NEONATAL LUPUS ERYTHEMATOSUS (NLE): REVIEW OF CASES AT TEXAS CHILDREN'S HOSPITAL OVER A 10-YEAR PERIOD

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Background: Neonatal lupus erythematosus (NLE) is a passively acquired autoimmune disease, typically caused by maternal anti-Ro and anti-La antibodies in infants from mothers with Systemic Lupus Erythematosus (SLE) and Sjogren's Syndrome (SS). Classically, it is associated with a discoid rash and congenital heart block. Less recognized symptoms of NLE include brain, liver and hematologic abnormalities. Here we review NLE cases diagnosed at TCH.

Materials/Methods: Using the EPIC SlicerDicer tool, we searched for cases of NLE treated from 1/1/2011 – 11/30/2021. Of 39 patients noted, 4 were excluded due to inadequate charting. Using retrospective chart review, data was collected for each infant including sex, ethnicity, age at presentation, maternal diagnosis, rash findings, lab abnormalities, EKG results and development of future autoimmune disease.

Results: Of the 35 cases, 17 were male. Ethnic distribution included 34.3% African American, 28.5% Caucasian, 20% Hispanic, 11.4% Asian and 5.7% biracial. The median age of presentation was 6 weeks. Most moms had diagnoses of SLE or SS (n=23) vs those without known autoimmune disease (n=12). Antibodies were found in 21/23 patients tested, including 1 with anti-Smith antibodies. Most NLE cases had discoid rash (n=33) and 5 required diagnostic skin biopsy. Hematologic abnormalities included 10 infants with neutropenia (ANC <1,000), 1 with severe thrombocytopenia requiring platelet transfusion and 2 with anemia. Transaminitis developed in 13 patients. Cardiac involvement occurred in 6 infants: 4 had complete heart block requiring pacemakers, of which 1 required a VAD, and 1st-degree heart block resolved in 2 cases. A rare manifestation included a child with obstructive hydrocephalus that improved without intervention. One child developed JIA, another autoimmune neutropenia and a third persists with elevated anti-Smith antibodies.

Conclusions: NLE is an uncommon diagnosis and can be devastating in patients with complete heart block. While skin manifestations are the most common clinical feature, they are typically self-limited and may be mistaken for fungal infections or eczema. Less commonly recognized manifestations include neutropenia and transaminitis requiring possible steroid therapy. Most patients do well with symptoms resolving by their 1st birthday. NLE should be considered in an infant with discoid rash or heart block even in mothers without SLE or SS. NLE patients may be at higher risk of developing future autoimmune disease and benefit from monitoring.

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