CASE REPORT

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Diagnostic dilemma and Management challenges of Neonatal Lupus with Complete Congenital Heart Block in a resource poor set-up – A case report

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Abstract

Neonatal lupus, distinct entity from systemic lupus erythematosus (SLE), is of the few rheumatic disorders manifesting in the neonate. Congenital heart block is a severe manifestation of lupus with other complications being cytopenia's, cutaneous rash, hepatitis and neuropsychiatric alterations. It occurs as a result of transplacental passage of maternal anti-SSA [Ro], anti-SSB [La] and anti-RNP [ribonucleoprotein] antibodies. Here, we report a case of asymptomatic mother who delivered Lupus baby with complete congenital heart block and was later diagnosed retrospectively as having the autoimmune disease. Therefore, early suspicion, diagnosis and management is very important to save a life but it is a real challenge in a resource poor country like India. Here, we are mainly focussing on the diagnostic dilemmas, management challenges and identifying asymptomatic mothers in a resource poor set-up.

Keywords: Neonatal lupus, Complete congenital heart block, Anti-SSA [Ro] antibody, Anti-SSB [La] antibody.

INTRODUCTION

Neonatal lupus is a passively acquired autoimmune disease which usually persists for the first six months of life, until neonate clears antibodies. Inflammatory manifestations like cytopenias, cutaneous rash and hepatitis also gradually resolves. Congenital heart block is a permanent complication bearing mortality rate of 18% and requiring pacemaker implantation in two-thirds of newborn.[1]

The maternal auto-antibodies transfer occurs as early as 11 weeks but damage to the conducting system occurs only after heart development is complete, evidenced by the lack of structural and anatomical abnormalities in the affected newborn's heart.[2] Ro and La are intra-cellular antigens which are released due to unregulated apoptosis. These antigens are attacked by antibodies leading to activation of immune system causing inflammation, fibrosis and calcification of the conducting system.[3]

The risk of congenital heart block with maternal autoantibodies, mainly Ro antibodies is roughly 2% [4] and the recurrence risk in subsequent pregnancy can reach 19%. Therefore, proper follow-up and pregestational counselling is essential. In mothers with Systemic lupus erythematosus (SLE) or Sjogren's syndrome, the prevalence of Ro antibodies is approximately 40 % and 60 – 100 % respectively.[5]Not all the mothers carrying Ro autoantibodies have the same risk of developing congenital heart block in newborn, several studies have shown that the risk depends on the serum levels of autoantibodies during pregnancy.[6]

The various cardiac manifestations in neonatal lupus includes: heart block, cardiomyopathy, valvular dysfunction and endocardial fibroelastosis. The conduction system abnormalities range from prolongation of PR interval to complete heart block beginning at 16 weeks of gestation detected by in-utero fetal

echocardiogram. The fluorinated corticosteroids [dexamethasone or betamethasone], intravenous immunoglobulins [IVIg], plasmapheresis, terbutaline [beta agonist] or hydroxychloroquine have been used in pregnant women to prevent occurrence of fetal cardiac abnormalities.

CASE REPORT

A primigravida mother with 38 weeks of gestation in labour pains was admitted to our hospital for delivery. Fetal heart rate monitoring showed persistent fetal bradycardia with heart rate around 45 beats per minute, emergency caesarean section was performed and a male newborn, weighing 2.7 kg was delivered. APGAR score was 5 at 1 minutes and 8 at 5 minutes. Newborn was shifted to neonatal care unit.

On examination, cry, reflex and activity were fair, capillary refill time (CRT) was less than 3 seconds, oxygen saturation (SpO2) was 92 % at room air. Chest auscultation revealed bilateral good air entry and respiratory rate of 48 cycles per minute. Cardiovascular system examination revealed heart rate varying from 38 bpm to maximum rise of 52 bpm varying from sleep to crying activity respectively. S1, S2 heard with ejection systolic murmur audible in all the areas. All peripheral pulses were palpable.

Table 1: Investigations IN Newborn

Complete blood count	Haemoglobin: 13 g%, Total Leucocyte Count: 8400/cumm, Platelet count: 2.1 lakh/cumm.
C- reactive protein	7.34 mg/l
Electrolytes	**Na**+: 137mmol/l, **K**+: 4.6 mmol/l
Urea	15 mg/dl
Creatinine	0.4 mg/dl
Liver function test	SGOT: 31U/l, SGPT: 20U/l, alkaline phosphatase: 241U/l, albumin: 3.4 g/dl
Chest X ray	Normal
Arterial blood gas (ABG) analysis	PH: 7.34, pCO2: 34 mmHg. pO2: 108mmHg
2D ECHO with Doppler	Atrioventricular dissociation with slow ventricular rate, patent foramen ovale with bidirectional flow was present and mild tricuspid regurgitation was seen.
Electrocardiogram (**FIG-1**)	Third degree atrioventricular block with narrow complex QRS showing atrial rate of 115 beats per minute and ventricular rate of 45 beats per minute.

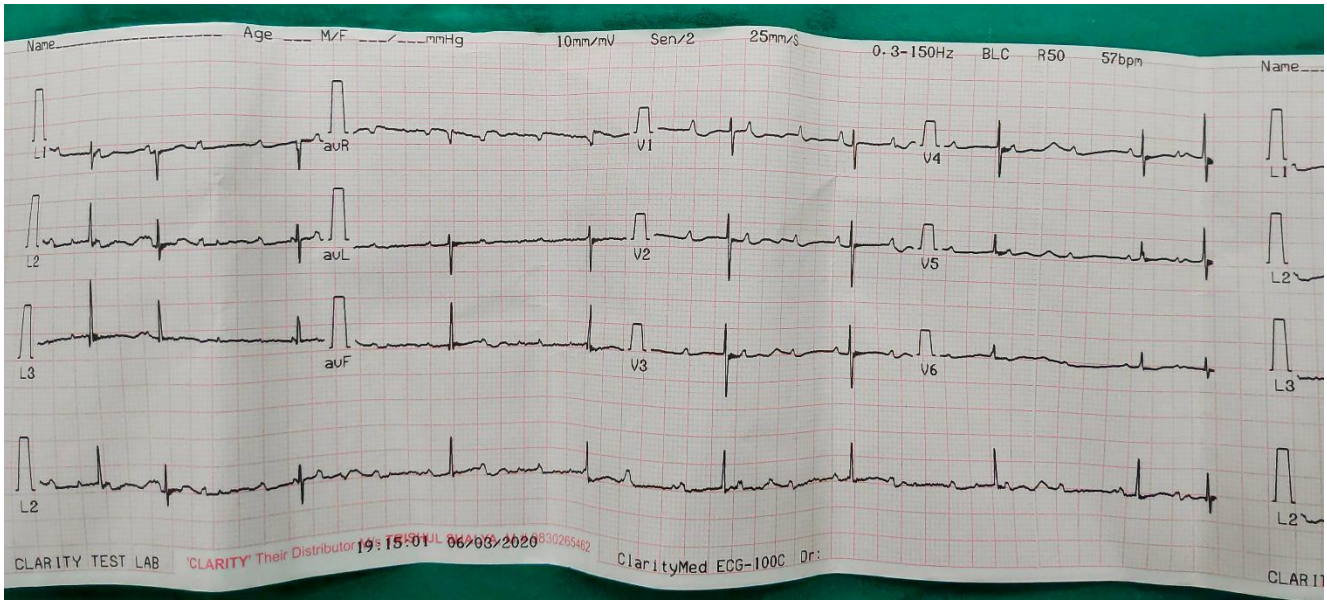

Figure 1: Electrocardiogram

Intravenous injection of atropine and infusion adrenaline was tried to increase heart rate but there was no response.

Maternal history revealed no regular antenatal visits taken. Available documents showed only one antenatal visit in rural hospital during 32 weeks of gestation showing blood pressure recorded of 118/70 mmHg, haemoglobin:10 g %, total leucocyte count of 6200/cumm with 64 % neutrophils and 31% lymphocytes. Urine examination showed absence of albuminuria and proteinuria. TSH was 2.78 mIU/ml [normal]. Antenatal USG scan showed single live intrauterine fetus of approximately 32 weeks 4 days, with fetal heart rate recorded as 157 bpm.

On detailed examination and history taking of mother, she was asymptomatic with no history of skin rash or joint pain. For confirmation of the diagnosis of neonatal lupus following investigations of the mother were done which revealed anti-nuclear antibodies (ANA) levels of 6.15 OD ratio[positive], anti-SSA-Ro IgG titres of 200 RU/ml [positive] and anti-SSB-La IgG titres of 44.1 RU/ml [positive]. The diagnosis of maternal autoimmune disease was done.

As investigations in newborn revealed no cytopenias and hepatitis features, supportive care was given. Cardiac pacing is the definitive treatment and the newborn was referred to the higher centre for the same.

DISCUSSION

The main challenge of managing this condition lies during antenatal period. Review of literature shows that 50% of mothers are asymptomatic, 20-30% have previous diagnosis of autoimmune disease [7] and remaining 20% do not have any auto-antibodies. This shows a large proportion involves mothers who are asymptomatic and early identification in this group is challenging especially in a resource poor set-up. Some of the key points in history for early identification includes: recurrent miscarriage, fetal growth restriction, preterm delivery, recurrent history of arterial and venous thrombosis, pre-eclampsia and neonatal lupus in previous pregnancy. [8] The fetal echocardiography assessments made at 18-20 weeks to rule out complete heart block resulting in fetal bradycardia is an accurate test in asymptomatic mothers.[9]

The routine screening of all the asymptomatic mothers with autoantibodies is not feasible but the identification of above-mentioned key points along with findings of fetal bradycardia in echocardiography necessitates testing for SSA/Ro and SSB/La autoantibodies helping in early identification and management in asymptomatic mothers.

The management challenges are seen in pregnant women with active lupus/lupus nephritis or anti-Ro/La/ antiphospholipid antibodies and should be considered as a high-risk group and managed in centres with appropriate experience.

The diagnostic dilemmas in pregnant women lies with differentiating preeclampsia from auto-immune nephritis. This may be challenging as they have symptoms in common like hypertension, proteinuria, severe headache and visual disturbances.

CONCLUSION

In summary, we report a case of neonatal lupus in asymptomatic mother with autoimmune disease. In developing countries like India, with poor awareness, resources and lack of trained doctors many cases are not diagnosed early during antenatal period where the appropriate treatment to prevent neonatal lupus is possible.

Conflict of Interest

The authors declare that there is no conflict of interest.

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Author's Contribution

All authors have contributed equally to manuscript writings

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