

Clinical Characters of Neonatal Lupus Erythematosus

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DESCRIPTION

Subacute Cutaneous Lupus Erythematosus (SCLE) skin lesions and congenital heart block are the most common symptoms of Neonatal Lupus Erythematosus (NLE). Anti-Ro/SSA, anti-La/ SSB, or anti-U1RNP autoantibodies are present in the maternal blood. Anti-Ro/SSA autoantibodies are the most common, accounting for around 95% of cases. Autoantibodies are passed from mother to kid through the placenta. When maternal autoantibodies are no longer detectable in the newborn, the skin condition goes away. NLE provides the strongest clinical evidence that autoantibodies are involved in at least some aspects of lupus erythematosus, but there is still no conclusive proof that autoantibodies are involved in the disease process. Skin illness normally appears after birth, is temporary, and leaves no scars. Cardiac illness starts in the womb, and heart blocks are virtually usually permanent. Many babies need pacemakers, and roughly 10% of them die from cardiac illness problems. Transient liver damage or thrombocytopenia has been found in certain cases.

NLE patients normally have a normal infancy but may develop autoimmune disease later in life. It's unclear whether the later onset of autoimmune illness is a common or exceptional occurrence. Mothers of babies with NLE may be asymptomatic at first, but eventually develop autoimmune disease signs. Sjogren's syndrome is the most common constellation of symptoms in our group of about 30 moms of babies with NLE. The majority of newborns that are exposed to anti-Ro/SSA autoantibodies during pregnancy do not develop NLE. There is no test to predict which babies will be impacted in the future. Treatment during pregnancy is still debatable, and if used, it should be reserved for foetuses with potentially fatal diseases. Topical treatment for skin condition and, if necessary, pacemaker implantation for heart block are the only treatments available after birth. For severe interior disease, systemic steroids may be prescribed.

The heart of a newborn with neonatal lupus erythematosus is usually structurally normal. Congenital atrioventricular block, which can emerge as first, second, or even third-degree AV block,

is the most common and characteristic cardiac presentation. Usually appears between 18 and 24 weeks of pregnancy. Sinus bradycardia, QT prolongation, cardiomyopathy, congestive heart failure, myocarditis, and structural or valvular abnormalities are some of the other cardiac manifestations (ventricular septal defects, ostium secundum type atrial septal defects, patent foramen ovale, patent ductus arteriosus, pulmonary stenosis, pulmonary valve dysplasia, fusion of the chordae tendineae of the tricuspid valve). In pregnancy, second- and third-degree AV block is present with foetal bradycardia and a ventricular rhythm of 40 to 80 beats per minute. The heart rate is fewer than 100 beats per minute after birth, which is rarely associated with indications of congestive heart failure (diaphoresis, pallor, peripheral edema, prominent jugular veins and crackles on auscultation of the lungs). Nonspecific indications such as intermittent gallops, fluctuation in the loudness of the first heart sound, intermittent cannon waves, and murmurs can be detected during cardiac auscultation. A severe AV block, which causes myocardial dysfunction due to endocardial fibroelastosis and myocardial fibrosis, is related with right ventricular pacing and consequent ventricular asynchrony and dysfunction in 5% to 10% of neonates.

NLE neonates should be treated in a tertiary care facility. Participation of a multidisciplinary team is also possible. Patients with NLE who have cardiac involvement need to be monitored on a frequent basis to determine heart function and the necessity for a pacemaker. For those who are unable to adjust for a sluggish heart rate, a pacemaker is frequently required. Serial echocardiogram should also be scheduled to check for a prolonged PR interval. If the cardiac involvement is severe, the young child's activity may need to be limited.

Sunscreens may be beneficial in the treatment of cutaneous lupus erythematosus, but a newborn is less likely to be overly exposed to the sun. Solar exposure, however, should be avoided if at all feasible. Parents should be urged to apply sunscreen well before sun exposure and to use a sunscreen with a high SPF that is water resistant and gives broad-spectrum (UV-A) coverage. Solar avoidance should be advocated as a behaviour modification. Clothing that provides protection is highly

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desirable. Disease prevention strategies that avoid irreversible scarring are a top focus. Mild topical corticosteroids can be used to treat NLE skin lesions. Antimalarial drugs have the potential for toxicity and have a slow onset of effect, thus they are probably not appropriate for treating this temporary condition. For remaining telangiectasia, laser therapy may be considered. In the treatment of NLE, systemic corticosteroids and immunosuppressive drugs are generally not recommended. Children with NLE require on-going monitoring, particularly before adolescence and if their mother has an autoimmune condition. Even if the child is not at a higher risk of developing SLE, the onset of an autoimmune disease in early childhood should be a cause for concern.

Neonatal Lupus Erythematosus is a clinical syndrome characterised by cutaneous, cardiac, and systemic problems in newborns whose mothers have autoantibodies to Ro/SSA and La/SSB. The disorder could have significant consequences. Neonates with NLE should be treated at a tertiary care facility, and multidisciplinary team participation may be required.