

BACKGROUND

Neonatal lupus erythematosus (NLE) is a passively acquired autoimmune disease caused by the transplacental passage of maternal SSA/anti-Ro and SSB/anti-La antibodies who may have a diagnosis of Systemic Lupus Erythematosus (SLE) and/or Sjogren's Syndrome (SS) or be unaffected carriers of these antibodies. NLE is associated with discoid rash and congenital heart block. Less recognized features include brain, liver and hematologic abnormalities. More severe lab abnormalities and cardiac involvement may require close monitoring and treatment.

PURPOSE

Review NLE cases from Texas Children's Hospital over a 10-year period and describe clinical features in affected infants.

METHODS

- EPIC SlicerDicer tool
- Searched Term: diagnosis- neonatal lupus erythematosus
- Search Dates: 1/1/2011 - 11/30/2021
- Retrospective chart review
 - 39 patients
 - 4 excluded: 2 inadequate charting, 1 diagnosed at other institution, 1 patient not yet born
- Data Collected:
 - Sex
 - Ethnicity
 - Age at presentation
 - Maternal diagnosis
 - Rash findings
 - Lab abnormalities
 - EKG results
 - Development of future autoimmune disease

RESULTS

- A total of 35 patients were evaluated
- Male 49% (n=17), Female 51% (n=18)
- Ethnic distribution:
 - African America 34.3% (n=12)
 - Caucasian 28.5% (n=10)
 - Hispanic 20% (n=7)
 - Asian 11.4% (n=4)
 - Biracial 5.7% (n=2)
- Median Age of presentation: 6 weeks
- 21/23 infants tested had SSA/anti-Rho, SSB/anti-La or anti-Smith antibodies

Maternal Diagnoses	Frequency
SLE	37.1% (n=13)
Sjogren's Syndrome	11.4% (n=4)
SLE + Sjogren's Syndrome	14.3% (n=5)
Other Autoimmune Disease	2.8% (n=1)
No Hx Autoimmunity	34.3% (n=12)

Fig 1: Maternal Diagnoses

NLE Symptoms	Frequency
Rash	94.2% (n=33)
Neutropenia	28.6% (n=10)
Anemia	5.7% (n=2)
Thrombocytopenia	2.8% (n=1)
Complete Heart Block	11.4% (n=4)
1 st Degree Heart Block	5.7% (n=2)
Transaminitis	37.1% (n=13)
Obstructive Hydrocephalus	2.8% (n=1)

Fig 2: Frequency of NLE Symptoms/Complications



Fig 3: Discoid rash (face) at 6 weeks of age, develops on sun-exposed areas



Fig 4: Discoid rash (abdomen) at 6 weeks of age

DISCUSSION

While NLE is associated with maternal SLE and SS, approximately 1/3 of mothers did not have an autoimmune diagnosis. NLE is associated with discoid rash and congenital heart block. In our patient population, heart block was noted in 6 (18%) patients. Complete heart block occurred in 4 patient requiring pacemakers. A ventricular assist device occurred in 1 patient awaiting cardiac transplantation. The most common clinical manifestation of NLE in our cohort was skin rash in 33 (92%) of patients. Rash frequently develops at ~6 weeks of age from sun exposure, or sooner if phototherapy is required. NLE skin rash is self-limited and can be mistaken for fungal infections or eczema. Five patients required diagnostic skin biopsies. Less commonly recognized manifestations that occurred in our cohort included neutropenia (28.6%), anemia (5.7%), thrombocytopenia (2.8%) and transaminitis (37.1%). Two patients went on to develop future autoimmune disease: one with Juvenile Idiopathic Arthritis and another with Autoimmune Neutropenia.

CONCLUSION

NLE is an uncommon diagnosis and can be devastating in patients with complete heart block. Discoid rash is a common presentation, but blood counts, liver enzymes and neurologic evaluation should be considered. NLE should be considered in an infant with discoid rash and/or heart block even in the absence of maternal SLE or SS. Most patients do well with symptoms resolving by their 1st birthday. Patients with history of NLE may be at risk of developing future autoimmune disease and may benefit from monitoring.

REFERENCES

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2. Hon KL, Leung AK. Neonatal lupus erythematosus. *Autoimmune Dis*. 2012;2012:301274. doi:10.1155/2012/301274