOCK AS A CLUE FOR A MATERNAL AUTOIMMUNE DISEASE

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BACKGROUND

Neonatal lupus is a rare autoimmune disease characterized by passive transfer of antibodies such as anti-Ro/SSA or anti-La/SSB across the placenta to the fetal circulation causing clinical manifestations in the neonate, regardless of maternal illness. The most serious complication is congenital heart block (CHB). The disease causes 80–95% of the severe atrioventricular blocks in the neonatal period. Treatment varies according to the degree of block, despite limited data. Second-degree CHB is treated with fluorinated corticosteroids and/or intravenous immunoglobulin. Complete blocks are not reversible and there is no indication for therapies, except when there is extranodal disease. Most pregnant women are asymptomatic at diagnosis of CHB. Sjögren's syndrome was diagnosed during pregnancy or less than 12 months after delivery in 14% of mothers in a case series of 49 women with fetuses with CHB.

CASE REPORT

A 37-year-old woman (gestational age of 31 weeks) was admitted with a history of complete CHB since 18 weeks, in addition to severely restricted intrauterine growth (< 3rd percentile). Laboratory workup showed positive anti-Ro in high titers (> 240 U/mL), positive anti-La (18 U/mL), positive ANA (nuclear large/coarse speckled 1/320), normal serum complement levels and negative antiphospholipid antibodies. Dexamethasone 4 mg/day was started at 20 weeks in an outpatient service due to autoimmune CHB. Patient reported previous uneventful pregnancy 4 years before and recent diagnosis of subclinical hypothyroidism. She also reported xerophthalmia for 10 years, under chronic use of lubricating eye drops. There were no other complaints or laboratory alterations. On ophthalmologic examination, a 0 mm Schirmer's test bilaterally, normal salivary flow, anterior and posterior blepharitis 3+/4, tear film break time of 3 s in right eye and 2 s in left eye, bad tear film distribution, lacrimal meniscus absent 3 min after fluorescein instillation and Inferior punctate keratitis in both eyes. Minor salivary gland biopsy showed chronic lymphocytic sialadenitis (focus score of 2). Therefore, she was diagnosed with primary Sjögren's syndrome, prescribed lubricating eye drops every hour and progressive reduction of corticosteroids (complete CHB without extranodal disease).

CONCLUSION

This case reported highlights that identification of fetal atrioventricular block raises the need for investigation of maternal autoimmune diseases.

KEYWORDS

Sjögren's syndrome, Congenital heart block, Neonatal lupus, Anti-SSA/Ro, Anti-SSB/La.