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Case Report

Neonatal Lupus Erythematosus-Beyond Conduction Defects

Prem Alva[1], Shrishail Kumbar[2], Ashvij Shriyan[3], Aswathy Rajan[3]

[1]A.J. Hospital and Research Centre, Clinic of Pediatric Cardiology, Mangalore, India
[2]Sanjeevani Multispeciality Hospital, Clinic of Pediatrics, Bellary, India
[3]A.J. Institute of Medical Sciences, Department of Pediatrics, Mangalore, India

ABSTRACT

Neonatal lupus erythematosus is an uncommon disease; frequently undiagnosed, produced by the transplacental passage of autoantibodies such as anti-Ro/SSA and anti-La/SSB. Here, we discuss a case of a preterm baby with intracardiac calcification and right ventricular dysfunction detected antenatally and managed successfully with steroid and intravenous immunoglobulins.

Keywords: Lupus erythematosus, heart failure, neonatal critical care

Introduction

Neonatal lupus erythematosus (NLE) is a rare disease; frequently undiagnosed, which is caused by the transplacental passage of autoantibodies such as anti-Ro/SSA and anti-La/SSB1. NLE presents with multi-organ involvement but the skin and heart are the main targets. Mothers with anti-Ro/SSA and anti-La/SSB antibodies face a 2% risk of having a baby with myocardial calcification, cardiac dysfunction and heart blocks (1).

Case Report

A late preterm (36 weeks), appropriate for gestational age, male baby was delivered with a birth weight of 2,690 kg to a 31 year-old mother who was diagnosed as having systemic lupus erythematosus seven years prior to conception, which was confirmed by anti-Ro and anti-La antibodies. She was treated with oral prednisolone for one year initially and then started on oral hydroxychloroquine, which was continued. The initial ultrasound scan in the present pregnancy was carried out at four months of gestation and was normal. A second scan carried out at 32 weeks of gestation showed foetal cardiomegaly with pericardial effusion. Foetal echocardiography (ECHO) showed findings of a dilated right atrium (RA), right ventricle (RV) and multiple intracardiac calcifications predominantly in the right and left papillary muscles, and mild pericardial effusion with mild RV dysfunction.

In view of the foetal cardiac involvement, hydroxychloroquine was stopped and oral betamethasone was given for 2 weeks at a dose of 3 mg/day. Repeat foetal ECHO carried out at 36 weeks of gestation showed multiple intracardiac calcifications, grossly dilated RA & RV with significant RV dysfunction. Hence, the mother was advised to have an early delivery in order to avoid further complications.

Address for Correspondence
Aswathy Rajan, A.J. Institute of Medical Sciences, Department of Pediatrics, Mangalore, India
Phone: +91 812 355 09 52 E-mail: ar_aswathy@yahoo.com ORCID: orcid.org/0000-0002-5568-2569
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The baby was born by normal vaginal delivery and was placed in the neonatal intensive care unit due to tachypnea. ECHO carried out on day 1 of life showed a large patent ductus arteriosus (5 mm) with bidirectional shunt, dilated RA, RV with RV dysfunction and mild pericardial effusion (Figure 1). The baby was started on intravenous furosemide, dobutamine and oxygen support. Electrocardiogram (ECG) showed first degree heart block.

Initial haemoglobin, white blood cell counts and platelet counts were within normal limits. Liver enzymes were normal: Anti-Ro: >240 U/mL, anti-La: 0.7 U/mL and anti DNAse <10 IU/mL. As we noted significant cardiac lesions with ventricular dysfunction, intravenous immunoglobulin (IVIG) 1 g/kg/day was given. At 48 hours of life, the tachypnea decreased, however significant tachycardia persisted. At 72 hours of life, repeat ECHO showed patent foramen ovale with left to right shunt, dilated left atrium (LA) & left ventricle (LV) and mild RV. The LV dysfunction, LV ejection fraction was 45% with no pericardial effusion. Due to the low ejection fraction, syrup digoxin was started at a dose of 10 mcg/kg. Repeat ECHO on day 5 showed multiple intracardiac calcifications, patent foramen ovale with left to right shunt, mildly dilated LA & LV, fair LV function with good RV function. Clinically, the baby was stable and so the baby was moved to the mother's bedside on day 6 of life with oral digoxin and furosemide. On follow-up, ventricular function returned to normal by 3 months of age and the child was asymptomatic. All cardiac medications were stopped in view of this. However, the ECG was still showing a first degree heart block.

Discussion

Cardiac manifestations, being the second most common presentation in NLE, usually include conduction abnormalities and cardiomyopathy which may lead to heart failure in the new-born period (1). Cardiac damage in the form of calcification and collagen deposition secondary to existing blocks may be seen. In addition, damage to the valves and valve apparatus, which includes fibrosis and calcification of the papillary muscle, myocarditis, pericardial effusion and endocardial fibroelastosis are also known to occur (2).

Very few studies have shown findings beyond the conduction system. Cuneo et al. (3) reported two cases of atrioventricular valve insufficiency due to chordal rupture from the papillary muscles in a 34 weeks of gestation foetus and a 6-month old infant born to mothers who tested positive for anti-Ro 52 antibodies. Morais et al. (4) described a male infant born to a mother who had unexplained fever and vasculitic lesions on her extremities and face as having hyperechogenic lesions on the anterior papilar muscle of the LV and on the lateral cusp of the tricuspid valve. The occurrence of dilated cardiomyopathy has been reported to be between 5 to 28.6% in babies with NLE. The spectrum of neonatal lupus now includes many cases which have described the occurrence of endomyocardial fibroelastosis (5). Carolina Llanos et al. (6) conducted a study of 18 lupus autopsy cases. Three of these cases showed findings of papillary muscle fibrosis, microcalcification of the atrial septum, soft tissue adjacent to the atrioventricular node and ventricles, dilated atria and ventricles, valvular disease, pericardial effusion and endocardial fibroelastosis.

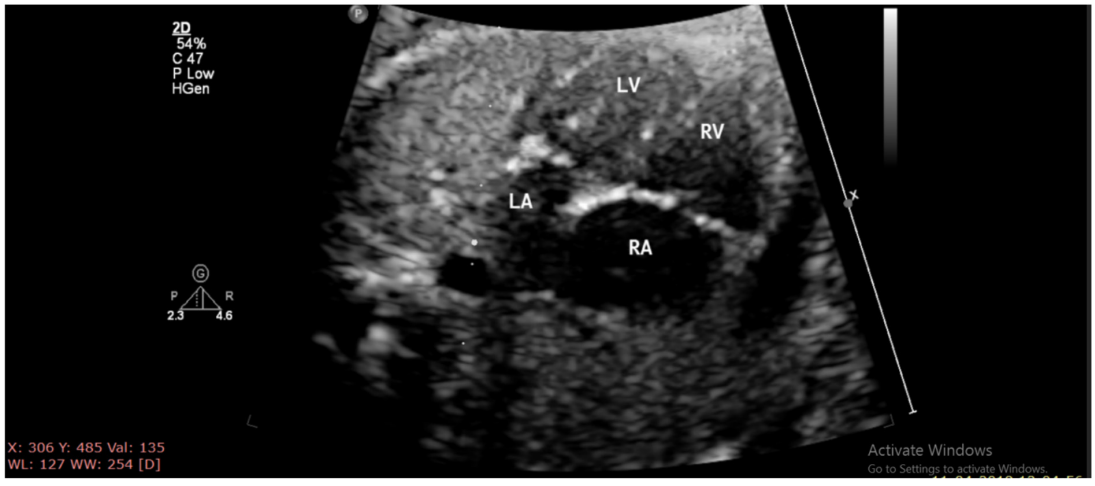

Figure 1. Echocardiogram on day 1 of life showing a 5 mm patent ductus arteriosus, dilated RA, RV with RV dysfunction and mild pericardial effusion
LA: Left atrium, LV: Left ventricle, RV: Right ventricle, RA: Right atrium

In this present case, foetal ECHO carried out at 32 weeks of gestation showed multiple intracardiac calcifications mainly in the papillary muscles with mild pericardial effusion and mild RV dysfunction. The mother was started on corticosteroids based on a recent study conducted by Buyon et al. (7) which showed that antenatal steroids help in preventing cardiac damage or its progression. Repeat ECHO at 36 weeks of gestation showed progressively dilated RA and RV, significant RV dysfunction in addition to multiple intracardiac calcifications. Post-delivery, the baby was started on inotropes and diuretics in view of the significant right ventricular dysfunction. In order to reduce further cardiac damage by transplacentally transferred maternal anti-Ro and anti-La antibodies, IVIG transfusion was given. Post IVIG infusion, an improvement in clinical status and ventricular function was noted along with resolution of the pericardial effusion. The use of IVIG has been studied alone or along with dexamethasone or plasmapheresis (8). Trucco et al. (9) observed improvement in ventricular systolic function in eleven neonates who received IVIG within the first few days with or without steroids. IVIG therapy is more beneficial if it is given early in the pregnancy. More importantly, waiting to provide IVIG until after birth may eliminate the added benefits of reducing maternal antibody exposure and myocardial damage in utero. However, a study by Alsaleem. (8) showed antenatal use of low dose IVIG at 400 mg/kg did not prevent the recurrence of heart block in a series of babies. More studies and data are needed to understand the spectrum of cardiac involvement in terms of ventricular dysfunction, multiple calcifications other than heart blocks and the role of IVIG in post-natal life.

Babies with lupus erythematosus need regular follow-up. Maternal anti-Ro/La antibodies usually disappear by 6 months of life. These babies can develop bradycardia, prolonged page rank interval, cardiomyopathy and/or heart block so serial monitoring by ECG and ECHO is necessary (1). NLE with cardiac involvement is associated with 20-30% mortality in neonatal life. In 10% of cases, transient ECG alterations were found during follow-up. Persistent anti-Ro/La antibodies warrant follow-up until adulthood. Additionally, the rate of subsequent pregnancies being affected is 12-25% (10).

Conclusion

NLE with cardiac involvement is a serious condition and therapy should either be targeted to eliminate the necessary factor (no antibody, no disease) or to modify the inflammatory component before it provokes irreversible cardiac damage. Combined steroid and IVIG therapy shows promising benefits.

Ethics

Informed Consent: Informed consent was taken prior to writing this case report from the patient's parents.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: P.A., A.S., Design: S.K., A.S., A.R., Data Collection or Processing: P.A., S.K., Literature Search: P.A., A.R., Writing: P.A., S.K., A.R.

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