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Autoantibodies to SS-A/RO in an Infant with Congenital Heart Block

C.P. Contractor

S.D. Mehta

S.M. Khambadkone

K.M. Dixit

Simin F. Irani

Neonatal lupus erythematosus is a rarely reported condition characterized by congenital heart block or transient cutaneous lupus or both(1). Mothers of these infants may have (connective tissue diseases) or may be asymptomatic, but consistently have circulating antibodies to SS-A/RO antigen.

Case Report

A new-born-girl was born to a third gravida mother by emergency cesarean section

at 38 weeks gestation. During labour, the fetal heart rate varied between 50 and 70 beats per minute. Examination at birth showed a full term baby weighing 2.95 kilograms, with a length of 49 cm and head circumference of 33 cm. The heart rate was 62 beats minute with all peripheral pulses well felt. The blood pressure was 60/45 mm of Hg in the right arm. Auscultation of heart was normal. There was no skin rash and the rest of the systemic examination was normal. Routine hematological investigations were normal. The chest roentgenogram was normal. The electrocardiogram revealed a complete heart block with an artial rate of 140/minute and ventricular rate of 60/minute. The 2 dimensional echocardiography done on the infant showed a structurally normal heart.

The mother did not have any evidence of skin rash, joint pains or urinary complaints. Investigations of the mother revealed normal hematological, hepatic and renal parameters. Serology for connective tissue disorders showed anti-nuclear antibody (ANA), anti-Sm antibody and anti-double standard DNA (anti-ds-DNA) antibody tests to be negative. However, antibodies to SS-A/RO were detected in a titre in 1 : 64. The neonate's blood, when examined, showed antibodies to SS-A/RO in a similar titre.

The neonate was given a trial of intravenous atropine under cardioscopic monitoring with no resultant change in heart rate.

Since the child had no evidence of

From the Division of Neonatology, Department of Pediatrics, K.E.M. Hospital, Bombay 400 012.

Reprint requests: Dr. Simin F. Irani, Professor of Neonatology, Division of Neonatology, K.E.M. Hospital, Parel, Bombay 400 012.

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hypotension or congestive cardiac failure, pacing was not advised. The child was feeding well and was discharged on the eighth day of life. Two months later, the child showed normal motor and mental development, the heart rate was 66/minute with no evidence of cardiac failure or hypotension.

Discussion

Congenital complete heart block is an uncommon disease, occurring in one in 20,000 pregnancies. Neonatal lupus related to transplacental passage of maternal SS-A/RO antibodies is proposed to be the most common cause of congenital heart block(2).

In women with anti SS-A/RO antibodies, the risk of delivering an infant with congenital heart block is three times higher than the 1 in 60 risk of lupus patients in general(3). Nearly 50% of infants with neonatal lupus syndrome show cardiac manifestations, of which isolated congenital heart block is the most common. Structural heart defects are seen in 25% of these patients(4).

The pathophysiology of congenital complete heart block in infants born to mothers with connective tissue disorders is probably immunological due to anti RO(SS-A) and anti La (SS-B) antibodies which are of the IgG class(5). The question arises why the mother's heart is not affected by the same antibodies. This may be due to the fact that the fetal heart is more vulnerable to damage than the mature heart or that it may possess phase specific antigens. Also, blocking antibodies of the IgA or IgM class may prevent an IgG antibody from causing damage to the mother without being transferred to the baby.

Congenital complete heart block is

generally a benign condition. However, persistent heart rate less than 50/min and Stokes Adams attacks carry a poor prognosis(6).

The drugs recommended in treatment are atropine and isoproterenol and if they fail, a pacemaker should be implanted without delay. Various newer modalities of treatment including corticosteroids, immunosuppressants and plasmapheresis have been tried in mothers with connective tissue diseases in an attempt to prevent adverse effects on the fetus.

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