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Case Report

Silent maternal autoimmunity unmasked by neonatal atrioventricular block: a case of neonatal lupus

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ABSTRACT

Neonatal lupus erythematosus (NLE) is a rare autoimmune condition resulting from transplacental transfer of maternal autoantibodies, most commonly anti-Ro/SSA and anti-La/SSB, and may present with cardiac, cutaneous, or hematological manifestations. We report a late preterm male neonate who presented on day one of life with respiratory distress and persistent bradycardia. Electrocardiogram revealed atrioventricular block with 2:1 conduction, while echocardiography showed no structural cardiac abnormalities. Autoimmune evaluation demonstrated elevated antinuclear antibodies, anti-Ro, anti-La, and anti-dsDNA antibodies, confirming the diagnosis of neonatal lupus. The neonate was managed with isoprenaline infusion followed by oral beta-blocker therapy, resulting in stabilization of heart rate. In view of these findings, the asymptomatic mother was evaluated and found to have elevated ANA, anti-Ro, and anti-La antibodies, suggestive of subclinical systemic lupus erythematosus. The infant remained clinically stable on follow-up, with appropriate neurodevelopment and later development of hypopigmented skin lesions, and is planned for pacemaker insertion at a later stage. This case highlights the importance of considering neonatal lupus in cases of unexplained neonatal bradycardia and emphasizes the need for evaluation of maternal autoimmune status even in the absence of symptoms.

Keywords: Neonatal Lupus, Atrioventricular block, Autoimmune disease, Bradycardia

INTRODUCTION

Neonatal lupus erythematosus (NLE) is a rare passively acquired autoimmune condition in neonates, associated with maternal autoantibodies directed against intracellular ribonucleoproteins.[1] The clinical spectrum includes cutaneous lesions, cardiac conduction abnormalities, hepatobiliary involvement, and hematological manifestations. Among these, congenital heart block represents the most severe and often irreversible complication, contributing significantly to morbidity. Importantly, many mothers remain clinically silent, and the diagnosis is often established only after evaluation of the affected neonate. Recognition of this entity is essential not only for neonatal management but also for identifying underlying maternal autoimmune disease and counseling regarding future pregnancies. Here, we report a case of neonatal lupus presenting with cardiac conduction abnormality in a late preterm infant, which led to the detection of previously undiagnosed maternal autoimmunity.

CASE REPORT

A male neonate, born at 36 weeks of gestation with a birth weight of 2.2 kg (appropriate for gestational age), was delivered via elective lower segment cesarean section to a G2P1L1 mother. The antenatal period was uneventful, with no documented maternal illness or prior serological abnormalities.

The neonate was referred to our neonatal unit on day one of life with complaints of respiratory distress. On admission, the baby had mild tachypnea with a heart rate of 89 beats per minute, which is low for age (normal neonatal heart rate: 120–160 beats per minute). Oxygen

saturation was maintained with minimal respiratory support. There were no cutaneous lesions, petechiae, hepatosplenomegaly, or clinical features suggestive of sepsis or encephalopathy. Systemic examination, including cardiovascular and neurological assessment, was otherwise unremarkable.

In view of persistent bradycardia, an electrocardiogram was performed, which revealed second-degree atrioventricular block with 2:1 conduction (Figure 1). A two-dimensional echocardiogram showed no structural cardiac abnormalities.

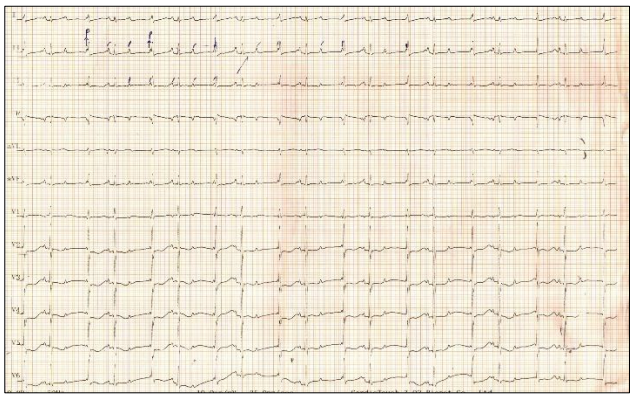

Figure 1: Electrocardiogram of the neonate demonstrating second-degree atrioventricular block with 2:1 conduction (black arrows indicating non-conducted P waves).

Further laboratory evaluation was undertaken to determine the underlying etiology. Autoimmune screening revealed elevated titers of antinuclear antibodies, anti-Ro/SSA, anti-La/SSB, and anti-dsDNA antibodies, consistent with neonatal lupus syndrome.

The neonate was managed in the neonatal intensive care unit with supportive care. Intravenous isoprenaline infusion was initiated due to symptomatic bradycardia, resulting in improvement of heart rate to above 110 beats per minute. After stabilization over three days, the infant was transitioned to oral oxprenolol, with maintenance of stable heart rate and hemodynamic status. Serial monitoring showed no recurrence of significant bradycardia or signs of cardiac failure. The neonate required a total NICU stay of 12 days. In view of persistent conduction abnormality, permanent pacemaker insertion was planned at a later age.

Given the positive autoimmune profile in the neonate, the mother was evaluated despite being asymptomatic. Serological testing revealed elevated antinuclear antibodies, anti-Ro, and anti-La titers. On further enquiry, she reported occasional episodes of photosensitivity during pregnancy, which had not been previously evaluated. A diagnosis of probable subclinical systemic lupus erythematosus was considered.

The infant remained clinically stable and was discharged with advice for regular follow-up. At subsequent visits, the baby demonstrated age-appropriate neurodevelopment. A hypopigmented rash over the lower thigh was noted during follow-up, further supporting the diagnosis of neonatal lupus. Over time, heart rate remained stable on medical management, which is consistent with reported transient improvement in atrioventricular conduction in some cases of neonatal lupus, although long-term pacing may still be required. Relevant laboratory parameters are summarized in (Table 1), and the clinical course during the first 15 days of life is illustrated in (Figure 2).

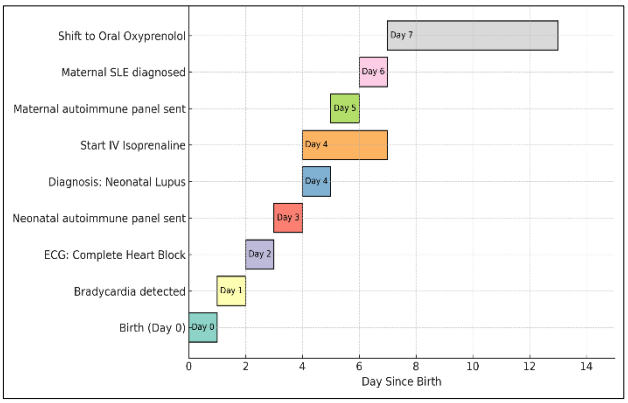

Figure 2: Gantt chart illustrating clinical timeline during first 15 days of life.

Table 1: Serial laboratory parameters demonstrating persistent autoantibody positivity and improvement in heart rate following initiation of therapy.

Parameter	Reference range	Day 1	Day 3	Day 7
Heart rate (bpm)	100–160	89	110	120
ANA titre	<1:80	1:320	1:320	1:320
Anti-Ro	<20	92.50	92.40	92.45
Anti-La	<10	20.64	20.64	20.64
Anti-dsDNA	<10	21.24	21.24	21.24

DISCUSSION

NLE is a passively acquired autoimmune disorder resulting from the transplacental transfer of maternal autoantibodies, predominantly anti-Ro/SSA and anti-La/SSB.[1] These antibodies target fetal tissues, particularly the cardiac conduction system and skin, leading to a spectrum of manifestations including congenital heart block, cutaneous lesions, hepatobiliary dysfunction, and hematological abnormalities.[2] Among these, cardiac involvement represents the most severe complication and is associated with significant morbidity.

Cardiac manifestations of NLE most commonly present as varying degrees of atrioventricular block, typically detected antenatally or in the early neonatal period.[3] The occurrence of conduction abnormalities in the presence of

a structurally normal heart, as observed in our case, strongly supports an immune-mediated etiology. Unlike cutaneous manifestations, which are usually transient, cardiac conduction defects are frequently irreversible and may necessitate long-term pacing.[4]

A key clinical feature of NLE is that a substantial proportion of mothers remain asymptomatic at the time of diagnosis. It is estimated that approximately 40–60% of mothers of affected infants have no prior history of autoimmune disease, with neonatal presentation serving as the first clue to underlying maternal autoimmunity.[5] In the present case, the diagnosis in the neonate prompted evaluation of the mother, leading to the identification of previously unrecognized autoimmune seropositivity, thereby highlighting the sentinel role of neonatal findings.

Management of neonatal lupus with cardiac involvement is primarily supportive. Pharmacological agents such as beta-agonists (e.g., isoprenaline) may be used for temporary stabilization of heart rate in cases of symptomatic bradycardia.[6] However, infants with persistent or significant conduction abnormalities frequently require permanent pacemaker implantation, particularly when associated with hemodynamic compromise or sustained bradycardia.[7] In our patient, initial stabilization was achieved with medical therapy, and pacemaker insertion has been planned at a later stage.

In some infants, partial improvement or stabilization of heart rate may occur over time, possibly due to gradual clearance of maternal antibodies from the neonatal circulation.[8] Nevertheless, such improvement does not eliminate the need for long-term follow-up, as conduction abnormalities may persist or progress.

Importantly, early identification of maternal autoantibody positivity during pregnancy has significant preventive implications. The use of hydroxychloroquine in anti-Ro/SSA-positive mothers has been shown to reduce the risk of congenital heart block and recurrence of neonatal lupus in subsequent pregnancies.[9,10] This underscores the importance of antenatal screening and timely maternal intervention in high-risk pregnancies.

This case highlights the importance of considering neonatal lupus in neonates presenting with unexplained bradycardia, particularly in the absence of structural heart disease. It also emphasizes the need for comprehensive maternal evaluation and the role of preventive strategies in improving outcomes in future pregnancies.

CONCLUSION

Neonatal lupus should be suspected in any neonate presenting with unexplained bradycardia or atrioventricular conduction abnormality in the absence of structural heart disease. This case underscores the pivotal role of the neonate in unmasking occult maternal autoimmunity, even in clinically asymptomatic mothers. It reinforces the need for routine maternal evaluation in such scenarios, as early identification not only guides neonatal management but also enables timely preventive strategies, including hydroxychloroquine therapy, to reduce recurrence risk in future pregnancies.

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